Progressive Ascending Paralysis in an Adult Male diagnosed with POEMS Syndrome: A Case Report

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ABSTRACT

Progressive ascending symmetrical weakness is generally seen in patients with acute inflammatory demyelinating polyradiculoneuropathy. We report a case of a healthy 56-year-old male who presented with ascending bilateral lower limb weakness with severe painful paresthesias occurring for the past four weeks. On physical examination, the patient presented with symmetrical flaccid paralysis of the lower limbs, areflexia, proprioception impairment, and decreased sensation to light touch and pinprick. The patient also had hepatomegaly and bilateral lower extremity edema. Brain and spine MRI showed no acute pathology; however, MRI abdomen and pelvis revealed hepatomegaly and osteosclerotic lesions at the right ilium and acetabulum. Pertinent test results showed high TSH levels, low testosterone levels, high IgG levels, and M-protein spike on protein electrophoresis. In addition, electrodiagnostic studies demonstrated severe axonal polyneuropathy with demyelinating features at the lower extremities, whereas needle electromyography showed distal fibrillation potentials. The bone marrow and pelvic tumor biopsies revealed monoclonal plasma cells (plasmacytoma), and the patient was diagnosed with POEMS syndrome. With chemotherapy and radiation therapy to the right pelvis in concurrence with intense rehabilitation and pain management, the patient's neuropathic pain and sensorimotor function improved in the inpatient rehabilitation. POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes) is a rare multisystemic disease that occurs in the setting of a plasma cell disorder. Polyneuropathy is the most common clinical manifestation and a requirement for the diagnosis, where patients usually have ascending symmetrical motor and sensory deficiencies in the limbs and extremities. Although an uncommon cause of polyneuropathy, this case highlights known physical examination and laboratory findings associated with this syndrome, and therefore clinicians should be aware of POEMS syndrome and to include it in the differential diagnosis for ascending paralysis.

CASE DESCRIPTION

A healthy 56-year-old white male presented with feeling "weak all over" for past four weeks. Initially, the patient noticed foot dropping, and then felt weakness spreading over his legs to his thighs, leading to difficulty walking. His weakness in his legs has been associated with numbness in his feet and severe paresthesias. The patient noted mild abdominal pain and denies urinary or bowel incontinence. On physical examination, the patient presented with symmetrical flaccid paralysis of the lower limbs, areflexia, proprioception impairment, and decreased sensation to light touch and pinprick. The patient also had hepatomegaly and bilateral lower extremity edema. *Laboratory Results*: The lumbar puncture CSF, heavy metal, porphyria, and connective tissue screening were all normal. However, laboratory results did showed high TSH levels, low testosterone levels, high IgG levels, and M-protein spike on protein electrophoresis. *Imaging*: Magnetic resonance imaging (MRI) in the brain and spine was normal; however, the MRI revealed hepatomegaly and osteosclerotic lesions at the right ilium and acetabulum. Subsequently, the patient underwent a pelvic and bone marrow biopsy, where the pathology showed plasmacytoma. *Electrodiagnostic Study*: The study demonstrated severe axonal polyneuropathy with demyelinating features at the lower extremities, whereas needle electromyography showed distal fibrillation potentials. *Diagnosis*: From the clinical, laboratory and pathological findings, the patient was diagnosed with POEMS syndrome. *Rehabilitation and Treatment*: The patient was transferred to acute inpatient rehabilitation. With chemotherapy and radiation therapy to the right pelvis in concurrence with intense rehabilitation and pain management, the patient's neuropathic pain and sensorimotor function improved.

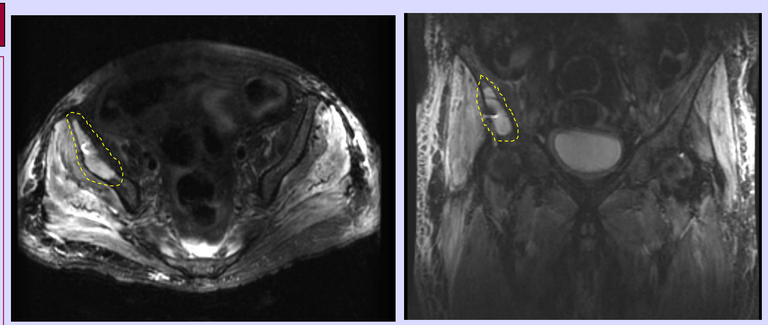

Figure 1. Magnetic resonance imaging of the pelvis reveals a destructive lesion measuring 7 x 5 x 2 cm in the right ilium above the right acetabulum

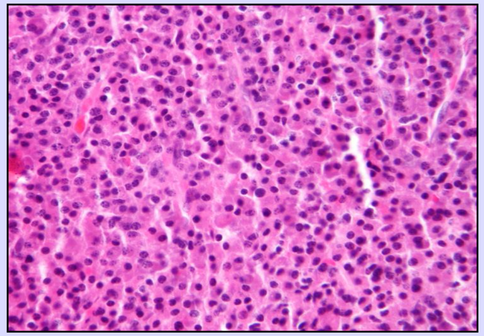

Figure 2. Biopsy of the right pelvic lesion demonstrates monoclonal plasma cell infiltration.

Major Criteria
Polyneuropathy
Monoclonal plasma cell-proliferative disorder
Minor Criteria
Organomegaly (splenomegaly, hepatomegaly, or lymphadenopathy)
Endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, pancreatic)
Skin changes (hyperpigmentation, hypertrichosis, hemangiomata, plethora, white nails)
Extravascular volume overload (edema, pleural effusion, or ascites)
Sclerotic bone lesions
Castleman disease
Papilledema
Thrombocytosis/polycythemia

Table 1. POEMS Syndrome diagnostic criteria. Diagnosis requires the two major criteria plus at least one minor criterion.

DISCUSSION

POEMS syndrome, also known as the Crow-Fukase syndrome or osteosclerotic myeloma, is a rare multisystemic disease that occurs in the setting of a plasma cell disorder with characterization of polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes. The pathogenesis of this syndrome is not clear. The suggested pathogenesis is minute changes in the vessel wall leading to increased vascular permeability, which this is probably mediated by cytokines, such as VEGF. Due to the uncertainty of the pathogenesis, treatment strategies and prognosis are heterogeneous.

Polyneuropathy is the most common symptom and is required for the diagnosis of POEMS syndrome. Most patients with this symptom have symmetrical motor and sensory deficiency in the extremities. It usually starts in the lower extremities with progressive proximal extension. Decreased deep tendon reflexes associated with various combinations of sensory symptoms are found. EMG shows signs of both demyelination and axonal degeneration. Nerve conduction abnormalities exhibit characteristic, patterns that can be summarized by a number of features including: (1) slow nerve conduction diffusely distributed in the intermediate nerve segment, (2) relatively preserved nerve conduction near the distal nerve terminals, (3) prominent axonal loss in distal lower extremity nerves, and (4) no conduction blocks (5) lower limbs having absent or attenuated amplitudes of compound muscle action potentials and absent sensory nerve action potentials compared with upper limbs. Distal fibrillation potentials are found on needle electromyography. These features are useful in differential diagnosis of POEMS syndrome from chronic inflammatory demyelinating polyneuropathy (CIDP), where CIDP has the presence multifocal conduction blocks.

Plain film radiographs are useful for locating lytic bone lesions caused by osteosclerotic myeloma. At least 95% of patients have osteosclerotic lesions, with more than half the patients having multiple lesions. Both osteosclerotic and osteolytic lesions may be present and may be of modest size.

Radiation therapy to the osteosclerotic lesions has been beneficial, where systemic and skin symptoms and even polyneuropathy improve. In addition, chemotherapy (melphalan and prednisone) is administered in patients with widespread osteosclerotic lesions. Often times, concurrent treatment is applied to POEMS patients.

CONCLUSION

We present an uncommon cause of polyneuropathy and ascending paralysis in a patient with POEMS syndrome, which most often overlooked. It is important that clinicians recognize the distinct signs, symptoms, electrodiagnostic, laboratory and imaging studies of POEMS syndrome in order to permit early and proper diagnosis, rehabilitation, and pharmacologic treatment.

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