

MISCELLANEOUS

M1 Acquired Hemophilia: A Report of Three Cases

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INTRODUCTION: Acquired hemophilia is a rare disorder, which occurs in 1 to 4 per million population. It is characterized by the development of auto-antibodies against the coagulation protein, Factor VIII. We report three adult Filipino female patients who develop bleeding diathesis which subsequently were found to be due to acquired inhibitors against Factor VIII.

CLINICAL PRESENTATION AND DIAGNOSIS: Patient A is a 28-year old female previously diagnosed with SLE who was admitted for hematuria following 7 months of vaginal bleeding, gum bleeding and easy bruisability. She was pale on admission with hematomas on the trunk and extremities. Tests for lupus anticoagulant (aPTT, KCT, dRVVT) were prolonged but an assay for Factor VIII inhibitor showed levels of 2.8 Bethesda units (BU)/ml.

Patient B is a 28-year old previously healthy female who presented with hematomas on the extremities and hemarthroses 10 days prior to admission. She was pale on admission, with hematomas on the trunk and extremities, and hemarthroses of the right shoulder and the left knee and foot. Tests for lupus anti-coagulant (LAC) were prolonged but an assay for Factor VIII inhibitor showed levels of 25 BU/ml.

Patient C is a 49-year old previously healthy female who presented with bilateral leg swelling and hematuria following 3 months of recurrent hematoma formation and easy bruisability. Aside from being pale and with hematomas on the trunk and extremities, she later developed hemarthroses of the right ankle and left hip. Kaolin Clotting Time was prolonged and the assay for Factor VIII inhibitor showed levels of 1.25 BU/ml.

MANAGEMENT AND OUTCOME: All patients received transfusion of blood products to acutely manage the bleeding. Steroids were started at 1 mg/kg/day following pulse doses of methylprednisolone. Patients B and C received additional immunosuppression in the form of Azathioprine. All patients were discharged improved following control of the bleeding diathesis.

SIGNIFICANCE OF THE STUDY: Acquired hemophilia is a potentially life-threatening disorder, with a mortality rate of 15-22%. Prompt recognition and treatment is necessary to avert serious complications.

M2 Pneumomediastinum in a Patient with Interstitial Lung Disease Associated with Dermatomyositis

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Poster Presentation. 12th PRA Annual Convention. Mactan Cebu. Jan 2005

OBJECTIVE: To present a case of pneumomediastinum in a patient with interstitial lung disease and associated dermatomyositis.

STUDY DESIGN: Case Report

SETTING: University of Santo Tomas Hospital

CASE SUMMARY: A thirty two year old female Filipino nurse presenting with slight dyspnea and crepitation on the anterior chest and neck area. She is a diagnosed case of dermatomyositis with interstitial lung disease, maintained on prednisone. Physical examination was showed presence of heliotrope rash, Gottrons sign, presence of V-sign, shawl sign and holster sign. Palpation on the neck showed crepitations on the anterolateral portion of the neck. Auscultation revealed bibasal crackles. She has no proximal muscle weakness. High Resolution of chest CT scan showed pneumomediastinum with subcutaneous emphysema on both sides of the neck, pulmonary emphysema, chronic interstitial lung disease with interstitial infiltrates. Management was supportive with patient undergoing methylprednisolone pulse therapy and patient was discharged improved.

DISCUSSION: Pneumomediastinum is a rare occurrence among patients with dermatomyositis. Predisposing factors include presence of concomitant interstitial lung disease, young age, on chronic steroids and presence of cutaneous vasculopathy. Pathogenesis include infections, disruption of mucosal barrier, alveolar rupture due to an increase in intra-alveolar pressure or a decrease in peribronchovascular interstitial pressure. Treatment is largely supportive.

M3 Antiphospholipid Antibody Syndrome in a Patient with Dermatomyositis: A Case Report

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OBJECTIVE: This case report aims to describe the clinical course and treatment of a rare case of a patient with an overlap of dermatomyositis and antiphospholipid antibody syndrome.

SUMMARY: This is a case of a 29 year old female who initially presented with a history of fever, proximal muscle weakness, and appearance of Gottron's papules and rashes on the face, trunk and extremities. Pertinent work-ups included a positive antinuclear antibody, anti-Jo1 and anti-RNP antibodies. CK-MM level was elevated and muscle biopsy was consistent with an inflammatory myopathy. Patient was then diagnosed to have dermatomyositis and was treated with corticosteroids. Six years after the diagnosis, she had onset of progressive dyspnea and Raynaud's phenomenon. Work-up done revealed moderate pulmonary hypertension on 2D-echo and pulmonary scintigraphy showed findings consistent with pulmonary thromboembolism. Activated partial thromboplastin time was prolonged and did not correct with mixing. Kaolin clotting-time test and dilute Russell's viper venom time were also both prolonged. The patient was treated with Enoxaparin and later shifted to high-intensity coumadin. A vasodilator and a diuretic together with steroids were also given. She had regular follow-up with a local physician with an INR ranging from 3-4. Despite this, patient had recurrence of dyspnea and eventually expired.

CONCLUSION: The course of a patient with the novel overlap of dermatomyositis and antiphospholipid antibody syndrome was presented. This is the second case in literature of the concurrence of dermatomyositis with APAS. She was treated with corticosteroids and other immunosuppressive drugs for recurrent flares of dermatomyositis however the continuous state of hypercoagulability secondary to the lupus anticoagulants resulted in the patient demise despite treatment with coumadin. Future studies may provide which therapy is best for this overlap syndrome and determine the definite pathogenesis and prognosis in patient with this association.

M4 Clinical Profile of Patients with Polymyositis/Dermatomyositis Seen at UP-PGH

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OBJECTIVES: To describe the clinical features of patients with polymyositis/dermatomyositis (PM/DM) and to determine the laboratory features, treatment modalities and outcome of these patients.

METHODS: Case records of patients with polymyositis/dermatomyositis seen at University of the Philippines-Philippine General Hospital between 1995 to 1999 were retrieved. Included were those with a final diagnosis of polymyositis/dermatomyositis using the criteria of Bohan and Peter in 1975. Data were extracted and recorded using a chart review form.

RESULTS: Based on our census, there were 31 patients diagnosed to have PM/DM from 1995 to 1999. Of these, 25 (81%) of the charts were retrieved. Twenty-two patients had dermatomyositis; 12 (48%) of whom having a definite DM, 6 (24%) with probable DM, and 4 (16%) with a possible DM. Three patients had polymyositis with one patient with possible PM. Mean age of patients diagnosed to have PM/DM was 43 years. There were more females (18) than males (7). The most common clinical manifestations were proximal muscle weakness as seen in 23 (92%) of patients, dysphagia in 17 (56%), and fever in 11 (44%). Heliotrope rash was the most common skin manifestation found in 11 (50%) of DM patients, followed by Gottron's sign in 9 (36%). Systemic lupus erythematosus was the most common associated connective tissue disease. Elevated CK/ MM/Total level was the most common laboratory finding as seen in 21 (84%) of patients. Prostatic cancer, papillary adenocarcinoma of the ovaries and acute myelogenous leukemia were malignancies identified in 4 (16%) of the patients.

Treatment entailed use of steroids in 23 (92%) of patients while combination treatment using steroid methotrexate was given in 1 patient. In this series, 14 (56%) of patients improved, 5 (20%) went on remission, 1 (4%) had no improvement, 2 (8%) died of pneumonia, and 3 (12%) went home against advice.

CONCLUSION:

In this review, DM was more commonly observed than PM. The usual patient is female in the 5th decade of life. Common clinical manifestations were proximal muscle weakness, dysphagia, heliotrope rash and elevated Creatinine Kinase MM/Total. The usual treatment regimen involved the use of steroids with good results.

M5 Evidence-based Management of Chronic Low Back Pain

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In the review of literature on the management of chronic low back pain, there were many pharmacologic and non-pharmacologic therapeutic modalities that have been extensively investigated. However, in these trials, there were study design issues identified that have led to inappropriate comparisons among studies or incorrect conclusions.

The author reviewed the current rational evidence-based therapeutic approaches to the treatment of chronic low back pain. Beneficial effects were seen with exercise, the use of nonsteroidal anti-inflammatory drugs, opioids, anti-depressants, herbal medicine, massage, acupuncture, use of back schools and intensive multidisciplinary programs. There were mild beneficial effects seen with the use of spinal manipulation or changing behavior while injection therapy, transdermal electrical stimulation (TENS) and the use of lumbar corsets showed undetermined efficacy

M6 Tuberculosis Among Filipino Patients with Rheumatic Diseases on Infliximab Therapy

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14th PRA Annual Convention. Cagayan de Oro. Jan 2007

OBJECTIVE: Biologic agents are expected to provide superior efficacy over traditional disease modifying drugs but with better safety profile. However, there is cause for concern for susceptibility to tuberculosis (TB) infection or reactivation among patients on TNF inhibitors, which may have a special impact in endemic countries like the Philippines. This paper describes the spectrum of tuberculosis infections in a cohort of Filipino patients who had used infliximab for a rheumatic disease indication.

METHODS: Retrospective case - cohort. Cases of tuberculosis (TB) were obtained from a registry of Filipino patients who had received infliximab therapy for a rheumatic disease indication from January 2002 to December 2005. Tuberculosis (TB) infection was defined as any of the following: (a) active TB: based on typical infiltrates by chest x-ray, positive acid-fast bacilli (AFB) smears of sputum or other specimens, histopathological findings of granulomatous inflammation with/ without caseation necrosis, or clinical symptoms strongly suggestive of TB infection e.g. prolonged fever, with clinical response - adjudged by the attending rheumatologist - to at least one month of anti-TB treatment; (b) latent TB: by a positive tuberculin skin test (TST ≥ 8 mm), absent clinical symptoms and normal chest x-ray; (c) TB exposure: known exposure to household member with active TB, TST < 8 mm or unknown, absent clinical symptoms; and (d) previous TB: a history of active TB which was adequately treated, absent clinical symptoms.

RESULTS: Among the 62 Filipino patients with rheumatic disease who underwent screening for tuberculosis and received infliximab, 2 had prior treated TB and 4 had a TST ≥ 8 mm with normal chest x-ray. The latter were placed on isoniazid prophylaxis. Among the 64 patients who underwent infliximab therapy, 5 (7.8%) patients developed active tuberculosis (TB) while receiving maintenance doses of

infliximab therapy. The dose of infliximab ranged from 2 to 6 mg/ kg per infusion, and duration from infliximab initiation to TB diagnosis averaged 6.7 months + 5.4 SD (range 3 to 15 months). Sites of TB included pulmonary, submandibular lymphadenopathy and an abnormal TST with or without fever or chest x-ray findings. Infliximab was discontinued in all patients as they completed anti-TB therapy; all patients responded satisfactorily to at least six months of anti-TB therapy.

SUMMARY: Although tuberculosis remained a risk and adverse event, this did not seem to significantly impact on morbidity provided vigilance was exercised in monitoring, with prompt initiation of anti-TB therapy.

M7 National Nutrition and Health Survey (NNHeS): Prevalence of Rheumatic Diseases among adult Filipinos

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Phil J of Internal Medicine Nov-Dec 2006; 44(6); 297-303.

OBJECTIVE: The objective of this study is to determine the prevalence of arthritis and specific rheumatic diseases (osteoarthritis, rheumatoid arthritis, gout, systemic lupus erythematosus) in the Philippines.

METHODOLOGY: Using a multi-staged cluster sampling methodology, this study evaluated 4753 adults aged 20 or more, from 2636 households, 79 provinces and 17 regions. The survey instrument was a validated interviewer-assisted questionnaire, containing items modified from a pre-validated ILAR-COPCORD screening instrument for musculoskeletal complaints.

RESULTS: The prevalence of arthritis in general was at 6.5%. The prevalence rates of osteoarthritis, rheumatoid arthritis and gout in adult Filipinos were 0.5%, 0.6%, and 1.6% respectively. No case of systemic lupus erythematosus was identified.

DISCUSSION: These data on rheumatic diseases from the second NNHeS may be used for policy formulation and program prioritization. In addition, the information may serve as baseline measures to evaluate the burden of illness brought about by musculoskeletal complaints. A third NNHeS is planned in the year 2008.

Keywords: rheumatic diseases, osteoarthritis, rheumatoid arthritis, gout, systemic lupus erythematosus, survey, prevalence, Philippine arthritis prevalence

M8 Rheumatologic Manifestations of Renal Disease at the University of Santo Tomas Hospital

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Results presented at the 12th APLAR meeting 2-5 August 2006, Kuala Lumpur Malaysia

OBJECTIVE: Determine the prevalence of rheumatologic manifestations in patients with renal disease

DESIGN: 5-year review of registry of renal disease patients with rheumatologic manifestations from the Sections of Nephrology and Rheumatology at the University of Santo Tomas Hospital (2001-2006)

MAIN OUTCOME MEASURES: Frequency of rheumatic disease manifestations in patients with acute renal failure, chronic kidney disease, and post renal transplant patients

RESULTS: Included were data from 69 patients with renal disease classified into azotemia (30 patients), chronic kidney disease (CKD) < 5 years and > 5 years (29 and 10 respectively). There was no renal transplant patient. The mean age \pm SD was 61.5 \pm 16.7, 55 \pm 14 and 58 \pm 14 among azotemic patients, CKD < 5 years and > 5 years respectively. The mean disease duration \pm SD among the azotemic patients was 56.6 \pm 41.5 months while those with CKD were 53 \pm 55 and 124 \pm 67 months. The mean creatinine level \pm SD was 2.74 \pm 1.5, 8.3 \pm 3.9 and 5.7 \pm 3.3 mg/dl among azotemic patients, CKD of < 5 years and > 5 years respectively. The most common co- morbid conditions included hypertension (55%) and diabetes mellitus (37%). Among the azotemic patients, the most common rheumatologic manifestations were

metabolic bone disease (36 %), soft tissue rheumatism (STR) (26%), and crystal induced arthropathy (23%). Metabolic bone diseases included osteoporosis and calciphylaxis. Among CKD patients > 5 years disease duration, the most common manifestation was metabolic bone disease (60%), crystal induced arthropathy (20%) and STR (20%) while in patients with < 5 years disease duration, crystal induced arthropathy (45%) was the predominant condition, STR 14%, and infections 14%. The most common infection was osteomyelitis.

CONCLUSION: Metabolic bone disease appears to be the most common rheumatologic manifestation among renal disease patients seen in our institution. This review recommends early identification and initiation of preventive measures and effective management schemes for this complication.

M9 Orbital Myositis Successfully Treated with Steroids and Cyclophosphamide: a Report of Two cases

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1. **Poster Presentation. 13th PRA Annual Convention. Manila, Jan 2006**
2. **Poster Presentation. 12th APLAR Congress. Kuala Lumpur, Malaysia, August 2006**
3. **APLAR Journal of Rheumatology 2006; 9(suppl.1 P329):A156.**

OBJECTIVE: To present two cases of idiopathic orbital myositis successfully treated with systemic steroids and oral cyclophosphamide.

DESIGN/SETTING: A case series of two private outpatient cases in a tertiary hospital

CASE 1. A 37 year old female with periorbital pain and swelling of the right eye later involving the left eye, accompanied by diplopia of one year duration. MRI showed right medial rectus hypertrophy, suggestive of pseudotumor (myositis) of the right orbit. She had received high dose prednisone, methotrexate and azathioprine without benefit. On referral to Rheumatology, she was given 3 daily doses of pulse methylprednisolone 1 g/day, and started on cyclophosphamide 150 mg daily; prednisone was continued at 50 mg/day. By the 12th week of therapy, she had decrease in pain and swelling of both eyes with improved visual acuity. She no longer experienced diplopia, and prednisone had been tapered to 20 mg/day. She regained full motility of her left eye, although the right eye remained immobile to the right external gaze. By the 24th week, the left eye was totally asymptomatic, with improved vision and extraocular motility of the right eye, and prednisone had been tapered off. She is now maintained on 50 mg/day of cyclophosphamide.

CASE 2. A 31-year old male with sudden onset of periorbital pain and swelling, ptosis, chemosis with some extraocular motility limitation of the right eye. MRI revealed enlarged extraocular muscles on the right more than the left. He had received high dose prednisone and radiotherapy with transient improvement. On referral to Rheumatology, he was given pulse methylprednisolone 1 g/day for 3 days, started on cyclophosphamide 150 mg daily, and continued on prednisone 60 mg/day. By the 8th week, he had decreased periorbital pain and swelling, decreased chemosis and ptosis with increase in right orbital aperture, and improvement of the extraocular muscle motility. Prednisone was tapered off, and he is currently maintained on 150 mg/day of cyclophosphamide.

SUMMARY: We presented two Filipino patients with idiopathic orbital myositis unresponsive to high-dose daily steroids combined with methotrexate or azathioprine, and radiotherapy. Both patients showed satisfactory clinical improvement with pulse methylprednisolone and oral cyclophosphamide.

Research Grant: None

M10 Aerosolized Iloprost in Pulmonary Hypertension Secondary to Connective Tissue Diseases: A Philippine General Hospital Experience

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1. **Future in Rheumatology: From Bench to Bedside. (Abstract Book P275). APLAR 2004. p.150**

2. **Annals of the Rheumatic Diseases. The EULAR Journal. July 2007; 66 (Supplement II SAT 0159): 477**
3. **Poster Presentation. EULAR Congress. Barcelona, Spain, June 2007**
4. **Oral Presentation. 13th PRA Annual Convention. Manila. Jan 2006**

BACKGROUND: Several studies have shown that aerosolized iloprost, a stable analogue of prostacyclin, causes a reduction in the mean pulmonary artery pressure (PAP) and improvement in the clinical parameters among patients with pulmonary hypertension.

OBJECTIVE: To report a local experience with inhaled iloprost in secondary pulmonary hypertension.

METHODS: This is a descriptive study of 6 cases admitted at the UP-PGH, pay and charity wards, from 2003-2005. Chart reviews were done and demographic data, primary diagnosis, pulmonary hypertension defined as a pulmonary artery pressure of >20 mmHg at rest, and functional classification utilizing the New York Heart Association (NYHA) classification and the 6 minute walk test (SMWT) are described. Changes in the NYHA functional classification, PAP by 2D echocardiography and SMWT, pre and post-inhalational therapy are hereby reported.

RESULTS: Six patients with secondary pulmonary hypertension (4 scleroderma and 2 systemic lupus erythematosus (SLE) were included in the review. Two scleroderma patients had the CREST syndrome. Diagnoses were based on the ACR Criteria for classification of SLE and ACR Criteria for Diagnosis of Scleroderma. All were females and mean age was 41 years. All patients had breathlessness and by NYHA classification, 1 was Class IV, 2 were Class III and 3 cases, Class II. All patients had a PAP of >20 mmHg at rest, with mean of 41mmHg (lowest of 40 and highest of 70mmHg).

All patients received inhaled iloprost at a dose of 2.5-5.0 ug every 3 hours except during sleeping hours for an average of 5 doses a day using an ultrasonic nebulizer provided Schering Philippines Corp.

Only 2 patients had recorded SMWT pre and post treatment. Both recorded 30% improvement of distance covered. This test was withheld in the Class IV patient, but post treatment she was able to cover 20 metres in 3 min. One other patient had chest pain and refused the test. All patients showed a reduction in the PAP and improvement in the NYHA functional class. Among the side effects noted were cough, flushing, jaw claudication case and dizziness which resolved on decreasing the dose.

CONCLUSION Aerosolized iloprost showed beneficial symptomatic effects in this series of 6 cases and has mild, transient side effects. This drug has a potential for use in patients with secondary pulmonary hypertension from Connective Tissue Diseases.

M11 Pulmonary Arterial Hypertension Among Filipino Patients with Connective Tissue Disease

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1. **Oral Presentation. 50th Annual General Assembly & Scientific Meeting of Japan College of Rheumatology (JCR). Nagasaki, Japan. 2006**
2. **Poster Presentation. 13th PRA Annual Convention. Westin Hotel, Manila. Jan 2006**
3. **Modern Rheumatology Suppl 16:159,2006**

OBJECTIVE. To describe the clinical features, various therapies used, and clinical course of pulmonary arterial hypertension in a group of Filipinos with connective tissue diseases.

DESIGN: Case series

SETTING: Cases seen at 2 tertiary care hospitals: Section of Rheumatology, Clinical Immunology and Osteoporosis at University of Santo Tomas Hospital, and Section of Rheumatology, Allergy and Immunology at the St. Luke's Medical Center.

PATIENTS/PARTICIPANTS: We reviewed the records of patients diagnosed with pulmonary arterial hypertension (PAH), defined as a systolic pulmonary artery pressure (sPAP) greater than 30mmHg measured by two dimensional echocardiogram with Doppler study, looking into their presenting features, underlying connective tissue disease, pharmacologic intervention, and clinical course

RESULTS: A total of 31 patients were identified and included in the analysis, of which there were 30 females and 1 male. There were 13 patients with systemic lupus erythematosus (SLE), 10 with scleroderma, and 7 with mixed connective tissue disease (MCTD) and 1 with primary antiphospholipid

syndrome (APS). The average age at PAH diagnosis was 37 ± 13 years (mean \pm SD) with a range of 14 to 62 years and mean duration of illness from CTD to PAH diagnosis was 51 ± 53 months with a range 0 to 224 months. Twelve (12) patients had died at the time of this report, with a median duration of 15.5 months (range 1 to 57 months) from PAH diagnosis to mortality; of these, 6 had scleroderma, 5 had SLE and 1 with primary antiphospholipid syndrome. The following therapies were used in this group of patients: low molecular weight heparin, warfarin, calcium-channel blockers, aspirin, cyclophosphamide pulse therapy, bosentan, intravenous or inhaled iloprost, and sildenafil.

CONCLUSION: We have described the clinical profile of pulmonary arterial hypertension in a group of Filipino patients with connective tissue disease, most commonly SLE. Varied forms of pharmacologic therapy were used among these patients. Mortality remains high, particularly among those with underlying scleroderma. Early recognition and treatment are crucial in order to provide a better outcome for these patients.

M12 Clinical Experience with Infliximab among Filipino Patients with Rheumatic Diseases

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1. **APLAR Journal of Rheumatology 2006;9: 150-56**
2. **Paper Presentation. 13th PRA Annual Convention. Westin Hotel, Manila. Jan 2006**

AIM. To describe the clinical experience with infliximab among Filipino patients with rheumatic diseases, specifically disease indications, dose regimens, clinical response, and adverse events.

METHODS. We reviewed the data of Filipino patients who were given infliximab by rheumatologists for a rheumatic disease indication. The case report form included demographic profile, underlying rheumatic disease, co-morbidities, concurrent medications, dose and frequency of infliximab, clinical efficacy, and adverse events. The frequency of doses, intervals between doses, and discontinuation status were recorded.

RESULTS. Included were 64 patients (35 females), with a mean age of 44 years. Most (41%) had rheumatoid arthritis, followed by psoriasis/ psoriatic arthritis (31.2%) and ankylosing spondylitis (17.2%). Average disease duration from diagnosis to initiation of infliximab therapy was 7.6 years \pm 6.7 SD. Among 35 patients, the interval between maintenance infusions ranged from 6 to 13.6 weeks, with a mean of 8.27 weeks. Clinical response was good to excellent in more than 80% of patients. Discontinuation rate was 10.9% and 28.1% at 3 and 12 months, respectively. Infusion-related adverse events were mild and transient, and 14 (21.8%) cases of infection resolved with appropriate therapy. Infliximab was temporarily withheld in 5 (7.8%) patients with active tuberculosis.

SUMMARY. These findings substantiate the superior clinical efficacy of infliximab and manageable adverse events among Filipinos with rheumatic diseases. It also demonstrated dose regimens in clinical practice in a third world setting with limited resources.

M13 Philippine Guidelines on the Screening for Tuberculosis Prior to the Use of Biologic Agents

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1. **APLAR Journal of Rheumatology 2006;9:184-192**
2. **Oral Presentation. 12th APLAR Congress. Kuala Lumpur, Malaysia, August 2006**

AIM: To develop practice guidelines in tuberculosis screening of patients and their households and close contacts, prior to the use of biologic agents.

METHOD: A technical research committee formulated an evidence-based raft, based on existing literature regarding the tests used in tuberculosis screening among immunocompromised patients. The evidenced-based draft was then circulated to an expert panel. An *en banc* meeting of the panelists was held and a

consensus was declared if more than 50% agreed on a recommendation. Issues not resolved by consensus were discussed by correspondence and voted upon. The guidelines were presented in a public forum and feedback by stakeholders were reviewed and integrated into the final draft.

RECOMMENDATIONS: 1. Patients for biologic therapy should be screened for latent and active tuberculosis prior to initiating treatment. 2. All patients who are candidates for biologic agents should be screened by tuberculin skin test for latent TB, and a chest radiograph for active tuberculosis. 3. Household and close contacts of candidate patients should be screened for active tuberculosis. 4. All household and close contacts of candidate patients should be screened for active TB using chest radiograph. 5. Treat latent and active tuberculosis according to local guidelines. 6. Delay treatment with biologic agents in patients with latent or active tuberculosis. 7. Administer tuberculosis prophylaxis to the patient for biologic therapy exposed to household contact with active tuberculosis.

CONCLUSION: These recommendations emphasize the importance of screening patients, household and close contacts for latent and active tuberculosis prior to initiating biologic therapy.

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M14 The Long and Winding Road: A Case of Relapsing Polychondritis

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13th PRA Annual Convention. Westin Hotel, Manila. Jan 2006

Relapsing polychondritis (RP) is a unique, rare autoimmune disorder in which the cartilaginous tissues are the primary targets of destruction but the immune damage can spread to involved noncartilagenous tissues like the kidney, blood vessels, and so forth. The annual incidence in Rochester, Minnesota was noted to be 3.5 cases per million. Buckner noted an incidence of 4 cases per million in the Pacific Northwest. The manifestations of the disease can take many different forms and the pathogenesis is still unclear. It may occur in a primary form or it may be associated with other disease states.

This is a case of 15 yo male, who was earlier diagnosed as juvenile chronic arthritis since the age of nine. He initially presented with clinical features consistent with juvenile chronic arthritis and treated as such. He was admitted several times in the past due to fever to fever, joint pain and swelling. Several years later, he had involvement of the aural and nasal cartilages resulting floppy ears and saddle nose deformity (figure 3 & 4). This time a diagnosis for relapsing polychondritis was made according to the diagnostic criteria of *Macadam et al* (Table 2).

This case report highlights the difficulty of diagnosing this extremely rare disease and summarizes important aspects of relapsing polychondritis with a focus on recent information regarding clinical manifestations would assist in its timely diagnosis, thereby improving the prognosis and quality of life for affected patients.

M15 Neuropathic shoulder and elbow arthropathy associated with Syringomyelia and Arnold-Chiari Malformation

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Poster Presentation. 12th PRA Annual Convention. Cebu. Jan 2005

Neuropathic arthropathy of the upper extremity joints is a rare disease, usually associated with syringomyelia in 25% of cases. Arnold-Chiari malformation associated with syringomyelia causing

neuropathic arthropathy is infrequently reported. We present a case of neuropathic shoulder and elbow arthropathy secondary to syringomyelia associated with Arnold-Chiari Malformation.

A 57-year-old female had a ten-year history of numbness and weakness of the right arm, with deformity of the shoulder and elbow. She also had right-sided hearing loss and imbalance. She denied any trauma. Examination showed muscle atrophy, decreased strength and sensation, swelling and limited range of motion of the shoulder and elbow. She had a positive Romberg sign and wide-based gait. Laboratory tests were normal. Radiographs showed destruction, degenerative changes, bone fragmentation affecting the shoulder and elbow joints; and dislocation of the humeral head. Nerve conduction studies showed right brachial plexopathy. Magnetic resonance imaging (MRI) showed a C2-T4 syrinx, Arnold-Chiari malformation and cerebellar tonsillar herniation. Patient underwent decompressive suboccipital craniectomy, C1-2 laminectomy and duraplasty. One month later, patient had partial recovery of strength, and improved hearing and balance.

Neuropathic arthropathy develops in 25% of cases with syringomyelia. Mostly affecting joints of the upper extremity, the usual presenting feature is joint swelling. Diagnosis is usually delayed. Radiographs are notable for bone destruction and joint disorganization. Treatment requires management of the underlying disease with functional rehabilitation of the affected joints. Less than five cases of syringomyelia with Arnold Chiari Malformation causing neuropathic arthropathy have been reported. We conclude that Arnold-Chiari associated with syringomyelia is a rare cause of neuropathic arthropathy. Upper extremity neuropathic arthropathy warrants investigation of the spinal cord by MRI to assess for causal lesions

M16 Hereditary Multiple Exostoses in a 27 Year-old Female: A Case Report

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Poster Presentation. 12th PRA Annual Convention. Cebu. Jan 2005

OBJECTIVE: To present a case of hereditary multiple exostoses in a Filipino patient.

STUDY DESIGN: Case report

SETTING: University of Santo Tomas Hospital

CASE SUMMARY: A 27 year old saleslady, female was admitted because of fever and myalgia. Physical examination revealed hyperemic conjunctiva and flushed face. Musculoskeletal examination of the extremities, showed the presence of bony protuberances, fixed and hard, found on both forearms, distal portion of left thigh and on both shoulders. History showed the presence of prominent bony protuberances in other members of the family. She was treated empirically as Upper Respiratory Tract Infection. Plain Radiographs of upper and lower extremities revealed multiple bony exostoses on the metaphyses of both femurs, which was diagnosed to be osteochondroma. Pertinent family history and multiple bony exostoses found comprises a diagnosis of Hereditary Multiple Exostoses.¹ Watchful expectancy of complications were advised to the patient.

DISCUSSION: Hereditary multiple exostosis is an autosomal dominant condition which affects mostly males in the family. It is more common among those with Caucasian ancestry. Management is usually conservative with surgical intervention done to relieve complications arising from compressive symptoms. Malignant degeneration is rare. Asian patients afflicted with these conditions are uncommon with case reports affecting mostly Taiwanese and Indian ancestry.

M17 Infliximab Therapy for Filipinos with Rheumatic Disease: Preliminary Experience with Three Patients

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Poster Presentation. 11th PRA Annual Convention. Manila Hotel. Jan 2004

OBJECTIVE: To describe the clinical response to infliximab (IFX) therapy of three Filipinos with rheumatic disease refractory to traditional treatment.

METHODS: We reviewed three patients with rheumatic diseases: two males with ankylosing spondylitis (AS) and one female with rheumatoid arthritis (RA), vasculitis, and ulcerative colitis (UC), who were refractory to conventional treatment with non-steroidal anti-inflammatory agents (NSAIDs), disease modifying anti-rheumatic drugs (DMARDs) and steroids, and who received IFX therapy. Patients were followed up for a minimum of 42 weeks after initiation of treatment. Response to treatment using Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) in the 2 patients with AS, and Medical Outcomes Study Short Form Health Survey (SF-36) and Health Assessment Questionnaire (HAQ) in all 3 patients and any adverse reaction, infection, or concomitant medical problem were recorded. All patients and all household members were screened for tuberculosis (TB) prior to initiation of IFX therapy.

RESULTS: The three patients showed marked clinical improvement based on BASDAI, HAQ and SF-36 measurements noted as early as the 2nd and 3rd IFX infusion. A rebound of illness was noted in one patient whose interval of infusion was delayed but recovery and improvement immediately followed each subsequent infusion. The same patient developed tuberculosis after the 49th week of IFX therapy requiring temporary discontinuation of therapy.

M18 Bilateral Optic Neuropathy in a Patient with Crohn's Disease: A Case Report

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Rheumatology Section, Department of Medicine UP-PGH, 2004

Crohn's disease is a disorder of unknown cause involving any location of the alimentary tract from mouth to anus. Numerous complications may occur distant from the bowel. Numerous complications may occur distant from the bowel.

OBJECTIVES: To present a case of bilateral optic neuropathy associated with Crohn's disease and to discuss its possible etiopathogenesis, clinical features, diagnostic work-up and management.

CASE: We report a 68-year old woman with bilateral optic neuropathy and biopsy proven Crohn's disease diagnosed 3 months prior to the ocular involvement. Three months prior to admission, patient underwent right hemicolectomy for intestinal obstruction. Histopathologic examination of the specimen disclosed chronic active ileocolitis with ulcers and fissures compatible with Crohn's disease, active phase with reactive epithelial changes; reactive sinus histiocytosis on all 23 lymph nodes. She was discharged with mesalamine (Salofalk). A month prior to admission, she stopped taking mesalamine due to drug unavailability. A week prior to admission, she complained of arthralgia, low grade fever, bitemporal headache, and occasional epigastric pain. Two days prior to admission, she complained of blurring of vision of the right eye on awakening. A day prior to admission, the fellow eye had blurring of vision of the right eye on awakening. A day prior to admission, the fellow eye had blurring of vision on awakening. Fundoscopic examination revealed macular branch artery occlusion of the right eye and optic disc edema of the left eye. Visual acuities were finger counting at 3 feet in the right optic disc edema of the left eye. The rest of physical examination was unremarkable except for cachexia. Laboratory work up showed leukocytosis with neutrophil predominance, elevated ESR, urinary tract infection for which she received fluoroquinolone (ofloxacin). She was started on intravenous methylprednisolone 200 mg every 8 hours for 3 days followed by oral prednisone 60 mg/day. Mesalamine was restarted. Her visual acuity on discharge improved to counting finger at 4 feet in the right eye and light perception on the left eye. She had regular follow up every three months. Prednisone was gradually tapered to present dose of 10mg/day. Her final visual acuity a year after the diagnosis improved to 20/25 in the right eye but the left eye had no light perception. There was no recurrence of Crohn's disease after a year of follow-up.

CONCLUSION: We reported a case of bilateral optic neuropathy and discussed its possible etiopathogenesis and management. Awareness of the association of optic neuropathy and Crohn's disease would promote optimal management.

M19 Antiphospholipid Antibody Syndrome in a Patient with Systemic Sclerosis: a Case Report

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Rheumatology Section, Department of Medicine UP-PGH

Phil J Internal Medicine Nov-Dec 2000. 38(6): 331-334.

OBJECTIVES: This case report aims to discuss the clinical course of a patient with diffuse systemic sclerosis (SSc) with severe pulmonary hypertension and secondary antiphospholipid antibody syndrome (APAS), and to discuss treatment options for a such a case.

SUMMARY: We describe the third case in literature of APAS in a patient with SSc. This is a 40-year-old female who initially presented with a three-year history of Raynaud's phenomenon and digital pitting and a year and a half later developed progressive diffuse skin thickening. She later had a chronic progressive course of thromboembolism, primarily in the lungs, later diagnosed to be due to APAS. The patient was treated for the scleroderma and was anti coagulated for the APAS but still died, underlying the difficulty in diagnosing and treating such rare cases.

M20 Acute Rheumatic Fever in Filipino Adults: A 5-year Review (1994-1998)

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1. **Phil J Internal Medicine Nov-Dec 2000. 38(6): 295-300.**
2. **Published at APLAR Journal of Rheumatology, 1999**

OBJECTIVE: To determine the clinical profile of acute rheumatic fever in adults seen at the University of the Philippines-Philippine General Hospital, Section of Rheumatology, and to compare this data with childhood rheumatic fever and with previous local reports on adults done in 1977.

DESIGN: Descriptive study

METHODOLOGY: All possible cases of acute rheumatic fever seen by the Section of Rheumatology, UP-PGH from 1994-1998, were reviewed. Patients 18 years old and above who satisfied the modified Jones' criteria were included.

RESULTS: Ten of 26 cases reviewed were included. The age range was 20-40 years old, with 60% below 30 years of age. Arthritis was the most common major manifestation (90%) while carditis was seen in only 30%. Erythema marginatum was seen in 30% while subcutaneous nodules and chorea were present in 10%. All patients had fever. Increased ESR and CRP were common (80% and 75%, respectively). A female preponderance was noted. There was no specific seasonal occurrence observed. The incidence of carditis was significantly lower in adults than in children while arthritis was much more common. The other manifestations occurred at about the same rate in both age groups. The previous local report on adult rheumatic fever done by Paulino-Abundo and Carpio noted more carditis and less arthritis compared to our study. Possible explanations to the discrepancies were discussed.

Key words: Rheumatic fever, acute, adult, Filipino

M21 Frequency Distribution of Rheumatic Conditions Seen by Private Specialists: Philippine Rheumatology Association Survey 1998-1999.

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Philippine Rheumatology Association

Phil J. Internal Medicine, 38: 243-247, Sept-Oct, 2000

OBJECTIVE: The study aims to describe the frequency distribution of the different rheumatic diseases seen by medical practitioners (rheumatologists and rehabilitation specialists in particular) in their private practice.

STUDY DESIGN: Descriptive, cross sectional study.

SETTING: Areas of private practice by specialists whether in the city or province (from north to south).

PARTICIPANTS: The respondents included private specialists who were members of the Philippine Rheumatology Association (PRA).

RESULTS: A total of 2,484 patients were seen by the respondents, of which 2,467 patients were considered evaluable. Of the 2,467 evaluable patients, 1,781 (72.2%) had a confirmed diagnosis while 294 (11.9%) had a probable diagnosis. Osteoarthritis (n=525) was the most common rheumatic disease of the 1,781 cases with confirmed diagnosis, while gout (n=385) and soft tissue rheumatism (n=360) were identified as the second and third most common rheumatic diseases respectively. In the pediatric age group with confirmed diagnosis, SLE (n=33) was the most common rheumatic disease followed by JRA (n=30).

CONCLUSION: This series demonstrated the frequency distribution of the different rheumatic diseases seen by medical practitioners (rheumatologists and rehabilitation specialists in particular) in their private practice. Osteoarthritis was the most common among the rheumatic disease followed by gout as most common rheumatic disease. In the pediatric age group with confirmed diagnosis, SLE was the most common rheumatic disease followed by RA.

M22 Functional Disability Profile of Arthritic Patients Seen at the Rheumatology, Outpatient Department, University of the Philippines, Philippine General Hospital

Melissa I. Untalan, MD and Evelyn Osio-Salido, MD
Rheumatology Section, Department of Medicine UP-PGH, 1997

OBJECTIVES: (1) to evaluate the functional disability of patients with arthritis seen at the Rheumatology section, Outpatient department, University of the Philippines, Philippine General Hospital; and (2) to assess the ease of administration of the modified Health Assessment Questionnaire Functional Disability Index Filipino version to patients with arthritis.

METHODS: The modified HAQ FDI Filipino version which was translated and validated in the Philippines in 1993 was used in this descriptive study. All patients at least 19 years old with arthritis for at least 6 months with an arthritic attack at the time of consultation, were included. A one on one interview was conducted by only 1 investigator. The FDI was computed by adding the scores then dividing by the total number of components answered.

RESULTS: A total of 57 consecutive arthritis patients were entered. Seven of these were new patients; 73.7% were females; 75.4% were married. The longest disease duration is 18 years and the shortest is 6 months. Rheumatoid arthritis (40.3%) accounted for most of cases seen, followed by osteoarthritis (19.3%) and gout (17.5%). Three fourths (73.7%) had mild functional impairment and the rest had moderate impairment. No patient had severe impairment. With regards to the instrument itself, it took only 2 to 3 minutes to administer despite some vague terms which needed clarification.

CONCLUSIONS: (1) Patients seen at our section are generally mildly impaired (2). The modified HAQ FDI Filipino version, when administered to a population with arthritis, is easy to administer.

M23 The Prevalence of Rheumatic Diseases in a Filipino Urban Population: a WHO-ILAR COPCORD Study

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Journal of Rheumatology Sept 1997; 24(9): 1814-9.

OBJECTIVE: To determine the prevalence of musculoskeletal complaints and rheumatic diseases in a Filipino urban community

Design: Descriptive cross-sectional two-phase survey design. Phase I (screening) used to face-to-face interview while phase II (examination) involved actual case identification of the rheumatic diseases.

SETTING: An urban community in Metro Manila

PATIENTS/PARTICIPATIONS: Multi-stage cluster sampling of 670 households (3065 adults)

SURVEY METHODOLOGY: A pilot study was conducted to pretest the questionnaire, field procedures, sampling design and data management plan. Standardized translated COPCORD Questionnaire screened the number of cases with musculoskeletal complaints. Identification of cases of rheumatic diseases was based on the American College of Rheumatology epidemiologic criteria.

RESULTS: 3006 completed the questionnaire (phase I-response rate (98%). 489 of the respondents had musculoskeletal complaints. Functional disability was reported in 24% among these respondents. 353 (phase II-response rate 72%) were examined revealing 295 with true rheumatic conditions. Twenty six persons had no abnormalities while 32 had non-rheumatic conditions on physical examination. The most common rheumatic diseases were osteoarthritis (n=124) and soft tissue rheumatism (n=115).

CONCLUSIONS: The prevalence of musculoskeletal complaints was 16% of the adult population in a Filipino urban community. The prevalence of osteoarthritis was 41/1000 adults for soft tissue rheumatism.

M24 Rheumatic Diseases in the Philippines: Validation of the COPCORD CORE Questionnaire Against Physical Examination

Stella Cortes-Fabia, MD, Leonila F. Dans, MD, Llewelyn Hao-Tanopo., MD, Jose Paulo Lorenzo, MD., Josephine Abao-Lim, MD

Rheumatology Section, Department of Medicine UP-PGH, 1993

BACKGROUND: The COMMUNITY ORIENTED PROGRAM FOR THE CONTROL OF RHEUMATIC DISEASES (COPCORD) Core Questionnaire or CCQ has been used as a screening test for Rheumatic Diseases. Several Validation studies in different patient populations have been done showing a Sensitivity of 55-92% and a Specificity of 67-78%.

OBJECTIVE: This study would validate the CCQ in the Filipino Community comparing the Questionnaire against the Physician's Physical Examination as the Gold Standard.

STUDY DESIGN: Descriptive cross-sectional epidemiological survey was done at a hospital setting that is Out-Patient of University of the Phil-Philippine General Hospital.

RESULTS: Of the three hundred respondents, a prevalence rate of 49% was noted. It had a sensitivity of 91% and specificity of 51%. The positive and negative predictive value were 64% and 86% respectively.

CONCLUSION: CCQ has an excellent sensitivity and adequate specificity in screening for Rheumatic Diseases in the Filipino Community.

M25 Outpatient Rheumatic Disease Registry at the University of Santo Tomas Hospital January-December 1994

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Phil J. Internal Medicine, 34: 23-28, Jan-Feb 1996

The distribution of rheumatic diseases may vary depending on genetic, racial, environmental and other factors. This paper describes the prevalence of rheumatic diseases seen at the Rheumatology and Clinical Immunology outpatient services of the University of Santo Tomas Hospital from January to December 1994. Included were all patients whose rheumatologic diagnoses were confirmed by any of the rheumatology consultants or fellows in training.

A total of 2,138 patients (1542 females, 776 males) were entered in the registry. The most common was soft tissue rheumatism (38.6%), of which tendinitis and bursitis were most prevalent. Osteoarthritis made up 18% of the total diagnoses, followed by back pain syndromes (15%). Gout and rheumatoid arthritis were seen in 9.4% and 4.3% respectively. Systemic Lupus Erythematosus made up majority of the connective tissue diseases (2.5%)

Awareness of the geographical distribution of the rheumatic diseases is important for prioritization of directive efforts towards their diagnoses and therapy.

M26 Rheumatic Manifestations of Malignancy: A One-year Survey of Cancer Patients Seen at the UP-PGH Medical Center

Leonila F. Dans, MD and Victoria L. Torralba, MD
Rheumatology Section, Department of Medicine UP-PGH, 1989

Musculoskeletal syndromes have been reported as presenting manifestations of occult neoplastic processes. To describe the different rheumatologic manifestations associated with certain malignancies in the local setting, a survey of diagnosed cancer patients seen at the UP-PGH Medical Center, from July 1988 to July 1989 was done. Of 189 patients (81 children and 108 adults) interviewed, 50 cases (26.5%) were most common underlying malignancies noted were acute leukemia (n=24), lymphoma (n=7), and (n=44) had symptoms prior to oncologic diagnosis, with an average time interval of 3.5 months from the onset of symptoms to final diagnosis. The most common presentation was oligoarthritis, with involvement of the shoulders, knees and ankle joints. Other Rheumatologic symptoms elicited were vasculitis, polymyositis, shoulder-hand syndrome and localized myalgia. Results of this study are compared with reported cases in foreign literature.

M27 Experience Survey on Tenoxicam and Epidemiological Research on Rheumatism Among Five Thousand Filipino Patients.

Penserga E., MD, Ngelangel C., MD
Phil J of Int Med, 32(5), Sept-Oct, 1994

M28 Successful Use of Diltiazem in Calcinosis Caused by Connective Tissue Disease. Case Report

Torralba, Tito P., MD, Li-Yu, Julie;MD, Navarra, Sandra T.G.V. , MD

Journal of Clinical Rheumatology. 5(2):74-78, April 1999

The mechanism for calcinosis in connective tissue disease remains unclear. Various therapies such as warfarin, colchicine, steroids, and bisphosphonates have been tried. However, despite some benefit in early cases, improvement generally is not seen in advanced cases of calcinosis. Several recent studies strongly suggest a favorable response of calcinosis to diltiazem, a calcium channel antagonist. This report concerns two Filipino women, one with dermatomyositis, the other with undifferentiated connective tissue disease, who showed significant reduction of widespread calcinosis after treatment with diltiazem.

M29 Calcium Apatite Crystals in Synovial Fluid Rice Bodies

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Published in Annals of Rheumatic Diseases 2002;61:387-390

BACKGROUND: Rice bodies can occur in the joints in many rheumatic conditions, but they are most common in rheumatoid arthritis. They are generally believed to occur rarely in patients with osteoarthritis, but one study reported rice bodies with apatite crystals.

OBJECTIVE: To report on a series of joint fluids with rice bodies containing apatite clumps and examine their clinical pictures.

METHODS: All synovial fluid analysis reports for 10 years were reviewed for rice bodies and eight patients were reported on. A series of patients with a variety of diseases with synovial fluid rice bodies

found to contain calcific material is described. All were examined by compensated polarised light and alizarin red stain, and four were examined by electron microscopy.

RESULTS: The eight patients all had alizarin red S chunks embedded throughout the rice body. Transmission electron microscopy disclosed the presence of a matrix of collagen, fibrin, and amorphous materials containing typical apatite crystals. Clinical diagnoses, radiographic findings, and leucocyte counts varied, but six of the eight patients had had previous repeated corticosteroid injections into the joints.

CONCLUSION: Aggregates of apatites may be more common than previously recognised in rice bodies as they are not routinely sought. Whether they are a result of joint damage or depot steroid injections and whether that might contribute to further joint injury now needs to be investigated.

Keywords: calcium apatite; synovial fluid; rice bodies

Abbreviations: CPPD, calcium pyrophosphate dihydrate; OA, osteoarthritis; RA, rheumatoid arthritis; SF, synovial fluid

M30 Interstitial Lung Disease among Filipinos with Connective Tissue Disease

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St. Luke's Medical Center and University of Santo Tomas Hospital, Philippines, 2008

AIM: To describe the clinical features, radiographic findings, therapies and clinical course of Interstitial lung disease (ILD) in Filipinos with connective tissue diseases (CTDs).

METHODS: We retrospectively reviewed the records of patients diagnosed with ILD using the 2002 American Thoracic Society (ATS)/ European Respiratory Society (ERS) International multidisciplinary consensus classification of the idiopathic interstitial pneumonias. All patients had underlying CTDs, defined by respective American College of Rheumatology (ACR) criteria, and seen at 2 tertiary referral centers (St. Luke's Medical Center and University of Santo Tomas Hospital) in the Philippines.

Results: Of the 35 patients (32 women), 7 had systemic lupus erythematosus (SLE), 7 with scleroderma, 5 with mixed connective tissue disease (MCTD), 5 with dermatomyositis (DM/PM), 6 with rheumatoid arthritis (RA), 3 with undifferentiated connective tissue disease (UCTD) and 2 with overlap syndrome. The average age at ILD diagnosis was 48 ± 14 years (mean \pm SD), and mean duration of illness from CTD to ILD diagnosis was 26 ± 42 months. Dyspnea was the most common manifestation (29, 83%), 29% (n=10) had concomitant pulmonary hypertension (PAH), 43% (n=15) developed serositis, 23% (n=8) had intermittent cyanosis and 60% (n=21) had chronic cough. Radiographic findings included the following: 17 usual interstitial pneumonia (UIP), 19 nonspecific interstitial pneumonia (NIP), 3 bronchiolitis obliterans organizing pneumonia (BOOP), 5 diffuse alveolar damage (DAD), and 4 lymphocytic interstitial pneumonia (LIP). Five patients developed malignancies: 2 breast, 1 thyroid, 1 liver and 1 non-Hodgkin's lymphoma. Two patients had died at the time of this report, 1 UCTD and 1 DM, both with 1 month duration from ILD diagnosis to mortality. Therapies used in this group of patients for both ILD and CTD included: oral and pulse steroids, hydroxychloroquine, azathioprine, cyclophosphamide, penicillamine, colchicine, methotrexate, sildenafil, iloprost, and leflunomide. Three patients received biologics (2 infliximab and 1 rituximab) and 1 was treated with intravenous immunoglobulin.

CONCLUSIONS: We have described the clinical profile, radiographic findings of ILD in a group of Filipino patients with CTD. Varied forms of pharmacologic therapy were used for these patients. Early recognition and aggressive therapy especially during the "inflammatory" stages of ILD is crucial to more favorable outcomes.

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