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Rare Juvenile Primary Systemic Vasculitis

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6. ANCA-ASSOCIATED VASCULITIS: Granulomatosis with polyangiitis (Wegener`s, GPA) and Microscopic polyangiitis (MPA)

6.1 What is it?

GPA (previously called Wegener's granulomatosis) is a chronic systemic vasculitis affecting mainly the small blood vessels and tissues in the upper airways (nose and sinuses), lower airways (lungs) and kidneys. The term "granulomatosis" refers to the microscopic appearance of the inflammatory lesions that form small multi-layered nodules in and around the vessels.
MPA affects smaller vessels. In both diseases, an antibody called ANCA (Anti-Neutrophil Cytoplasmic Antibody) is present; hence, the diseases are referred to as ANCA-associated diseases.

6.2 How common is it? Is the disease in children different from the disease in adults?

GPA is an uncommon disease, especially in childhood. The true frequency is not known but it probably does not exceed 1 new patient in 1 million children per year. More than 97% of reported cases occur in the white (Caucasian) population. Both sexes are affected equally in children, whereas in adults men are affected slightly more often than women.

6.3 What are the main symptoms?

In a large proportion of patients, the disease presents with sinus

congestion that does not improve with antibiotics and decongestants. There is a tendency for crusting of the nasal septum, bleeding and ulcerations sometimes causing a deformity known as saddle-nose. Airway inflammation below the vocal cords can cause narrowing of the trachea, leading to a hoarse voice, nosebleeds, chronic ear inflammation mimicking infection), and respiratory problems. The presence of inflammatory nodules in the lungs results in symptoms of pneumonia with shortness of breath, cough and chest pain.
Kidney involvement is initially present in only a small proportion of patients but it becomes more frequent as the disease progresses, causing abnormal urine findings and blood tests for kidney function, as well as hypertension. Inflammatory tissue can accumulate behind the eye balls, pushing them forward (protrusion), or in the middle ears, causing chronic otitis media. General symptoms such as weight loss, increased fatigue, fevers and night sweats are common, as are various skin and musculoskeletal manifestations.
In MPA, the kidney and lungs are usually the main organs affected, although any organ can be affected.

6.4 How is it diagnosed?

Clinical symptoms of inflammatory lesions in upper and lower airways, together with kidney disease, typically manifested by the presence of blood and protein in the urine and increased blood levels of substances cleared by the kidneys (creatinine, urea), are very suspicious of GPA. Blood tests usually indicate increased non-specific inflammatory markers (ESR, CRP) and elevated ANCA titers. The diagnosis may be supported by a tissue biopsy.

6.5 What is the treatment?

Corticosteroids in combination with cyclophosphamide are the mainstay of the induction treatment for childhood GPA/MPA. Other agents suppressing the immune system, such as rituximab, can be chosen according to the individual situation. Once disease activity settles, it is kept under control with "maintenance therapy", usually with azathioprine, methotrexate or mycophenolate mophetil.
Additional treatments include antibiotics (commonly long-term co-trimoxazole), blood pressure lowering agents, drugs against blood clot

formation (aspirin or anticoagulants) and painkillers (non-steroidal anti-inflammatory drugs, NSAIDs).