

VASCULITIS

V1 Clinical Features of Adults and Children with Henoch Schonlein Purpura

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OBJECTIVES: Henoch-Schönlein purpura (HSP) is an immunoglobulin-mediated vasculitis affecting small sized blood vessels. Although HSP has been extensively studied in children, much less is known about its natural history in adults. The aim of this study is to compare the clinical features of HSP among the adults and children, seen in a tertiary care hospital

METHODS: This is a retrospective study, by reviewing medical records patients with HSP in Rheumatology Clinics of the University of Santo Tomas Hospital, Manila from April 2002 to March 2007. Patients diagnosed as HSP, based on clinical criteria as proposed by Michel et al., including palpable purpura, bowel angina or diffuse abdominal pain, gastrointestinal bleeding, hematuria - without any known drug precipitants and regardless of the criteria for age - were included in this study. Patients were divided into two groups according to their age at HSP diagnosis: childhood onset (≤ 20 years old) and adulthood onset (> 20 years old).

RESULTS: A total of 52 patients with HSP were included in this study, consisting of 40 (77%) children (≤ 20 years), and 12 (23%) adults (> 20 years). The male to female ratio was 1.4:1.0 in children, and 1.0:1.0 among adults. The mean age of disease onset was 9.0 ± 4.6 years (range 2-18) in children and 37.8 ± 12.6 (range 21-54) in adults. All patients had skin manifestations described as purpuric lesions. Abdominal involvement was noted in 34 (85.0%) children and 7 (58.35%) adults, $p = 0.062$, kidney involvement in 20 (50%) children and 6 (50%) adults, $p = 1.0$ and joint involvement among 31 (77%) children compared to 10 (83.3%) adults $p = 0.50$. Most (17, 33 %) patients had onset of disease between January to March of the year.

CONCLUSION: Purpuric lesions were seen in all patients, joint involvement in 77 to 83%, abdominal involvement in 58 to 85%, kidney involvement in 50%. Despite a trend for more frequent abdominal involvement in children, none of the clinical characteristics statistically differed between children and adults with HSP.

V2 Successful Treatment of Mesenteric Vasculitis in an Adult Henoch Schonlein Purpura Patient

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Henoch Schonlein purpura (HSP) is a small vessel vasculitis which is uncommon in adults. The presentations of adult-onset disease are different from those seen in childhood. We report a 44-year-old man with HSP who presented with colicky abdominal pain, hematemesis, hematochezia, arthralgia, and skin rash. Endoscopy was done which showed erythema in the antrum, blood clots in the 2nd part of duodenum and duodenitis. He was started on pulse methylprednisolone therapy at 500mg/IV day for 3 days with subsequent resolution of signs and symptoms.

V3 Successful Treatment of Churg-strauss Syndrome with Rituximab: A Case Report

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1st Place, Poster Presentation. 14th PRA Annual Convention. Cagayan de Oro. Jan 2007

B cell depletion with rituximab, a genetically engineered chimeric anti-CD20 monoclonal antibody has been shown to be effective in certain chronic immunological diseases including rheumatoid arthritis, systemic lupus erythematosus, and antineutrophil cytoplasmic antibody (ANCA) associated vasculitis.

Several published reports have shown its effectiveness in Churg-Strauss Syndrome. We report a case of a 56 year old Filipino male, with a prior history of asthma and sinus abnormalities, who presented with peripheral eosinophilia, leukocytoclastic vasculitis, and severe axonal distal symmetrical sensorimotor polyneuropathy. Sural nerve biopsy revealed vasculitic neuropathy. He had received intravenous immunoglobulin, methylprednisolone pulses and high dose oral steroids but without clinical improvement. He was then given 2 doses of rituximab 500 mg combined with 500 mg cyclophosphamide, administered 2 weeks apart, without adverse events. A month later, his neurologic functions had significantly improved with increasing ambulation, and prednisone was effectively tapered to 10 mg daily.

V4 Acute renal failure secondary to rapidly progressive glomerulonephritis in an adult Filipino Female with Henoch-schonlein Purpura

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SIGNIFICANCE: A disease once thought to be confined to children; Henoch-Schonlein purpura is increasingly being diagnosed in adults. This case underscores the need to consider classic syndromes even in patients who, because of age or other demographic factors, are relatively low risk. In general, HSP is a benign self-limited disorder. Despite the usually benign course some patients develop rapidly progressive glomerulonephritis and die of renal failure. We presented a case of rapidly progressive glomerulonephritis in a patient with Henoch-schonlein purpura who was managed successfully.

THE CASE: An unusual case of a 45 year old Filipina who presented with purpuric rashes over the upper and lower extremities, arthralgia, abdominal pain, oliguria, hematuria, edema and hypertension. She had azotemia, anemia, hematuria, proteinuria and metabolic acidosis. Skin biopsy revealed a leukocytoclastic vasculitis with vascular deposition of IgA by direct immunofluorescence.

High dose steroids at 1mg/kg/day of Prednisone was started. On persistent azotemia Methylprednisolone Pulse Therapy was given. She was also started with Azathioprine 50mg BID and underwent hemodialysis. Improvement of urine output, resolution of purpuric rashes, bipedal edema, hematuria, arthralgia and abdominal pain were noted post therapy. Three months post discharge, patient had no bipedal edema, no purpuric lesions, no hematuria, no oliguria with no recurrence of signs and symptoms mentioned. Serum creatinine was within normal limits, and blood pressure was controlled. There was persistence of microscopic hematuria thus renal biopsy was planned.

V5 Clinical Value of Saddle Nose Deformity in 2 Cases of Wegener's Granulomatosis

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- 1. Oral Presentation. Department of Medicine-UP-PGH Research Forum. Oct 2006**
- 2. Poster Presentation. 14th PRA Annual Convention. Cagayan de Oro. Jan 2007**

SYNOPSIS: We are presenting two female Filipino patients with saddle nose deformities, eye lesions and pulmonary nodules. Biopsy of lesions showed granulomatous inflammation without caseation necrosis. Both patients fulfilled the criteria for Wegener's granulomatosis; the first two cases of documented Wegener's granulomatosis in our center. This widens the differentials for saddle nose deformity to include Wegener's granulomatosis on top of other common causes.

CLINICAL PRESENTATION: Patient A, a 36-year old female, presented with nasal stuffiness, gradual flattening of nasal bridge, painful red eyes, blurred vision, and purulent non-bloody eye discharge for 2 months. Patient B, a 53 year old female, presented with 10-year history of cough, colds and occasional purulent post-nasal drip. She had 6 months duration of nasal stuffiness, weight loss, and undocumented fever followed by collapse of the nasal bridge, dyspnea and hemoptysis.

PHYSICAL FINDINGS: Both patients had saddle nose deformity. Patient A had red palpebral conjunctivae, red papule on bulbar conjunctiva, and purulent discharge at the right eye. Patient B had a right corneal ulcer, nasal septum erosion, leg ulcer near the lateral malleolus, and a gluteal ulcer with good granulation.

LABORATORY WORK-UP: For patient A, imaging studies showed a pulmonary nodule and nasal septal erosions. Sputum and tissue (eye and nose) AFB smears were negative. On biopsies, there was granulation tissue on the conjunctivae and acute inflammation of the nasal mucosa.. Cytoplasmic ANCA was positive. For patient B, imaging studies showed a solitary pulmonary nodule and polysinusitis with secondary obstruction of the left osteomeatal complex. Punch biopsy of the leg ulcer showed granulomatous inflammation with necrosis. Skin biopsy showed fibrinoid necrosis. She had hematuria, pyuria, and granular casts on urinalysis. Sputum AFB smear was negative.

DIAGNOSIS: Both fulfilled at least 2 of the 4 classification criteria of the American College of Rheumatology for Wegener's granulomatosis. Patient A had a pulmonary nodule, nasal inflammation with cartilage collapse, and a positive c-ANCA. Patient B had a pulmonary nodule, nasal inflammation, granulomatous inflammation, and urinary sediments.

TREATMENT: Both patients were treated with combined immunosuppressives (high dose steroids with oral methotrexate for A and high dose steroids and intravenous cyclophosphamide for B)..

OUTCOME: Both patients had improvement in nasal stuffiness after two months. In addition, for Patient B, there was healing of the leg ulcers and decrease in urinary sediments.

SIGNIFICANCE: Saddle nose deformity in the Philippines is usually attributed to trauma, leprosy, or tuberculosis. The two cases presented widen the differential diagnosis for saddle nose deformity to include a rarer cause: Wegener's granulomatosis. This is suspected in the presence of nasal erosions rather than perforations (seen in trauma) with other findings reflecting glomerulonephritis and pulmonary involvement. Treatment of this condition differs significantly from treatment of other more common causes of saddle nose deformity

RECOMMENDATIONS:The finding of saddle nose deformity requires assiduous investigation to establish its cause from a wide range of diseases. Wegener's granulomatosis is rare in the Philippines but it exists. In the presence of nasal deformity, pulmonary and renal manifestations, this condition must be thought of.

V6 Clinical Profile of Henoch Schonlein Purpura Among Adult Filipinos Seen at the Philippine General Hospital

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

OBJECTIVES: This paper aims to present the clinical profile of Henoch-schonlein Purpura (HSP) in Filipino patients.

METHODS: This is a retrospective review of adult patients aged ≥ 16 years admitted for HSP at the Philippine General Hospital from January 2003 to December 2006. The patients were diagnosed to have HSP if they satisfied the criteria set by the American College of Rheumatology (ACR) which includes patient age younger than 20 years, palpable purpura, abdominal pain or gastrointestinal bleeding, extravascular or perivascular granulocytes on biopsy.

RESULTS:There were 6 females and 5 males who met with the ACR criteria for HSP. Ages ranged from 16-56 years with a mean age of 30.9 years.

The most common presenting symptom was rash in 6/11(54%) of patients and abdominal pain in 3/11(27%). All patients subsequently developed rashes and abdominal pain. Rashes were mostly purpuric and non-pruritic. There was musculoskeletal or renal involvement in 7/11 patients (64%). Nausea, vomiting, fever, diarrhea, hematochezia and melena were also noted.

There was leukocytosis with predominance of segmenters in 8/11(73%), thrombocytosis in 4(36%), hematuria in 7/11(64%), proteinuria in 6/11(54%), positive fecal occult blood in 3/9(33%) and elevated acute phase reactants in 3/4(75%). Two had skin biopsy with findings of leukocytoclastic vasculitis with IgA immunofluorescence.

Treatment involved steroids in 10/11(91%) patients; initial dose ranged from 0.75mg/kg/day to 1mg/kg/day. Azathioprine was added to patients with nephritis (3/11, 27%). Abdominal pain and rashes started to resolve within 5 to 10 days of treatment.

CONCLUSION:Our cases of adult Filipinos with HSP had similar clinical manifestations with those reported in the medical literature. Purpuric rash was the most common presenting manifestation. One patient had acute renal failure requiring hemodialysis.

V7 A Filipino Male with Hepatitis B associated Polyarteritis Arteritis Nodosa Successfully Treated with a Combination of Steroids,Lamivudine and Plasma Exchange: A Case Report

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A 38-year-old Filipino male was diagnosed with hepatitis B associated polyarteritis nodosa presenting as progressive asymmetric sensorimotor polyneuropathy of the lower extremities. He was initially given pulse methylprednisolone 1 gram daily for 3 days, followed by oral steroid and lamivudine 100 mg per day. Because of progression of the neuropathy, plasma exchange was also started. He steadily improved on this regimen, with return of his motor strength by the fourth week of therapy. Oral steroids and serial plasma exchange were subsequently tapered with continued improvement, and decrease in viral load. This case illustrates that a combination of steroids, lamivudine and plasma exchange is effective in the treatment of hepatitis B associated polyarteritis nodosa.

V8 Prevention of Relapse of Cutaneous Vasculitis with Empiric Anti-Microbial: A Report of Three Cases

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1. **Poster Presentation. 12th APLAR Congress. Kuala Lumpur, Malaysia, August 2006**
2. **APLAR Journal of Rheumatology 2006; 9(suppl.1) A277**

BACKGROUND AND OBJECTIVE: Cutaneous vasculitis is mostly idiopathic, requiring prolonged use of corticosteroids and/or immunosuppressives. We describe 3 cases of recurrent cutaneous vasculitis given high dose corticosteroids and empiric antimicrobial agent.

Case 1. An 8 year old female, presented with progressive painful gangrene of fingertips with leukocytosis ($17.5 \times 10^9/L$), elevated anti-streptolysin - O (ASO) 400 IU/ml and erythrocyte sedimentation rate (ESR) 32 mm/hr; C-reactive protein (CRP), antinuclear antibody (ANA), rheumatoid factor were negative and complement (C3) was normal. Biopsy showed leucocytoclastic vasculitis. Improvement with pulse methylprednisolone, dapsone and high dose prednisone was noted upon tapering steroids. Vasculitic lesions, fever, cyanosis with impending gangrene of the fingers and toes were noted. Blood cultures grew *Staphylococcus Aureus*. Intravenous cloxacillin, gentamycin, steroids and heparin were given with clinical improvement. Monthly prophylactic amoxicillin for 10 days enabled tapering and discontinuation of steroids. No recurrence of lesions noted for 32 months.

Case 2. A 19 year old female came with an 8 year history of recurrent painful, erythematous nodules and livedo reticularis on lower extremities. Histopathology showed nodular vasculitis. Recurrence noted on steroid tapering. Oral cyclophosphamide, dapsone, and empiric antibiotic were given separately with transient improvement. Laboratory tests disclosed hemoglobin 11.1 mg/dl, increased platelets, ASO titer 1:400 and ESR 55 mm/hr. Chest x-ray, urinalysis and C3 were normal. Methylprednisolone pulse therapy was given and she was maintained on high dose prednisone. Cutaneous exacerbations were noted on tapering of prednisone. Prophylactic amoxicillin for 7 days monthly led to improvement with no recurrence for 34 months.

Case 3. A 7-year old female presented with pain, swelling and development of multiple vasculitic lesions on lower extremities. There were leukocytosis (WBC $24.100 \times 10^9/L$), anemia (hemoglobin 92 mg/L), elevated ESR (142 mm/hr) and CRP levels. Initially, she was responsive to high dose steroids but recurred upon tapering. Improvement with methylprednisolone pulse therapy plus 10 day course of broad spectrum

antibiotics was noted. Monthly benzathine penicillin, prednisone and methotrexate were given. A year later, fever with erythematous lesions on all extremities was noted. Histopathology disclosed leucocytoclastic vasculitis. Methylprednisolone pulse therapy with gradual tapering of prednisone and monthly prophylaxis of amoxicillin for 10 days resulted in improvement over 34 months.

SUMMARY: The extended use of antimicrobial agent on a monthly prophylaxis regimen led to tapering and discontinuation of steroids. This strongly suggests the potential use of antibiotic prophylaxis for such individuals.

V9 Correlation of Positron Emission Tomography with Disease Activity among Patients with Takayasu's Arteritis"

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1. **Poster Presentation. European League Against Rheumatism. Vienna, Austria. 2005**
2. **Oral Presentation. 12th PRA Annual Convention. Cebu. Jan 2005**

OBJECTIVES: Takayasu's arteritis is a rare disease predominantly affecting the large arteries particularly the aorta and its main branches. The gold standard for investigation is angiography. Because of the rarity in presentation, data for treatment were culminated from case reports and case series of other studies. What is more difficult is the question of among the patients with the disease, when do you initiate or stop treatment as far as aggressive immunosuppressants in controlling disease activity? There is no current standard parameters to monitor disease activity, thus it is the aim of this paper to describe the patients with Takayasu's arteritis who underwent Positron Emission Tomography (PET). It is the main objective of this paper to describe the patient's clinical course and its PET findings.

DESIGN: Review and describe the patients who presented with Takayasu's arteritis who underwent PET scan at St Luke's Medical Center.

SETTING: the study setting is in a tertiary hospital where the level of clinical care is from a private practice.

PARTICIPANTS: Patients were included in the study if they fulfilled the American College of Rheumatology classification criteria for Takayasu's arteritis. Those patients with no PET scan were excluded from the study.

RESULTS: The PET findings provided information in terms of describing disease activity when correlated with its clinical course.

CONCLUSION: There seems to be a correlation between disease activity and tracer uptake among patients with active disease. PET scan can thus be used for establishing disease activity, which can greatly help in deciding the need for immunosuppressants.

V10 Asymptomatic Cardiac Involvement in Churg Strauss Syndrome

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

Cardiac involvement is the leading cause of mortality in 48% of patients with Churg-Strauss Syndrome (CSS). Congestive heart failure (CHF) is the usual manifestation. We present two cases of patients with CSS with asymptomatic cardiac involvement.

Case 1. A 51-year-old male, diagnosed with asthma two years prior, had worsening asthma and rashes. He had diffuse lung wheezes, purpura and hemorrhagic bullae, digital gangrene and polyneuropathy. Laboratory tests showed WBC $38.2 \times 10^9/l$ with 62% eosinophilis, ESR 90mm/h, positive pANCA. Chest x-ray showed bibasal infiltrates. Skin biopsy showed leucocytoclastic vasculitis with extravascular eosinophils. Patient denied chest pain. However, electrocardiogram (ECG) showed ST elevation in leads

V1-V4 and complete right bundle branch block. Troponin I was 11.8 ng/ml. 2-D Echocardiogram revealed hypokinesia of the interventricular septum, anterior and lateral walls of the left ventricle, and ejection fraction of 32%. Intravenous corticosteroids and cyclophosphamide resulted in overall improvement. Patient refused coronary angiography and catheterization.

Case 2. A 51-year-old female, diagnosed with CSS 4 years prior, developed hemorrhagic blisters, sinusitis, and left foot drop. Complete count showed WBC $7.5 \times 10^9/l$ with 16% eosinophilia. ECG showed T wave inversions in leads V2-V6. 2D-Echocardiogram showed left ventricular hypertrophy with impaired relaxation. Prednisone 50mg daily improved her symptoms and ECG findings.

Both patients fulfilled American College of Rheumatology criteria for the diagnosis of CSS with features of asthma, eosinophilia, neuropathy, sinusitis, extravascular eosinophils and lung infiltrates. Myocarditis with ventricular dysfunction and pericarditis are the most common cardiac complications. Up to 50% of patients have ECG abnormalities. Both patients described had no symptoms referable to their cardiac disease yet both had abnormal ECGs and 2D-Echocardiograms. We conclude that cardiac involvement in CSS may be asymptomatic and at least a baseline ECG is warranted in these patients.

V11 Large Vessel Vasculitis and Leprosy: a Case Report

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1. **Future in Rheumatology: from Bench to Bedside. (Abstract Book P322). APLAR 2004. p.168**
2. **Poster Presentation. 11th APLAR Congress. Korea, Sept 2004**
3. **Oral Presentataion. Department of Medicine-UP-PGH Reseach Forum. Oct 2005**

A 22-year old female nonsmoker presented with paresthesia, claudication, skin lesions, absent pulses on her lower extremities, and toe gangrene. Skin biopsy revealed leprosy and duplex scan documented severe stenosis of the femoral arteries. This is the first reported case of vasculitis involving large blood vessels in the setting of leprosy. The possible association of mycobacterial disease and primary systemic vasculitis is discussed.

V12 MCTD with Severe Raynaud's Phenomenon, Gangrene and Cutaneous Leucocytoclastic Vasculitis: A Case Report

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1. **Future in Rheumatology: From Bench to Bedside. Abstract Book (P379). APLAR 2004. p. 190-191**
2. **Poster Presentation. 11th APLAR Congress. Korea, Sept 2004**
3. **Free Poster Presentation. 35th PCP Annual Convention. Shangrila Hotel, Manila. May 2005**
4. **Phil J of Internal Medicine. 43(3): 137-140. May-June 2005**

PURPOSE: To describe a unique case of leucocytoclastic vasculitis in patient diagnosed with Mixed Connective Tissue Disease

METHOD: Case Report

RESULTS: We describe a case of a 28 year-old male with history of stroke and newly diagnosed to have Mixed Connective Tissue Disease, who developed rapidly evolving ulcers and Raynaud's phenomenon in all distal extremities. The patient presented clinically with pain, paresthesia and numbness of distal extremities, swollen hands, digital cyanosis and ulcers which eventually progressed to gangrene. He also had coexistent proximal muscle weakness involving the shoulder musculature. Serological work-up revealed a +4 ANA-speckled pattern and anti U1-RNP, anemia, thrombocytosis and negative antiphospholipid antibodies. Hepatitis profile was negative. pANCA and cANCA were likewise negative. EMG-NCV showed axonal sensory neuropathy which may be due to vasculitis. Muscle and sural nerve biopsies were unremarkable, however, biopsy of the skin lesions showed leucocytoclastic vasculitis.

The patient was initially treated with nifedipine, ASA and intravenous prostacyclin for 3 days. Digital gangrene was progressive, thus methylprednisolone pulse therapy was administered. Symptoms improved and progression of digital gangrene stopped. He was then maintained on Prednisone initially at 1 mg/kg then tapering dose, Nifedipine, ASA and Clopidogrel and was discharged improved.

MCTD can present in a variety of cutaneous manifestation. One case report featured skin biopsy that showed pustular leucocytoclastic vasculitis. The coexistence of vasculitic ulcers and MCTD has been postulated to be a result of localization of immunoglobulin and complement components in diseased vessel walls, ANCA, as well as other inflammatory mediators have been implicated. Exact mechanism is still ill-defined. The presence of vasculitic lesions is a poor prognostic factor in MCTD.

CONCLUSION: Cutaneous leucocytoclastic vasculitis in the setting of diagnosed MCTD is rarely described in medical literature. Awareness of such an association is important in early diagnosis and management.

V13 Churg-Strauss Vasculitis Associated with Antiphospholipid Antibodies

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Poster Presentation. 9th PRA Convention. Edsa Shangri la Hotel, Manila. Jan 2002

SYNOPSIS: We have presented the first case of Churg-Strauss Vasculitis associated with antiphospholipid antibodies

CLINICAL PRESENTATION: This is a 32-year old male, admitted for severe pain of both lower extremities and abdominal pain. Patient has bronchial asthma and allergic rhinitis

PHYSICAL FINDINGS: Patients was cachectic with severe atrophy of the muscles of the extremities. There was a decrease in motor strength of 3/5, sensory loss and severe tenderness of the abdomen and gastrocnemius muscles

LABORATORY WORK UP: Pertinent laboratory results are: eosinophilia (0.162), ESR (48mm/hr), DRVVT (Control=38 seconds Test=80 seconds), ACA IgM (110 MPL units/ml, up to 12.5 MPL units/ml). Eosinophilia is consistent with CSV and abnormal DRVVT and IgM to a possible antiphospholipid antibody syndrome (APAS).

DIAGNOSIS: Patient satisfied 4 of the 6 criteria for the classification of CSV> Biopsy of the removed jejunum is also consistent with CSV

TREATMENT: Exploratory laparotomy revealed a gangrenous jejunum. He was given high dose corticosteroids for CSV and anticoagulation for APAS. He was also given tramadol, carbamazepine, gabapentin and mecobalamin for pain and neuropathy.

OUTCOME: The patient's leg pains did not improve despite aggressive treatment. He succumbed due to intracerebral hemorrhage

SIGNIFICANCE: The prognosis of CSV is good. Whether the presence of antiphospholipid antibody in CSV affects the outcome is hard to determine

RECOMMENDATIONS: It is worthwhile to look into the association of CSV and the presence of antiphospholipid antibodies.

V14 Chapter on Takayasu Arteritis

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Published in NORD Guide to Rare Diseases.

Tan-Ong, M, Hoffman, G. Takayasu Arteritis. Edited by Gruson, E. In-depth Guide to Rare Diseases. The National Organization for Rare Diseases (NORD) Guide to Rare Disorders. Philadelphia: Lippincott Williams & Wilkins, 2003, p. 35.

DEFINITION: Takayasu arteritis is an idiopathic systemic inflammatory disease that may lead to gradual segmental stenosis, occlusion, dilatation, and/or aneurysm formation of the aorta and/or the coronary or pulmonary arteries. **SYNONYMS:** Pulseless disease; Young female arteritis; Middle aortic syndrome; Martorell syndrome; Occlusive thrombo-aortopathy; Nonspecific aorto-arteritis.

DIFFERENTIAL DIAGNOSES: Vascular infections; Spondyloarthropathies with aortitis; Buerger disease; Behçet syndrome; Cogan syndrome; Kawasaki disease; Sarcoidosis; Giant cell arteritis; Ehlers-Danlos syndrome; Marfan syndrome; Fibromuscular dysplasia; Neurofibromatosis; Ergotism; Radiation fibrosis.

SYMPTOMS AND SIGNS: The disease ranges from being entirely asymptomatic to having a morbid, disabling, or fatal outcome. Vascular ischemic symptoms are the hallmark. Disease manifestations include bruits, claudication, diminished or absent pulses, asymmetric blood pressure measurements, hypertension, carotodynia, pain over inflamed arteries, Raynaud phenomenon, angina, myocardial infarction, aortic regurgitation, congestive heart failure, pericarditis, palpitations, dizziness or lightheadedness, syncope, transient ischemic attacks, cerebrovascular accidents, seizures, amaurosis fugax, diplopia, blurred vision, iritis, episcleritis, hemoptysis, dyspnea, pleural effusions, arthralgias (rarely synovitis), abdominal pain, nausea, vomiting, erythema nodosum, and rarely, pyoderma gangrenosum and panniculitis. One or more nonspecific systemic symptoms such as fever, night sweats, malaise, fatigue, weight loss, myalgias, arthralgias, neck pain, or cervical lymphadenopathy occur in approximately half of patients at the onset of disease.

ETIOLOGY/EPIDEMIOLOGY: Although the exact etiopathogenesis of Takayasu arteritis is unknown, it is clear that vessel injury is caused by lymphocytes and macrophages. The disorder occurs most often in Asia, although cases have been recognized worldwide. Approximately 90% of patients are females of child-bearing age, but males may also be affected.

DIAGNOSIS: Diagnosis rests on a high index of suspicion, especially in the presence of ischemic manifestations in young individuals. Angiography is the procedure of choice.

TREATMENT: Standard Therapies: For control of immunoinflammatory features, glucocorticoids are given for approximately 1 month and tapered to the lowest dose required to control disease. Whenever chronic glucocorticoid therapy is employed, bone-preserving therapy should be added. If glucocorticoid therapy cannot be tapered without disease exacerbation, cytotoxic agents such as oral cyclophosphamide daily, azathioprine daily, or methotrexate weekly are added. If disease is well controlled after the addition of cytotoxic agents, corticosteroids are gradually tapered to discontinuation. Prophylactic agents for *Pneumocystis carinii* should accompany combination corticosteroid and cytotoxic therapy. Angioplasty or revascularization may be indicated in some patients. Elective surgery is generally recommended during periods of disease quiescence.

Investigational Therapies: Tacrolimus, mycophenolate mofetil, cyclosporin, and anti-TNF therapy (etanercept and infliximab) are being investigated.

V15 Takayasu's arteritis: vascular interventions and outcomes.

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1. *Journal of Rheumatology* 2004 Jan; 31(1):102-6.
2. Poster Presentation. 10th International Vasculitis and ANCA Workshop, Cleveland, Ohio, U.S.A. on April 27, 2002
3. Abstract published in the *Cleveland Clinic Journal of Medicine* Vol. 69 Supplement II –188;
4. Poster Presentation. 2003 Annual Scientific Meeting of the American College of Rheumatology, Orlando, Florida, U.S.A.
5. *Arthritis and Rheumatism* 2003; 48 (9): s202.

OBJECTIVE: To provide an analysis of outcomes of vascular interventions in 20 patients with Takayasu's arteritis (TA) who received care at the Cleveland Clinic Foundation between 1979 and 2001.

METHODS: We performed a retrospective chart review. The primary outcome measure of our review was patency of vessels as assessed by repeat invasive angiography or magnetic resonance angiography. The secondary outcome measures included periprocedural complications, morbidity, and mortality.

Interventions included bypass grafts, patch angioplasty, endarterectomy, percutaneous transluminal angioplasty (PTA), or stent placement.

RESULTS: Sixty-two revascularization procedures were performed in 20 patients. Followup evaluations were available for 52 procedures. Eleven of 31 bypass grafts restenosed or occluded between one day to 168 months after surgery. Three of 7 PTA and 5 of 7 stents restenosed or became occluded after 1-72 months and 2-45 months of followup, respectively. There were no deaths associated with revascularization procedures.

CONCLUSION: Despite providing short term benefit, endovascular revascularization procedures are associated with a high failure rate in patients with TA.

V16. Anti-Tumor Necrosis Factor Therapy in Takayasu Arteritis Resistant to Conventional Therapy.

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1. Oral Presentation. 10th International Vasculitis and ANCA Workshop, Cleveland, Ohio, U.S.A. on April 26, 2002

2. Abstract published in the Cleveland Clinic Journal of Medicine Vol. 69 Supplement II – 182-183.

BACKGROUND: Takayasu arteritis is an idiopathic systemic inflammatory disease that may large and medium-sized artery stenosis, occlusion, dilatation and/or aneurysm formation. A significant number of patients fail to achieve and sustain remission despite prolonged treatment with glucocorticoid and cytotoxic agents.

OBJECTIVE: To assess the utility of anti-tumor necrosis factor (anti-TNF) therapy in patients with Takayasu's arteritis who have been unable to maintain remission following conventional therapy.

PATIENTS AND METHODS: Open label pilot study of etanercept or infliximab in 5 patients with relapsing Takayasu arteritis. Outcome measures included clinical characteristics, laboratory abnormalities, vascular anatomy changes and ability to withdraw or reduce glucocorticoid and cytotoxic therapies in the absence of relapse.

SETTING: Tertiary care referral center

INTERVENTION: Etanercept doses varied from 25 mg twice a week to 50 mg twice a week. Infliximab was provided in one patient in doses up to 7 mg/kg every 8 weeks.

RESULTS: Patients included 1 male and 4 females, four Caucasians and 1 Far East Indian. The mean age was 28.2 years. Mean duration of disease prior to anti-TNF therapy was 5.2 years. Patients had previously experienced a mean of 5.5 relapses. Prior to trials of anti-TNF therapy, relapses had occurred when the mean daily prednisone dose was less than 21 mg. Patients had previously also failed to maintain remission on concurrent therapy with methotrexate, azathioprine, cyclosporin, mycophenolate mofetil and/or tacrolimus. Relapses lead to starting anti-TNF therapy. Time to achieving a degree of unprecedented improvement, that allowed for successful glucocorticoid reduction, ranged from 2 weeks to 2 months. Duration of follow up on anti-TNF therapy has been 29.6 (mean) months. One patient refused re-treatment with prednisone and achieved remission with only anti-TNF therapy. Three of the other 4 patients were able to discontinue prednisone after 4, 11 and 26 months following the start of anti-TNF therapy. The fourth patient continues to be in remission while taking 10mg of prednisone every other day. The mean period of continued disease remission, while receiving anti-TNF therapy, has been 19.4 months. Mean period of remission, without glucocorticoids, on anti-TNF therapy alone in 4 patients has been 12.8 months.

CONCLUSION: Anti-TNF therapy appears to be a useful adjunct to glucocorticoids in the treatment of Takayasu arteritis.

V17 Gastrointestinal Bleeding with Endoscopic Features of Mesenteric Vasculitis in Adult Henoch Schonlein Purpura

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OBJECTIVE: To report two cases of Adult Henoch Schonlein Purpura (HSP) presenting as gastrointestinal bleeding secondary to mesenteric vasculitis

CASE ABSTRACT: HSP is a small vessel vasculitis which is uncommon in adults. The presentations of adult-onset disease are different from those seen in childhood. We report two middle aged males with HSP who both presented with colicky abdominal pain, hematemesis, hematochezia, arthralgia, and skin rash. Endoscopies were showed erythema in the antrum, blood clots in the 2nd part of duodenum and duodenitis in the first case and colonic vasculitis in the second. Both received intravenous steroids either as pulse or as bolus with subsequent resolution of signs and symptoms.

CONCLUSION: Gastrointestinal bleeding as a complication of mesenteric vasculitis among HSP patients was seen in these two patients. Early recognition via prompt endoscopic procedure provided immediate therapeutic intervention with steroids thus preventing a potentially life threatening clinical sequelae.

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