University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2019 , Vol. 5(3)

TWO INTERESTING CASES OF VASCULITIC NEUROPATHY

THASEEN A
Department of Neurology,MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract : TWO INTERESTING CASES OF VASCULITIC NEUROPATHY Abstract Peripheral neuropathy is an important, and often the presenting clinical feature of the vasculitides. We report two cases presenting as peripheral neuropathy with connective tissue diseases. Case vignette 1A 19yr old female presented with benumbed sensation over the right leg foot, with tripping of right toes while walking for past 3 months, with similar complaints in the left leg 1 month later. On examination she had cushingoid features with purpuric rash over the right palm, and wasting over lateral aspect of right leg with distal hypotonia. She had involvement of both the common peroneal nerves and right tibial nerve. There was sequential involvement in the pattern of mononeuritis multiplex. Nerve biopsy was suggestive of vasculitic neuropathy. Skin biopsy showed fibrinoid necrosis of medium arteries involving intradermal portion suggestive of Polyarteritis nodosa. Case vignette 243year old female, presented with pins and needles and benumbed sensation involving left foot 4 months back which progressed up to the knee in 3 weeks, associated with weakness of left foot, and with similar complaints in the right foot a month later.
On examination she had hypotonia of left foot with weakness of dorsiflexion more than plantar flexion of both lowerlimbs. She also had decreased pain, touch, temperature over both foot upto one-third of leg, left more than right. Her RA factor and anti- CCP were positive suggestive of rheumatoid arthritis. ConclusionThese two cases are presented to enlighten the varying presentations of vasculitic neuropathy in the setting of connective tissue diseases.
Keyword :RA factor - rheumatoid factor, anti-CCP - anti cyclic citrullinated peptide.

TWO INTERESTING CASES OF VASCULITIC NEUROPATHY

Introduction:

Peripheral neuropathy is an important, and often the presenting clinical feature of the vasculitides. Its recognition is critical to attain an early diagnosis in these disorders where the final outcome can be greatly influenced by early therapeutic intervention. The vasculitides are a group of heterogeneous disorders which present with a variable and complex clinical picture. We report two cases presenting as peripheral neuropathy with connective tissue diseases.

Case vignette 1:

A 19yr old female presented with benumbed sensation over the right leg & foot, with tripping of right toes while walking for past 3 months. After about a week, she developed twisting of right foot while walking with unaware slippage of slipper from right foot. One month later she developed similar complaints in the left leg. She developed flailness of both lower limbs with buckling of knees while walking since last 2 weeks. Prior to this illness she had recurrent episodes of swelling over small joints, and was diagnosed as unclassified vasculitis.
She also had alopecia & photosensitive rash during the episode. She was treated with steroids and methotrexate, 5 yrs back for about 3 yrs. Her detailed general and neurologic examination revealed purpuric rash over right palm, cushingoid features, striae over abdomen and both thighs. Higher mental function and cranial nerves were normal.
Wasting was seen over lateral aspect of right leg with distal hypotonia, knee flexor weaker than extensor on the right, ankle dorsiflexion weaker than plantar flexion. In the left lower limb, knee flexors and foot dorsiflexors were weak with bilateral foot eversion weakness, ankle reflex bilaterally absent with rest of reflexes diminished. Pain, touch and temperature were diminished over the right lower limb on the lateral aspect of leg, and over the dorsum of foot by 60%. Vibration was diminished over both lateral malleoli. Pain, touch, temperature were diminished on the lateral aspect of left leg and over dorsum of left foot by 30%. Joint position sense was absent over the right toes. The pattern of motor and sensory involvement was suggestive of bilateral common peroneal and right tibial nerve involvement. Routine investigations revealed elevated ESR with normal counts. HIV and VDRL were negative. Antibody profile for vasculitis and serum cryoglobulin were negative. CSF analysis and C3, C4 levels were normal. NCS showed sensorimotor axonal changes in bilateral peroneal and right tibial nerves. Nerve biopsy of left sural showed intact perineurium, and epineurium showed prominent blood vessels with focal neovascularisation, perivascular lymphocytic infiltration.
Focal acute axonal breakdown was seen, myelin stain showed non-uniform loss of myelinated fibres with regenerating clusters, suggestive of vasculitic neuropathy. Skin biopsy showed fibrinoid necrosis of arteries involving intradermal portion suggestive of Polyarteritis nodosa.
She was started on IV methylprednisolone followed by oral steroids and IV cyclophosphamide. By two weeks she was ambulant without support and distal lowerlimb weakness started improving.

Case vignette 2:
43year old female, presented with pins and needles and benumbed sensation involving left foot 4 months back which progressed up to the knee in 3 weeks, associated with weakness of left foot. One month later she developed similar complaints in the right foot. She had recurrent small joint pain with swelling involving both the hands with early morning stiffness for the past 4 years. She is known case of DM on insulin. On examination she had hypotonia of left foot with weakness of dorsiflexion more than plantar flexion of both lowerlimbs. She also had decreased pain, touch, temperature over both foot upto one-third of leg, left more than right. There was no cranial nerve involvement or peripheral nerve thickening. Investigations revealed increased total count, polymorphs with elevated ESR. Her RA factor and antiCCP were positive. Her nerve conduction studies showed axonal changes involving bilateral tibial and common peroneal nerves. CSF study was normal and nerve biopsy of left sural was suggestive of vasculitic neuropathy. She was started on steroids and oral methotrexate and showed improvement by 6 weeks.

fig 1Purpuric rash over left palm in case 1

fig 2 Asymmetric polyneuropathy as left foot drop in case 2

Discussion:
Vasculitis affecting the peripheral nerves predominantly manifests as subacute, progressive, asymmetric sensorimotor polyneuropathy or mononeuritis multiplex, and more rarely as painful mononeuropathy, pure sensory neuropathy, neuropathy of the cranial nerves, plexopathy, or as autonomic neuropathy. Vasculitis of the peripheral nerves can also present with a distal symmetric neuropathy. Vasculitic neuropathy may occur as isolated or non-isolated (systemic) presentation together with involvement of other organs. Systemic vasculitis is categorized as either primary, for which there is no known cause (Takayasu syndrome, giant cell arteritis, classical panarteritis nodosa (PAN), thrombangitis obliterans, Kawasaki disease, Churg-Strauss syndrome (CSS), Wegener granulamotosis, cryoglobulinemic vasculitis, Behcet's disease, microscopic polyangitis, Schoenlein Henoch purpura), or secondary as a complication of an autoimmune connective tissue disorder, infection, sarcoidosis, malignancy, medication, radiation, or diabetes. The first patient was a case of PAN who manifested with mononeuritis multiplex, with sequential involvement of individual nerves, in a distal to proximal and asymmetric pattern.
The neuropathy was associated with pain in the distribution of the affected nerve, suggesting involvement of both motor and sensory modalities. Long nerves of the lower extremities are affected more frequently than those in the arms. Polyarteritis nodosa (PAN) is a systemic necrotizing medium-size-vessel vasculitis with variable clinical manifestations. Diagnosis is confirmed by histology or angiography. PAN, perhaps the most classical of the vasculitis with peripheral nerve involvement, is a rare disorder, the incidence in the general population ranging from 4.6 to 9.0 per million. It affects men and women of all racial groups with predominance between ages 40 to 60. Fifty to seventy (50 to 70%) percent of the diagnosed cases have neuropathy. Two percent (2%) of the patients showed cranial nerve involvement. Sensory complaints can vary from dysesthesia to pain. Motor deficits usually present abruptly. The severe and sometimes fulminant neuropathy associated with PAN can also be associated with aggressive systemic disease leading to organ failure. The second case was asymmetric peripheral polyneuropathy due to rheumatoid arthritis. Neuropathy an extra-articular manifestation in rheumatoid arthritis may not only result from entrapment or drug toxicity but also from vasculitis. Rheumatoid arthritis can evolve into rheumatoid vasculitis in 2-15% of the cases and half of these patients may develop vasculitic neuropathy. Peripheral neuropathy occurs in 10% of patients with RA. Vasculitis affects small to medium sized arteries, usually sparing arterioles, capillaries, or venules. Involvement of the PNS in ystemic vasculitis results from infiltration of the vasa nervorum or the epineural arteries by inflammatory cells. Infiltration of the vascular wall facilitates thrombosis and consecutive ischemia. The infiltration is facilitated by breaches in the blood nerve barrier, which is not as effective as the blood brain barrier.

Nerve biopsy:
Only few percentage of diagnosed patients have a positive nerve biopsy. Combined Nerve and muscle biopsy adds to the overall diagnostic yield than either alone. The absence of a positive tissue biopsy does not exclude the disorder. Biopsy of "symptomatic sites" seems to improve the diagnostic yield. Both of the patients desctribed, showed positive nerve biopsy.

Conclusion:
Vasculitic neuropathy should be considered if the clinical presentation is either subacute asymmetric axonal sensorimotor polyneuropathy, or painful mononeuropathies or mononeuritis multiplex. The first case of PAN manifesting as mononeuritis multiplex is presented, as it is a rare systemic vasculitis of the small or medium-sized arteries. The second case of RA presenting as asymmetric polyneuropathy is presented, as neuropathies are less common in RA, and they can become a frequent diagnostic encounter to the clinician, given the frequency of RA in the general population.

References:
1. Griffin, J., Vasculitic Neuropathy, Rheum Clin. 2003 Nov; 27 (4): 751-759.
2. Guillevin, L, et al, Polyarteritis Nodosa, Microscopic Polyangitis, and Churg-Strauss Síndrome: Clinical Aspects, Neurologic Manifestations, and Treatment. Neurol Clin. 1997 Nov;15(4):865-885.
3. Pagnoux C, Guillevin L. Peripheral neuropathy in systemic vasculitides. Curr Opin Rheumatol. 2005;17:41-48.
4. Schaublin GA, Michet CJ Jr, Dyck PJ, Burns TM. An update on the classification and treatment of vasculitic neuropathy. Lancet Neurol. 2005;4:853-865.

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Medicine and Medical Specialities

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Medicine and Medical Specialities

An Initiative of The Tamil Nadu Dr. M.G.R. Medical University
University Journal of Medicine and Medical Specialities