Microscopic Polyangiitis

Microscopic Polyangiitis is an autoimmune small vessel vasculitis affecting multiple organs in the body. Common causes of microscopic polyangiitis include autoimmune disorders, systemic lupus erythematosus, streptococcus infections and penicillin use. These conditions, infections and medications act as triggers to cause microscopic polyangiitis in patients who are genetically susceptible. The small vessel inflammation leads to disease manifestation in the brain, lungs, intestines and kidneys. Due to inflammation in the vessels of these organs, patients present with neurological defects, pulmonary symptoms, abdominal pain and possibly renal failure. A unique finding in this disease is that the vessels affecting the various organs occur at the same stage during the disease process. The skin manifestation is called a palpable purpura which are lesions that appear as red or purple discolorations on the skin that do not blanch on applying pressure. This disease can be detected by the presence of perinuclear anti neutrophilic cytoplasmic antibodies (P-ANCA) which are antibodies against myeloperoxidase in cells.



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Pathophysiology

Necrotizing Small Vessel Vasculitis

Necrosis-crow with Small Vessels-on-fire

A small vessel vasculitis characterized by segmental fibrinoid necrosis of the media with focal transmural necrotizing lesions.

Signs and Symptoms

Lungs, Kidneys, and Skin Commonly Affected

Lungs, Kidney, and Skin-suit-man

Microscopic polyangiitis commonly involves the lungs, skin, and kidneys. The most common patient presentation includes symptoms of fatigue, cough, dyspnea, palpable purpura, urticaria, and urinary abnormalities like hematuria and proteinuria.

Palpable Purpura

Palpable-paw-print from Purple-cat

Palpable Purpura are lesions that appear as red or purple discolorations on the skin that do not blanch when pressure is applied and can be felt on examination. These lesions are caused by inflammation and hemorrhage of small blood vessels. However, these lesions are not specific to microscopic polyangiitis and can be seen with other types of vasculitis.

Rapidly Progressive Cresentic Glomerulonephritis

Rapids with Crescent-kayak Glow-mare

Rapidly progressive (crescentic) glomerulonephritis (RPGN) is a clinical syndrome manifested by features of glomerular disease in the urine and by progressive loss of renal function caused by autoimmune disease. Patients present with the inability to produce urine (anuria) or production of abnormally small amounts of urine (oliguria) due to kidney injury, as well as myalgias, fever, fatigue, and other symptoms associated with the underlying cause.
There are three types of RPGN. Type I involves anti-glomerular basement membrane antibodies, Type II involves immune complex deposition, and Type III does not involve any immune complexes. Microscopic polyangiitis is a Type III RPGN, also known as pauci-immune RPGN. Diagnosis relies on biopsy showing glomerular crescents formed by the proliferation of parietal epithelial cells.

Diagnosis

Lesions are Same Age

Leeches from Same Age-egg

Usually, all lesions of microscopic polyangiitis are of the same age and in the same stage of inflammation, unlike polyarteritis nodosa, where lesions are at different stages of inflammation.

Non-granulomatous

Nun-granny-llama

Granulomas form in response to inflammation triggered by antigen activation of the body’s immune system. Macrophages and other cells act to wall off a causative agent within the granuloma and prevent further tissue damage. Although other vasculitis diseases involve granulomas, microscopic polyangiitis is

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notable for its lack of granulomatous inflammation.

Minimal Nasopharyngeal Involvement

Minimal Nose-pharaoh

The nasopharynx may be affected in microscopic polyangiitis, though it is less commonly involved, and this characteristic distinguishes it from granulomatosis with polyangiitis (formerly known as Wegener granulomatosis), in which the nasopharynx is involved in the majority of cases.

P-ANCA

PANCAkes

Anti-neutrophil cytoplasmic antibodies (ANCA) are autoantibodies detectable using immunofluorescent staining or immunoassays like ELISA. These antibodies are classified as either c-ANCA or p-ANCA, representing cytoplasmic and perinuclear antibodies, respectively. Microscopic polyangiitis is strongly associated with p-ANCA against myeloperoxidase (MPO), and patients suspected of having the condition should be tested for the presence of this antibody.

Treatment

Corticosteroids

Quarter-on-steroids

Immunosuppression is the main goal of therapy in vasculitis diseases like microscopic polyangiitis. Initial therapy includes glucocorticoids, like prednisone, in combination with another immunosuppressive agent, like cyclophosphamide or rituximab. However, corticosteroids should always be tapered off as soon as remission is achieved.

Cyclophosphamide

Cyclops-phosphate-P

This drug is an alkylating agent, meaning it cross-links DNA and triggers apoptosis. In addition to treating autoimmune conditions, it is also a widely used chemotherapy drug. It can be used initially in combination with corticosteroids and may be considered as part of an extended daily regimen. Be aware of severe cyclophosphamide side effects like hemorrhagic cystitis and myelosuppression.

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