Polyarteritis Nodosa

Polyarteritis nodosa (PAN) is an immune complex-mediated systemic vasculitis of medium and small arteries. This disease does not typically involve arterioles, capillaries or venules. It affects various organs in the body including the heart, liver, GI tract and kidneys, but spares the lungs. Unlike other types of vasculitis such as Churg Strauss syndrome or Wegener’s granulomatosis, PAN is not associated with antineutrophil cytoplasmic antibodies (ANCAs). However, PAN is associated with hepatitis B infections. In fact, 30% of patients with PAN have chronic hepatitis B infections, and antibody complexes to hepatitis B surface antigens are often found within inflamed vessels. PAN vessel changes are defined by what’s called segmental transmural necrotizing inflammation. While the disease process may be chronic, during the acute phase, the inflammation is characterized by infiltration of neutrophils, mononuclear cells and eosinophils, resulting in what’s called fibrinoid necrosis. Another differentiating characteristic that makes PAN unique compared to the other vasculitis is that different stages of inflammation can occur simultaneously due to the recurrent nature of this process. PAN is characterized by inflammation of the vessels which can lead to infarction, ulceration, or aneurysm formation anywhere the disease process is taking place. This disease can affect patients of all ages and can be intermittent, chronic or acute. Common manifestations include fever, weight loss, hypertension, melena, myalgia, malaise and peripheral neuritis. The disease process can be controlled by corticosteroids and cyclophosphamide.



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Pathophysiology

Immune Complex-Mediated Transmural Vasculitis

Immune Complexes on Vessels-on-fire

Polyarteritis nodosa is a vasculitis of the small and medium arteries, due to immune complex-mediated inflammation across all layers of the vessel wall.

Hepatitis B Association

Happy-tie-liver Bee

Around 30% of patients with polyarteritis nodosa have chronic hepatitis B, and the immune complexes found in vessels are often antibodies to hepatitis B surface antigen. This is a commonly tested association clinically and on exams.

Fibrinoid Necrosis

Fabric Necrosis-crow

This is a form of necrosis characterized by accumulation of protein-rich material in tissue, appearing pink and fibrin-like on histology. This type of necrosis is commonly associated with immune complex-mediated diseases.

Signs and Symptoms

Black Stool (Melena)

Black Intestine-stool

Also called melena, this is a form of bloody stool that results from bleeding in the gastrointestinal tract. The black color is derived from the oxidation of the iron in hemoglobin, when the blood is passing through the lower part of the intestinal tract. In polyarteritis nodosa, inflammation and vascular lesions in the upper intestinal tract cause this finding. This is in contrast to bleeding in the lower intestines, which may result in bright red blood or hematochezia.

Neurologic Dysfunction

Neuron Dysfunctioning

Polyarteritis nodosa also involves the nerves both peripherally and centrally. Peripheral nerve involvement results in pain, numbness and weakness. Central nervous system involvement can result in seizures.

Myalgia

Mayo-algae

Myalgia means muscle pain, and it can have various traumatic, metabolic, infectious and autoimmune etiologies. This is a common finding in polyarteritis nodosa because of the systemic inflammation going on in small and medium vessels.

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Fever

Fever-beaver

One of the most common clinical manifestations of polyarteritis nodosa, this finding is a result of the systemic inflammatory process occurring in small and medium arteries throughout the body.

Hypertension

Hiker-BP

High blood pressure (systolic above 140mm or diastolic above 90mm in adults) can occur due to various primary or secondary reasons. Although hypertension is extremely common in polyarteritis nodosa, the mechanism of the hypertension is poorly understood.

Diagnosis

Vessels / Lesions in Different Stages

Vessels and Leeches in Different Stages

Inflammation of different stages can be found simultaneously in polyarteritis nodosa, likely due to the recurrent nature of this disease.

Renal Microaneurysms (On Arteriogram)

Kidney with Micro-aneurysms

Arteriogram is a medical imaging technique used to visualize blood vessels. This is done by injecting a contrast agent into the lumen of the vessel, and using fluoroscopy to observe blood vessel characteristics as the contrast travels through the vessels. This appears as a "string-of-pearls" with angiography. Polyarteritis nodosa causes inflammation of the blood vessels, which can lead to infarction, ulceration, or aneurysm formation anywhere the disease process is taking place. These vessel changes are reflected on arteriogram as multiple aneurysms and constrictions. Polyarteritis nodosa can affect renal arteries and damage the kidneys. This results in various forms of clinical manifestations such as oliguria, edema, hypertension and even renal failure.

Spares Pulmonary Arteries (No Pulmonary Involvement)

Lungs safe inside Spare-tire

Polyarteritis nodosa affects various vessels in the body, but spares the arteries in the lungs. This is important clinically and on the exam, because other vasculitis diseases, such as Wegener's granulomatosis or Churg Strauss syndrome, involve the lungs, and this can be used to distinguish them from polyarteritis nodosa.

Treatment

Corticosteroids

Quarter-on-steroids

Treatment begins with suppressing the immune system, and corticosteroids are used because of their anti-inflammatory properties.

Cyclophosphamide

Cyclops-phosphate-P

Cyclophosphamide is a cancer medication classified as an alkylating agent, working to interfere with the growth of normal and neoplastic cells. It is used in polyarteritis nodosa as it decreases the immune system's response.

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