Vasculitis - Medium & Variable Vessel Sizes

Vasculitides are characterized by blood vessel inflammation with possible necrosis, ischemia, and organ damage. The vessels and organs affected vary by the specific vasculitic disorder. **General symptoms and signs** are due to systemic inflammation, which can cause fever, arthritis, arthralgia, fatigue, and weight loss. Skin lesions are associated with small and medium vessel vasculitides. Treatments include corticosteroids and immunosuppressants.

VASCULITIS - MEDIUM

POLYARTERITIS NODOSA
Polyarteritis nodosa is characterized by necrotizing inflammation that most commonly involves the medium muscular arteries, especially at their branch points. The resulting ischemia can affect multiple organ systems: – **Nervous system** involvement often produces mononeuritis multiplex or asymmetric polyneuropathy; sensory and motor deficits of the median, ulnar, and fibular nerves are common. – **Renal** involvement can lead to hypertension, oliguria, and even renal failure. – **Gastrointestinal** involvement can produce pain and malabsorption; when larger vessels, such as the celiac trunk or its branches are affected, fatal aneurysms can develop. – **Skin manifestations** can take various forms, including livedo reticularis (which presents as a purplish lace-like pattern), ulcers, subcutaneous nodules, or even gangrene. – **Heart failure** can occur due to coronary artery obstruction. Polyarteritis nodosa can be systemic or cutaneous. Some secondary forms are related to hepatitis B and C infections, or to rheumatoid arthritis. Most commonly affects men over 50 years of age.
KAWASAKI DISEASE
Kawasaki disease (aka, mucocutaneous lymph node syndrome) is characterized by **proliferative inflammation**, and tends to involve the medium muscular arteries. It has a predilection for the coronary arteries, which are affected in approximately 20% of cases Key cause of acquired heart disease in children. Large coronary aneurysms can be fatal, as they can cause myocarditis, cardiac tamponade, or, as we've illustrated here, coronary artery thrombosis. Typically presents as a self-limiting, acute febrile illness in children: – Fever – Conjunctivitis – Erythematous macular rash – Edema and desquamation, particularly of the limbs – Cervical lymphadenopathy – A red, inflamed throat – Strawberry tongue with dry, fissured lips.

VASCULITIS - VARIABLE VESSEL SIZE

BEHÇET DISEASE
Behçet disease is a chronic, relapsing vasculitis that affects vessels of all sizes. Mucosal inflammation is a common manifestation of the disorder, and often causes oral and genital lesions; ocular inflammation can produce uveitis or hypopyon. Cutaneous manifestations vary, and include pus-filled bumps, palpable purpura, erythema nodosum, and superficial thrombophlebitis. Typical age of onset is around 20 years old. Behçet disease is equally common in men and women, though it's often worse in men.
BUERGER DISEASE

Don't confuse Buerger disease with Berger's disease, a nephropathy caused by IgA accumulation.

Symptoms/Signs
Characterized by **inflammation and occlusive thrombosis** of the medium and small arteries and veins. Most commonly affects the extremities; signs and symptoms typically begin distally and move proximally. Ischemia from vessel occlusion often produces numbness, coldness, or tingling in the extremities, with claudication, then, pain at rest. Affected limbs are often cold, sweaty, and cyanotic, and ulcers form that progress to gangrenous.

Smoking/Management
Smoking is a major cause of Buerger disease, and smoking cessation is key to remission.
Histopathology
Histopathology reveals neutrophil infiltration and granulomatous formation resulting in vessel occlusion with relative sparing of the vessel wall. Arteriography may show a classic "corkscrew" appearance. See: Corkscrew Collaterals in Thromboangitis Obliterans (Buerger's Disease).

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CLINICAL CASES

CASE 1: VASCULITIS EVALUATION
A 3-year-old male was admitted last night through the emergency department following a 2-week history of fevers and a rash. No source of the infection was identified by the admitting physician. You are called to assist in determining the diagnosis. Upon review of the patient's chart, it is noted that his immunizations are up to date, there is no history of similar symptoms, recent travel, or known sick contacts. Blood and urine cultures were obtained in the emergency department. A significant amount of white blood cells were seen in his urine, though no bacteria were noted. On physical examination, his temperature is 38.6 degrees Celsius (101.5 degrees Fahrenheit), blood pressure 95/50 mm Hg, heart rate 90/min, respiratory rate 22/min, and oxygen saturation is 99 percent on room air. The patient's pupils are equal, round, and reactive to light and accommodation. His conjunctiva are injected bilaterally with notable perilimbal sparing, though without detectable matting, discharge, or exudate. The oropharynx is erythematous without tonsillar hypertrophy. His tongue and lips are bright red and swollen. The tympanic membranes are injected, but clear bilaterally. The patient's lungs are also clear, and his cardiac examination is unremarkable with no murmurs, rubs, or gallops detected. An erythematous polymorphous rash is noted on his trunk and extremities. Perianal erythematous desquamation is also noted without induration. The dorsums of the patient's hands and feet are noted to be edematous bilaterally. Further inspection demonstrates marked erythema of both the palms and soles of his feet. **Laboratory Results** Complete Blood Count Leukocyte Count 13,000 mm3 Hemoglobin 9.2 g/dL Hematocrit 35 percent Platelets 690,000 mm3 Liver Function Tests Alanine aminotransferase 71 U/L Aspartate aminotransferase 80 U/L Total Proteins 10 g/dL Total Bilirubin 0.9 mg/dL Albumin 2.2 g/dL Hematologic Studies C-Reactive Protein 30 mg/L Erythrocyte Sedimentation Rate 40 mm/h Based upon the history, physical examination, and laboratory results, what is the most likely diagnosis?
ANSWER

• Kawasaki disease (mucocutaneous lymph node syndrome)

EXPLANATION
This patient has Kawasaki disease (mucocutaneous lymph node syndrome), which is an acute, self-limited condition that causes inflammation of blood vessels throughout the body (medium vessel vasculitis). Coronary artery aneurysm and ventricular dysfunction are the most concerning potential sequelae of the disease. A number of criteria exist (many of which are seen in this clinical scenario) to assist with the diagnosis of Kawasaki disease. A fever of unknown origin must be present for at least 5 days, and 4 of the following must also be present: bilateral, nonexudative conjunctival injection with perilimbal sparing, oral mucousal membrane changes (including injected or fissured lips, injected pharynx, or strawberry tongue), peripheral extremity changes (including erythema of palms or soles, edema of hands or feet, and/or periungual desquamation), polymorphous rash, and cervical lymphadenopathy (at least 1 lymph node greater than 1.5 cm (0.6 inches) in diameter). The patient in this clinical scenario has all of these findings with the exception of cervical lymphadenopathy. There are also laboratory changes (all seen in this patient) expected with Kawasaki disease, which include white blood cells with no bacteria in the urine (sterile pyuria), elevated white blood cell count (leukocytosis), low hemoglobin (anemia), elevated platelets (thrombocytosis), elevated liver function tests (ALT and AST), low albumin (hypoalbuminemia), and increased inflammatory markers (C-reactive protein and erythrocyte sedimentation rate). A high white blood cell count and the presence of anemia and inflammation (signs of systemic inflammation) are cardinal signs of Kawasaki disease. Nail changes (Beau's lines) are commonly seen in the convalescent stage of the disease, and include a white-color to the nail plate (leukonychia), and proximal separation of the nail plate from the nail matrix due to a temporary cessation of growth (onychomadesis). Treatment is intended to decrease inflammation (specifically in the coronary arteries) to prevent the development of an aortic aneurysm. This patient should be given high dose intravenous immunoglobulin (IVIG) at 2 g/kg over 10 to 12 hours, as well as high dose aspirin (ASA) 80 mg/kg/day divided every six hours until he is afebrile for over 48 hours.

BOARD REVIEW

MEDIUM & VARIABLE VESSEL VASCULITIS
Getting ready for boards? Review these concise, bulleted high yield reviews for your exam.
USMLE & COMLEX-USA
USMLE Step 1 / COMLEX-USA Level 1 USMLE Step 2 / COMLEX-USA Level 2 USMLE Step 3 / COMLEX-USA Level 3

NURSE PRACTITIONER (NP)
Nurse Practitioner Licensing Exam
PHYSICIAN ASSISTANT (PA)
Physician Assistant Licensing Exam
INTERNAL MEDICINE (ABIM)
American Board of Internal Medicine (ABIM) Exam

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