An Unusual Case of Henoch Schönlein Purpura Presenting as Mononeuritis Multiplex

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Abstract
IgA vasculitis [Henoch Schönlein Purpura (HSP)] is characterized by leukocytoclastic vasculitis involving the small vessels with deposition of immune complexes that contain IgA. Clinical signs include purpura, arthralgia, glomerulonephritis and gastrointestinal involvement. HSP with nervous system involvemen is very uncommon. We report a 12 year old boy with IgA vasculitis who presented with typical rash and Mononeuritis Multiplex and responded well to steroid therapy. HSP presenting as Mononeuritis Multiplex has rarely been reported in children. Clinicians must know of this rare complication of HSP which, when present, may be well managed with steroids.

Keywords: Henoch Schönlein Purpura; IgA Vasculitis; Purpura; Mononeuritis Multiplex; Radial Mononeuropathy

Introduction
IgA vasculitis previously known as Henoch Schönlein purpura (HSP) is characterized by leukocytoclastic vasculitis involving the small vessels, with deposition of IgA containing immune complexes. Clinical signs include purpura, arthralgia, glomerulonephritis and gastrointestinal involvement [1]. The vasculitis of HSP can involve the nervous system and can add significant morbidity of the illness. Headache, mental status changes and seizures are the most frequent neurologic symptoms in HSP. Peripheral nervous system involvement such as mononeuropathies, polyradiculopathies is extremely rare presentation [2,3].

Case Report
We report a case of a 12 year old male who presented to us with complaints of palpable purpuric rash on his lower limbs for last ten days, inability to abduct his right arm, difficulty in walking due to weakness in limbs for last two days and abdominal pain for two days prior to admission. There was no history of abnormal movement of limbs, altered sensorium, fever, red colored urine or joint pains. His weight was 73kgs, height 165 cm, BMI was 26.81Kg/m2. His vitals were within normal range and he was hemodynamically stable. There was extensive palpable purpura involving his both lower limbs (Figure 1). Neurological examination revealed right arm abduction weak (power 2/5) along with preserved deep tendon reflexes and normal rest of the muscles of upper limb. Left upper limb was with normal tone and power. Lower limb examination revealed right Gluteus Maximus weak. Rest of the muscles in lower limb showed normal power. Higher mental functions and sensory system examination was normal.

A complete blood count with differential analysis, liver function test, renal function test, coagulation test, chemistry panels were done which were normal. Other relevant tests including creatinine phosphokinase (CPK), Antinuclear antibody (ANA), Anti-neutrophil cytoplasmic antibodies (ANCA), C3 and C4 levels were normal.

HBsAg, Anti HCV, and HIV were non reactive. Ultrasound abdomen revealed grade 1 fatty changes in liver, rest scan was normal. Urine analysis showed proteinuria with 6-8 RBC/hpf. Urine protein:creatinine ratio was 0.31. A skin biopsy from lower limb rash showed neutrophilic infiltrates around the small vessels suggestive of leukocytoclastic vasculitis. Nerve conduction study showed right Axillary Neuropathy with bilateral Peroneal Neuropathy. Possibility of Mononeuritis Multiplex without conduction block was kept. There was no relief in his neurological status for the next 3 days.

According to the EULAR/PRINTO/PRES criteria, he was diagnosed as HSP. In view of persistent neurological involvement he was started with IV Methyl-Prednisolone 1 gram a day for 5 days followed by oral Prednisolone 40 mg /day. The patient responded well to steroid therapy and power of the involved muscles showed improvement and he was discharged after 5 days of methyl-prednisolone. Subsequent neurological examination done two weeks later in follow up visit revealed near normal power in the involved muscle groups. His steroids were tapered subsequently over a period of four weeks. He is fine on a follow up of eight months.

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Figure 1: Extensive palpable purpura on both lower limbs.
Discussion

Vasculitic neuropathy may occur as isolated or together with involvement of other organs [1,2]. Vasculitis affecting the peripheral nerves commonly presents as subacute, progressive, asymmetric sensori-motor polyneuropathy or Mononeuritis multiplex, and sometimes as mono-neuropathy, pure sensory-neuropathy, cranial nerves neuropathy, or autonomic neuropathy.

Systemic vasculitis with involvement of the peripheral nerves can be further subdivided into primary (Churg-Strauss syndrome, Wegener granulomatosis [WG], Takayasu arteritis, Panarteritis nodosa, Thrombangiitis obliterans, Kawasaki disease, Behçet disease, microscopic polyangiitis, IgA vasculitis) or secondary systemic vasculitis (infection, sarcoidosis, malignancy, radiation, drugs, connective tissue diseases or diabetes). Churg strauss syndrome, WG and microscopic polyangiitis are the most common vasculitides that affect the peripheral nervous system [1,4].

Henoch Schönlein purpura or IgA vasculitis represents a multisytem vasculitis with occasional involvement of the CNS, manifesting as headache, seizures, ataxia, or altered mental state. Very rarely the Peripheral Nervous System is involved in the form of peripheral facial palsy, Guillain-Barre syndrome (GBS), brachial plexopathy or peroneal neuropathy [3,5,6]. Garzoni et al [5] did a systematic review of all HSP cases with neurological involvement who presented between 1969-2009. Peripheral facial palsy, GBS, brachial plexopathy were the commonest presentation in children with cranial or peripheral neuropathy group with Mononeuritis multiplex being reported in only one patient. Agras et al [6] reported a 12-year-old boy with HSP who presented with left lower brachial plexopathy who responded well to intravenous pulse steroid therapy.

Involvement of the Peripheral Nervous System in systemic vasculitis is as a result of infiltration of the vasa nervorum or the epineural arteries by inflammatory cells [6]. Infiltration of the vascular wall and resulting damage facilitates thrombosis and subsequent ischemia. Damage to the blood-nerve-barrier is induced by pro-inflammatory cytokines, oxidative stress-derived molecules, or metallo-proteinases [7]. Altered expression and function of adhesion molecules or leukocytes (primary T-cell) and endothelial cell activation play a key role in the pathogenesis [8, 9].

The age at disease onset is considered to be an important factor for disease severity and outcome in HSP [10] and this may be true with our case also as he was older as compared to the average HSP case. It has been reported that the incidence of nephritis and HSP recurrences increases with the age in childhood HSP [11,12].

Though the long term prognosis of HSP is attributable almost entirely to the kidney disease, sometimes extra renal involvement may produce substantial morbidity and mortality. Corticosteroids are advised in patients with Henoch Schönlein purpura complicating peripheral or cranial neuropathy [3,5,13]. Most of these conditions tend to have full spontaneous recovery. Our case responded well to steroid therapy.

Conclusion

HSP with nervous system involvement is very uncommon. HSP presenting as mononeuritis multiplex has rarely been reported in children. The clinicians must know of this rare complication of HSP, which may be well managed with steroids.

References


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