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Pan : A Vanishing Vasculitis

KEYWORDS

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ABSTRACT *Polyarteritis nodosa (PAN), is a collagen vascular disease of medium and small arteries affecting multiple organs and tissue having strong association with hepatitis B virus and hepatitis C virus . It may present in various forms affecting multiple systems. Here we present a case of Polyarteritis nodosa, who presented with mono-neuritis multiplex along with esophageal ulcerations .*

INTRODUCTION:

Polyarteritis nodosa (PAN), first described by Kussmaul and Maier in 1866 , 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 [1-3]. Classic polyarteritis nodosa, is a systemic vasculitis characterized by necrotizing inflammatory lesions that affects medium-sized and small muscular arteries, preferentially at vessel bifurcations, resulting in microaneurysm formation, aneurysmal rupture and thrombosis. Limited forms of polyarteritis nodosa is also known, and skin being the most common organ to be involved. Males, young adult to middle age, are more commonly affected. Diagnosis is mainly based on histopathology and vascular angiography along with a strong clinical background.

CASE HISTORY:

A 46 year male presented with complaints of low backache since 3 months along with right foot drop and weakness of bilateral lower limbs and hands since one and half month .He also had daily low grade fever and dysphagia since one month and developed left foot drop since 3 days .

On examination he was febrile, Pulse was 110 beats/min, BP 100/60mmHg and was pale. Neurological examination showed that he had hypotonia and wasting in all limbs. Power was 4/5 in all limbs except for ankle joint ,where power was 0/5. All deep tendon reflexes were diminished with absent ankle jerk . All sensations were decreased in both upper limbs and over both the soles and dorsum of feet. Higher functions and cranial nerves were normal.Jaw claudication was observed.

On Investigations, his hemoglobin was 8.0 gm.% ,total Leucocyte Count 18000/mm^3(N: 76% L:08 % E: 10%), platelet count 7.63 lakh/mm^3, ESR 109 mm/h, CRP 23.8 mg/L, R Factor –26.3 IU/ml. HIV, HBsAg and HCV were negative. Renal and liver functions were within normal limits. Urine Routine, Chest X Ray and USG abdomen pelvis were normal. MRI Cervical spine was normal.. Gastroscopy was done in view of severe dysphagia and was suggestive of esophageal ulcerations. Bone marrow was reactive and Antinuclear antibody, Protein electrophoreis, cANCA and pANCA (anti-nuclear cytoplasmic antibody) were all negative. With this clinical background and with a strong suspicion of polyarteritis nodosa , a CT Abdomen pelvis with renal angiography was done which showed beading and tortuosity in interlobar arteries in left kidney (Figure 1) , very typical of polyarteritis nodosa. Right sural nerve biopsy was also done which showed findings consistent with vasculitic peripheral neuropathy. He was then started on steroids and cyclophosphamide and responded well.

DISCUSSION:

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 immune cells, and the condition may be associated with various atypical presentations [1-3]. PAN may affect multiple organs, including kidneys, skin and gastrointestinal tract, as well as the central and peripheral nervous systems [4]. Patients may present with a variety of symptoms, the most common of which are fever , neuropathy and weight loss [5]. Mononeuritis multiplex is the most common neurological manifestation. Esophageal ulceration may also be seen in 13% of patients [6] ,as in our case. Histopathologic demonstration of vascular inflammation in medium-sized or small arteries is important to sustain the diagnosis of PAN .Typical Visceral angiography are seen in PAN which helps in confirming the diagnosis when histologic diagnosis of vasculitis cannot be achieved . Typical arteriographic lesions in PAN are arterial saccular or fusiform microaneurysms , which usually coexist with stenotic lesions, predominantly in kidney, mesenteric and hepatic artery branches[6]. Vascular lesions are characteristically segmental and occur mainly in branching points[7]. Steroids and cyclophosphamide form the main pillars of treatment . Cyclophosphamide is recommended to induce remission and azathioprine or methotrexate safer immunosuppressive agents, are advised to maintain remission[8].

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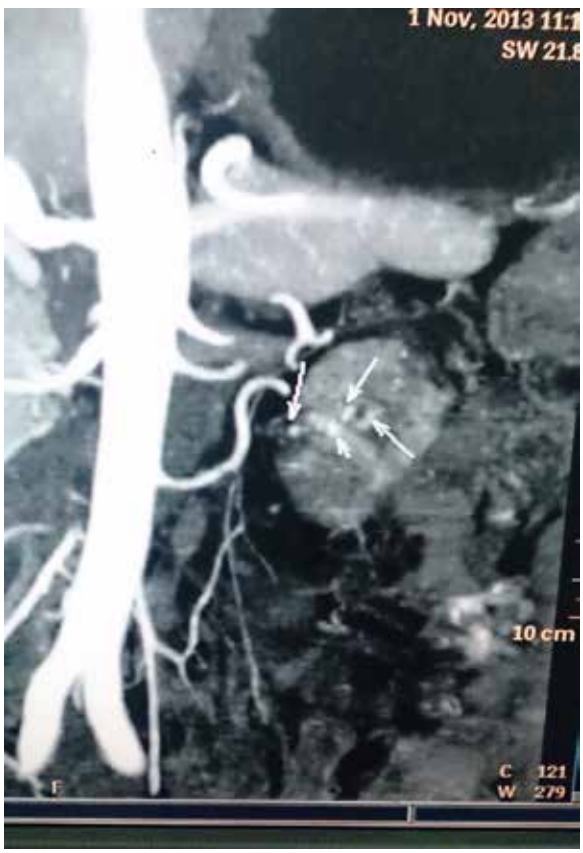
For hepatitis B and C associated PAN , steroids along with anti-virals form the treatment line. Surgery may be required for some disease complications, such as perforation, ischemia or hemorrhage of the gastrointestinal organs or kidneys [9]. Though PAN is a vanishing vasculitis, it still exists and a high index of suspicion, early diagnosis and early treatment is must to prevent further dreaded complications.

Figure 1



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