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POLYARTERITIS NODOSA – A SHORT REVIEW

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Abstract:
Polyarteritis nodosa is a rare disease resulting from blood vessel inflammation (vasculitis), causing damage to organ systems and featuring an extended range of possible symptoms. Polyarteritis nodosa (PAN), also known as Kussmaul disease or Kussmaul-Maier disease, is a vasculitis of the medium and/or small arteries that become swollen and damaged as a result of the attack by rogue immune cells, and the condition may be associated with various atypical presentations. The annual incidence of PAN varies between five and nine cases per million. Men are generally more affected than women in a 2:1 ratio, most frequently between the ages of 40 and 60 years. PAN is more common in people with hepatitis B infection. PAN may affect multiple organs, including skin, kidneys, and gastrointestinal tract, as well as the peripheral and central nervous systems. The inflammatory process causes necrosis of cells and structural components of the artery with aneurysms or stenosis formations. PAN often can culminate in necrosis or hemorrhage of the affected organ.

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INTRODUCTION

Polyarteritis nodosa (PAN), a form of vasculitis, is a serious blood vessel disease in which small and medium-sized arteries become swollen and damaged. This auto immune disease of unknown origin affects the arteries and blood vessels that carry oxygen and blood to the organs and tissues. More adults than children are affected with polyarteritis nodosa; while the disease can strike people of any age, it usually first appears in people between the ages of 40 and 50. Polyarteritis nodosa mimics many diseases and symptoms can vary widely[1]. The disease may be acute with a fever and on-going for a long time; milder but fatal within several months; or appear as a chronic, debilitating disease. The vessels of the kidneys, liver, heart, stomach, and intestines are most often affected. Treatment is focused on decreasing the inflammation of the arteries by suppressing the immune system.

SYMPTOMS

The Most Common Symptoms are:

Fatigue, Fever, Numbness, tingling or weakness of the hands and feet, Pain in the joints, especially the large ones. Skin rash with raised reddish-purple patches and knobs that can be felt along affected arteries, Abdominal pain, sometimes with nausea, and vomiting, Blood in the urine, Swelling, Weakness, Weight loss.

DISCUSSION

Classic PAN is an uncommon disease characterized by predominantly medium-sized artery inflammation leading to involvement of skin, kidney, peripheral nerves, muscle and gastrointestinal tract. Involvement of other organs like lungs, brain and heart is rare. The involvement of kidney is classically limited up to the level of spiral arteries and glomerulonephritis is not seen. This is unlike Microscopic polyangiitis (MPA/microscopic PAN), where systemic vasculitis is associated with small vessel vasculitis causing glomerulonephritis and alveolitis.

Several diagnostic criteria intended to be fulfilled in epidemiological studies or drug trials have been recommended for clinicopathological diagnosis of PAN[2]. The diagnostic criteria proposed by Ozen et al in 1992 for the diagnosis of childhood PAN included 2 major and 10 minor clinical and laboratory features without the immediate need of angiography and biopsy. In this retrospective study of 31 patients, there was a good correlation of these criteria with histopathological diagnosis of PAN. The authors however, proposed to use these criteria for early diagnosis after a prospective study validation. This is a clinical criteria aiding in an early diagnosis while awaiting a definite diagnosis by angiography or biopsy. One major feature in this study included renal parenchymal involvement with proteinuria, hematuria or rapidly progressive glomerulonephritis. As per the current understanding and the classifications proposed after the Ozen's criteria, the presence of proteinuria and or active urine sediments suggest the diagnosis of Wegener's granulomatosis and MPA, while classic PAN is associated with bland urine sediments. It may be likely that at least some patients from Ozen's study had either of these two conditions as also evidenced from pulmonary infiltrates or hemoptysis in some of these patients. MPA is a disease with autoantibody (p-ANCA) association in most, while classic PAN lacks the antibody in almost all, making further distinction between these two conditions[3]. Our patient had a presentation where acute onset of gangrene was attributable to classic PAN in view of unexplained hypertension (suggestive of renal artery involvement) with no evidence of glomerulonephritis. Other features aiding in the diagnosis were polymorphonuclear leucocytosis, elevated ESR, negative autoantibodies (ANA, ANCA), neuropathic pain and no apparent infectious etiology. Although histopathology and or angiography is important for confirmation of PAN, practical limitations, as in our patient, may lead to situations where one may provisionally diagnose the disease as per Ozen's clinical criteria without the aid from these specific tests. Despite lack of histopathology and angiography, our patient satisfied the required 3/10 of the ACR criteria for diagnosis of PAN but only 5 minor of the Ozen's criteria lacking major feature of renal parenchymal involvement, which we however contested above.

TREATMENT

Steroids, with or without cyclophosphamide is the recommended mode of therapy for PAN. Our patient showed significant improvement with early

institution of this therapy. Despite being a rare disease, PAN should be included as a differential diagnosis in children with gangrene and or unexplained hypertension to enable early diagnosis and proper management which can lead to reduced morbidity and mortality[4].

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