Uncommon Treatment of Mononeuritis Multiplex in a 37-year-old Male

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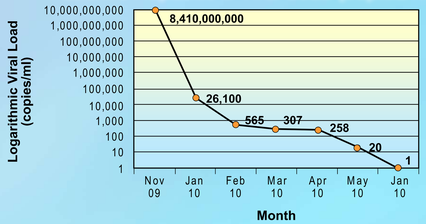
Introduction:

Mononeuritis multiplex is a painful condition characterized by asymmetric loss of sensory and motor function of individual peripheral nerves. It is a common presentation of a multitude of conditions, including diabetes mellitus, vasculitis, infections and various autoimmune disorders.

Case Presentation:

A 37-year-old Caucasian male with a history of hypertension secondary to autosomal dominant polycystic kidney disease (ADPKD) presented to the emergency department with a three-month history of progressive lower extremity weakness and paresthesias, leading to severe ambulatory dysfunction. Physical exam revealed stocking and glove pattern weakness in the distal upper and lower extremities bilaterally, along with severe pain to palpation. Diagnostic studies of an MRI of the brain and lumbar puncture were unremarkable. Laboratory data showed an elevated creatinine, transaminitis, an erythrocyte sedimentation rate of 80 and proteinuria. ELISA for HIV was negative, however, hepatitis B serology was positive for both surface and envelope antigens, indicating active viral replication. Suspicion for a vasculitic etiology of his symptoms led to an EMG and sural nerve biopsy, both of which had findings consistent with vasculitis. Collectively, his symptoms and diagnostic data met the criteria for polyarteritis nodosa (PAN), a vasculitis of medium-sized vessels. Treatment of PAN typically involves corticosteroids and the immunosuppressant medication, cyclophosphamide, with close monitoring of renal function. However, when PAN presents concurrently with active hepatitis B, the combination of corticosteroids, antiviral therapy and plasmapheresis is more favorable and was the course of action taken in this case. Upon discharge, the patient was continued on Entecavir, an antiviral therapy for chronic hepatitis B. Within weeks, the patient's paresthesias decreased, allowing him to ambulate with a cane.

Patient's Hepatitis B Viral Load (copies/ml)



American College of Rheumatology 1990 criteria for the classification of polyarteritis nodosa (PAN). Classified as PAN if at least 3 of the 10 criteria are present:

- Weight loss > 4 kg: Loss of >4 kg body weight since illness began, not related to dieting or other factors.
- Livedo reticularis: Mottled reticular pattern over the skin of portions of the extremities or torso.
- Testicular pain/tenderness: Pain or tenderness of the testicles, not due to infection, trauma or other causes.
- Myalgias, weakness or leg tenderness: Diffuse myalgias (excluding shoulder or hip girdle) or weakness of muscles or tenderness of leg muscles.
- Mono- or polyneuropathy: Development of mononeuropathy, multiple mononeuropathies or polyneuropathy.
- Diastolic BP >90 mmHg: Development of hypertension with the diastolic BP higher than 90 mmHg.
- Elevated BUN or creatinine: Elevation of BUN >40 mg/dl or creatinine >1.5 mg/dl, not due to dehydration or obstruction.
- Hepatitis B virus: Presence of hepatitis B surface antigen or antibody in serum.
- Arteriographic abnormality: Arteriogram showing aneurysms or occlusions of the visceral arteries, not due to arteriosclerosis, fibromuscular dysplasia or other non-inflammatory causes.
- Biopsy of small or medium-sized artery containing polymorphonuclear cells: Histologic changes showing the presence of granulocytes or granulocytes and mononuclear leukocytes in the artery wall.

*These criteria have a reported sensitivity of 82.2% and a reported specificity of 86.6% for the classification of polyarteritis nodosa compared with other vasculitides.

Source: Lightfoot RW, et al. "The American College of Rheumatology 1990 criteria for the classification of polyarteritis nodosa." Arthritis and Rheumatism. 33:1088-1093. 1990.

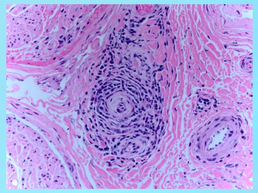


Image A: High powered vessel biopsy demonstrating transmural inflammatory cell infiltrates consistent with vasculitis.

Source: Case report patient

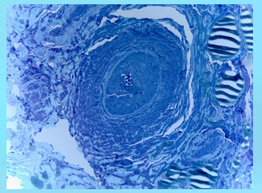


Image B: Thick section thrombosed vessel with surrounding inflammation consistent with vasculitis.

Source: Case report patient

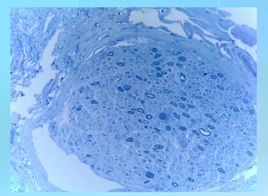


Image C: Thick section nerve fascicle demonstrating selective depopulation of myelin fibers.

Source: Case report patient

Frequency of Hepatitis B Virus-Associated Polyarteritis Nodosa Cases Among Polyarteritis Nodosa Cases According to Time of Occurrence

5-year Period	# of HBV-PAN Cases/ Total # of PAN Cases	%
1972-1976	5/13	38.5
1977-1981	13/25	52.0
1982-1986	41/84	48.8
1987-1991	21/77	27.3
1992-1996	20/56	35.7
1997-2002*	15/86	17.4

* This final period lasted for 6 years

Source: Guillevin, L. et al. "Hepatitis B Virus-Associated Polyarteritis Nodosa: Clinical Characteristics, Outcome, and Impact of Treatment in 115 Patients." Medicine. 84(5):313-322, September 2005.

Discussion:

The presentation of mononeuritis multiplex can mislead one into believing that the patient is suffering from a central neurological deficit. However, the patient's clinical presentation must be taken into account and prompt the physician to rule out other possibilities. In this case, a viral infection likely triggered the autoimmune disorder. Hepatitis B-associated PAN has been documented but is on the decline secondary to hepatitis vaccinations. It is believed that the viral antigen and antibody form an immune complex that is deposited in the vessel wall, leading to inflammation. In hepatitis B-associated PAN, plasmapheresis has been demonstrated to be effective in removing circulating immune complexes and may lead to hepatitis B seroconversion, thus leading to a more favorable outcome. Our patient's viral load was undetectable after weeks of antiviral therapy and correlated with his clinical improvement, proving that the combination of corticosteroids, plasmapheresis and antiviral therapy are effective in treatment of hepatitis B-associated PAN.

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