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Case Report

Small vessel vasculitis: a rare case report

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ABSTRACT

Small-vessel vasculitis is a rare immunological disease that affects arterioles, capillaries and venules. A 1-year-6-month-old male of non-consanguineous marriage came to the OPD with his mother with a complaint of swelling over bilateral legs for 3 days and rash over his body for 1 day. The swelling was increasing and gradually ascending upward. Systemic examination revealed S1, S2 +, and no murmur in CVS. Soft, NT, liver, spleen not palpable. The patient was admitted and routine investigations were sent. The case reported above had clinical spectrum with skin involvement as palpable purpura, nervous system involvement as peripheral neuropathy and proximal myopathy and hematological involvement, gastrointestinal involvement as vague abdominal pain.

Keywords: Vasculitis, Proximal myopathy, Wegener's granulomatosis, Vessel

INTRODUCTION

Vasculitides encompass group of inflammatory conditions affecting blood vessels with severe consequences including tissue ischemia, structural abnormalities, such as aneurysms/dissections, and end-organ damage.[1] Different forms are commonly classified based on the blood vessel size involved as large-vessel, medium-vessel, and small-vessel vasculitis. Small vessel vasculitis (SVV) is characterized by inflammation of parenchymal arteries, arterioles, capillaries and venules and is subdivided into antineutrophil cytoplasmic antibody-associated vasculitis, immune-complex-mediated vasculitis (including HSP), Wegener's granulomatosis (WG), microscopic polyangiitis (MPA), renal limited vasculitis, and Churg-strauss syndrome (CSS).[2,3] Vasculitis may be confined to a single organ such as skin/involve several organs simultaneously.[4]

CASE REPORT

A 1-year-6-month-old male of non-consanguineous marriage came to the OPD with his mother with a complaint of swelling over bilateral legs for 3 days and rash over his body for 1 day. The swelling was increasing and gradually ascending upward. Petechiae were mainly over the thigh and buttocks. The patient was vitally stable with pallor on examination. The patient was apparently all right 15 days back when he had an intermittent moderate undocumented fever for which he was taken to a local physician and treated with injectables (details not available). In the next 7 to 8 days fever settled. However, swelling over his left knee was noticed by his mother. History of antibiotic course in the recent past and present. The patient gave no history of oozing, similar complaints in siblings and parents, bruising tendency, change in bowel bladder habits, trauma and similar episode in past. Figure 1 represents the clinical features of the case.

Systemic examination revealed S1, S2 +, and no murmur in CVS. Soft, NT, liver, spleen not palpable. AEBE and chest were clear. No deficit was seen in the CNS system. Cutaneous examination revealed multiple well-defined haemorrhagic papules to plaque present over the bilateral lower limb. Groin, thighs, buttocks and bilateral upper limbs. Multiple well-defined hyperpigmented macules to

patches were present over the bilateral lower limb. The scalp was diffuse oedema present over the occipital region. Palms and soles, genitals, nails and oral symptoms were within normal limit.

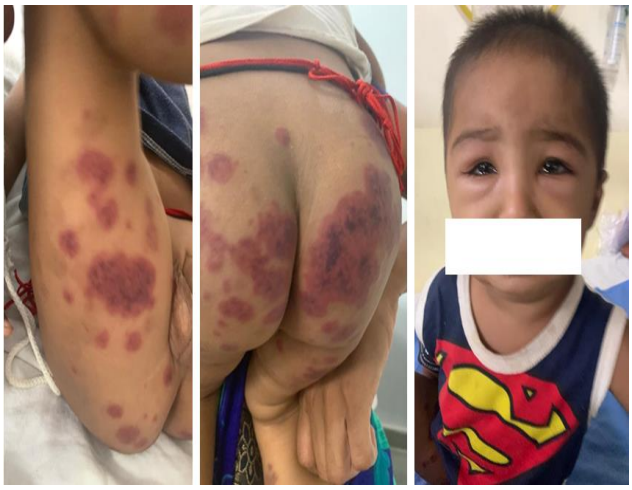

Figure 1: Clinical features of case.

The patient was admitted and routine investigations were sent. Urine examination was within normal limits. 2D echo showed the normal study. Histopathology revealed stratified squamous epithelium with pigmented basal cell layer. Subepithelial tissue showed neutrophilic inflammatory infiltrate in the wall of blood vessels. Histological features suggested leukocytoclastic vasculitis (Figure 2).

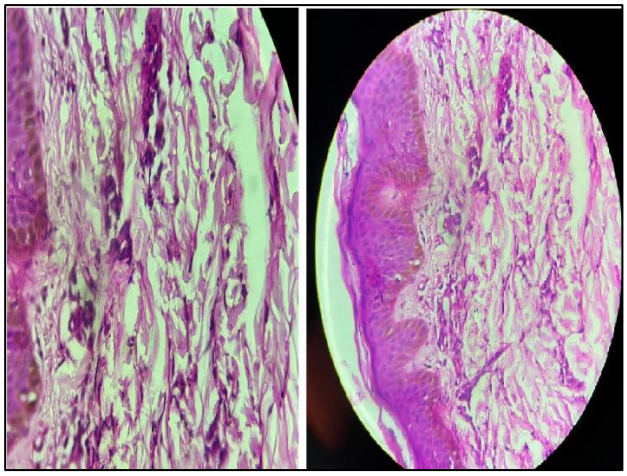

Figure 2: Histopathological features.

Treatment given during the hospitalization period were inj. Ceftriaxone (1ml=100 mg) 5.5 ml IV BD for 3 days, inj piptaz (1 ml=100 mg) 5 ML IV TDS for 7 days, syp azithromycin (5 ml=200 ml) 3.5 ml PO OD for 5 days. On follow-up after 10 days, the skin rashes were resolved, with continued treatment, presently patient has improved hematologically, and neuropathy and proximal myopathy has improved (Figure 3). The patient is now under regular follow-up.

Figure 3: Follow up after 10 days.

DISCUSSION

Vasculitis may affect the large, medium, or small blood vessels.[5] Small-vessel vasculitis can be characterised as ANCA-associated or non-ANCA-associated. Microscopic polyangiitis, Wegener's granulomatosis, Churg-Strauss syndrome, and drug-induced vasculitis are all symptoms of ANCA-associated small-vessel vasculitis. Vasculitis can affect a single organ, such as the skin, or it can affect numerous organs at the same time.[4] Vasculitis is a clinically and pathologically defined condition characterised by inflammation and damage to blood vessels.

According to the size of the vessel affected, vasculitis can be classified into Large vessel: polymyalgia rheumatica, Takayasu's arteritis, temporal arteritis; Medium vessel: Buerger's disease, cutaneous vasculitis, Kawasaki disease, polyarteritis nodosa; small vessel: Behçet's syndrome, Churg strauss syndrome, cutaneous vasculitis, Henoch Schönlein purpura, microscopic polyangiitis, Wegener's granulomatosis.[4]

Acute hemorrhagic edema of infancy (AHEI) is a leukocytoclastic vasculitis that is frequently misdiagnosed as Henoch-Schonlein Purpura (HSP). Although the current categorization of paediatric vasculitis does not include AHEI as a vasculitis identity, there were 100 instances of this condition recorded globally from the initial publication by Snow in 1913 and the year 2004.[6-9] AHEI is distinguished by the classic trio of visible purpuric skin lesions, edema, and fever, and it is more prevalent in male aged 4-24 months, with no ethnic predominance.[10,11]

CONCLUSION

The case reported above had clinical spectrum with skin involvement as palpable purpura, nervous system

involvement as peripheral neuropathy and proximal myopathy and hematological involvement, gastrointestinal involvement as vague abdominal pain. Although our case had no renal and pulmonary involvement, skin lesions and skin biopsy were diagnostic for SVV

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