INTRODUCTION:

Henoch Schonlein 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 occurs primarily in children. It is uncommon in people over 40 yrs. Adult onset HSP with nervous system involvement is still uncommon but clinically relevant. We present a case of 40 yr old male with adult onset HSP with neurological involvement in the form of left radial mononeuropathy.

Key-words:
Adult onset Henoch Schonlein Purpura (HSP), Mononeuropathy, Radial mononeuropathy

CASE REPORT:

We report a case of a 40 year old male chronic smoker with complaints of fever with palpable purpuric lesion on his lower limbs, arthritis in both knee and ankle joint and diffuse abdominal pain for 5 days prior to admission. A complete blood count with differential analysis, liver function test, renal function test, coagulation test, chemistry panels were done which was normal. Urine analysis showed minimal proteinuria but no RBC cast or cells. On the second day he developed subcutaneous edema over the forehead, his purpuric lesions were progressing proximally. He had an episode of blood in stool, for which stool occult blood came positive. His blood pressure recorded upto 160/100 mmhg. A skin biopsy of the lower limb showed neutrophilic infiltrates around the small vessels suggestive of leukocytoclastic vasculitis. On the third day of admission patient developed wrist drop on the left side suggestive of left radial nerve palsy. Nerve conduction study showed left radial mononeuropathy. According to the EULAR/PRINTO/PRESS criteria, he was diagnosed as HSP and started with IV methylprednisolone 1g/day for 3 days and continued oral prednisolone 40 mg/day. ANA, p-ANCA and c-ANCA was done to rule out polyarteritis nodosa which came as negative. The patient responded well to steroid therapy.

Figure 1: Skin biopsy showing neutrophilic infiltrates

Abstract

Henoch Schonlein 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 occurs primarily in children. It is uncommon in people over 40 yrs. Adult onset HSP with nervous system involvement is still uncommon but clinically relevant. We present a case of 40 yr old male with adult onset HSP with neurological involvement in the form of left radial mononeuropathy.
DISCUSSION:

Although the long term prognosis of Henoch-Schonlein syndrome is almost entirely attributable to the kidney disease, some rare extrarenal features may produce substantial morbidity and mortality. Reports available until the mid-seventies that deal with the CNS involvement in Henoch-Schonlein syndrome was aggregated by French authors in two reviews.

In Churg-Strauss syndrome, WG and microscopic polyangiitis, the most common vasculitides that affect the peripheral nervous system, the corresponding clinical patterns are mononeuritis multiplex, polyneuropathy, radiculopathy and neural plexopathy. In Henoch-Schonlein patients with cranial or peripheral neuropathy with or without hypertension following lesions were noted: peroneal neuropathy, peripheral facial palsy, Guillain–Barre syndrome, brachial plexopathy, posterior tibial nerve neuropathy, femoral neuropathy, ulnar neuropathy and mononeuritis multiplex. Vasculitis may cause inflammation in the walls of the vasa nervorum and induce critical ischemia to the nerves. However sometimes lesions may result from compression by hematoma or localized oedema.

Corticosteroids is advised in patients with Henoch-Schonlein purpura complicating peripheral or cranial neuropathy. However most of these conditions tend to have full spontaneous recovery.

CONCLUSION:

Henoch-Schonlein purpura in adult population is quiet uncommon. Adult onset HSP with nervous system involvement is still uncommon but clinically relevant. Hence we present this case report to help clinicians managing HSP with rare complications.

REFERENCES:

1. A. Bulun, R. Topaloglu, A. Duzova, I. Saatci, N. Besbas, A. Bakaloglu Ataxia and peripheral neuropathy: rare manifestations in Henoch-Schönlein purpura
2. Oxford Journals Nervous system dysfunction in Henoch–Schönlein syndrome: systematic review of the literature  Luca Garzoni1, Federica Vanoni1, Mattia Rizzit2, 3, Giacomo D. Simonetti2, 3, Barbara Goeggel Simonetti2, Gian P. Ramelli1 and Mario G. Bianchetti1
3. The clinical implications of adult-onset henoch-schonlein purpura. Warit Jithpratuck,1 Yasmin Elshenawy,2 Hana Saleh,1 George Youngberg,2 David S Chi,1 and Guha Krishnaswamy
4. An adult case of Henoch-Schönlein purpura complicating common peroneal nerve mononeuropathy.10.1007/s10165-008-0121-5, Modern Rheumatology, Kei Ohnuma, O
6. Henoch Schonlein purpura- S Lanzkowsky, L Lanzkowsky
7. Henoch Schonlein Purpura in adults –maria jose lopez miller, Javier Alberto Cavallasca
9. Dillon M J Henoch Schonlein purpura: recent advances
10. de Montis G , Turpin JC. Rheumatoid purpura and neurologic manifestations.