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Case Report

POEMS Syndrome - A Case Report

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Abstract

The acronym "POEMS syndrome", also known as Crow-Fukase disease, stands for Polyneuropathy, Organomegaly, Endocrinopathy, M-Protein, and Skin changes. It is a rare multisystem disorder, which may take several years to evolve fully, and patients suffer undiagnosed for long [1]. The combination of symptoms and signs is highly complex and some of the features are detected at sub-clinical level requiring high level of suspicion [2]. The clinical data on POEMS is still evolving with only a few case reports from India.

We herein report case of a 38-year-old male with peripheral neuropathy and ascites, which on later work up was diagnosed as POEMS syndrome in accordance with its diagnostic criteria [3-5]. Noteworthy, that the patient later also developed pulmonary Koch's. The case report is meant to enlighten the readers regarding the clinical spectrum of this uncommon entity and encourage appropriate investigations to confirm the diagnosis in scenarios of multisystem affection.

Keywords: POEMS Syndrome; Ascites; Plasma

Case Report

A 38-year-old male was admitted with progressive abdominal distension suggestive of ascites. The patient had been well until approximately 4 months back, when paresthesias of both feet, weakness of the lower legs, and bilateral foot drop, and abdominal distention all of which developed gradually. The patient had a history of slipping of slippers from feet, unable to feel the ground, painless trauma to feet.

General examination revealed mild pallor, no icterus, cyanosis, clubbing, lymphadenopathy. There was a bipedal pitting edema of the legs to the waist.

Vitals were stable with pulse rate of 70 beats per minute, and supine blood pressure of 122/76 mm Hg. Temperature was 36.3°C, respiratory rate 20 breaths per minute, and oxygen saturation 99% on ambient air. Positive findings on neurological examination were.

The left hand was mildly weak, particularly the abductor pollicis brevis, and strength was normal in the proximal arms. He had bilateral claw hands and decreased strength in the arms and legs. Strength in the ankle dorsiflexors measured 1 to 2; in the muscles of ankle eversion and inversion, 4−; in the extensor hallucis longus, extensor digitorum brevis, and toe flexors, 0; and in the plantar flexors, 4− to 4. Deep tendon reflexes were absent. Perception of vibration was absent in both first toes, decreased in both ankles, and normal at the knees and fingers. Position sense was diminished in both first toes. Romberg test was markedly abnormal; there was bilateral foot drop, Patient was unable to walk on his toes or heels.

Other systems were examined to reveal crackles at the lung bases. Bowel sounds were present, and the abdomen was tense, distended, and nontender. The remainder of the neurologic examination was normal.

Abdominal paracentesis was performed, 2 liters of ascitic fluid were removed, and abdominal discomfort decreased.

Radiological investigations like ultrasonography of the abdomen revealed increased echogenicity of the liver, splenomegaly (span, 16.6 cm), and ascites; the portal and hepatic vasculature was patent. Another striking observation was an generalised increased bone density, shiny bone shadows indicative of sclerosis in routine chest xrays, and also in vertebrae seen on thoracolumbar x-ray seen for lower limb weakness. There was no history of pathological fractures in the patient. Also no reports of Flourosis were present in his society. Calcium, phosphorus and PTH were all reported normal.

Thus, we had a middle aged male with progressive peripheral neuropathy and sclerotic bones, accompanied by hyperpigmentation of arms and feet, erectile dysfunction, pedal edema and ascites.

Tests of liver and renal function and measurements of serum electrolytes, thyroid hormones, glycosylated haemoglobin, vitamin B12, homocysteine, and methylmalonic acid were normal. Tests for rheumatoid factor; ANA; and GM1 antibodies were negative too.

Nerve conduction studies showed decreased sensory responses, absent suralnerve responses, prolonged F-wave latencies, and more prominent denervation in the tibialis anterior and medial gastrocnemius muscles, features consistent with a demyelinating polyneuropathy [6].

Computed tomography of the chest, abdomen, and pelvis after the administration of contrast material revealed scattered nodules (2-3 mm in diameter) in both lungs, diffuse lymphadenopathy (including a retroperitoneal paraaortic mass, 2.2 cm by 2.1 cm), ascites, a small pericardial effusion, and B/L pleural effusions.

Urinalysis revealed a specific gravity of more than 1.040, 1+ albumin, a total protein level of 320 mg per litre (reference range, 0 to 135), and a ratio of total protein to creatinine of 0.18; it was otherwise normal.

With further work up, clinical picture was fitting in various criteria of POEMS syndrome.

With clinical suspicion, serum electrophoresis was done which disclosed a monoclonal IgG spike. A low level of monoclonal IgG with lambda light chain is also characteristic of the POEMS syndrome [7]. However, in contrast to multiple myeloma, the levels of other immunoglobulin subclasses are not reduced in the POEMS syndrome. Overproduction of plasma cells in individuals with POEMS syndromes may result in the formation of sclerotic bone lesions and an overabundance of M-proteins in the blood [8]. In most cases, the specific type of plasma cell dyscrasia associated with POEMS syndrome is osteosclerotic myeloma, a variant of multiple myeloma [9].

Figure 1: Diagnostic criteria for the POEMS syndrome.

The patient was referred to an oncology centre for Radiation/ Stem cell therapy.

Conclusion

Henceforth, we understood that multisystem disorders require prudent testing and analysis to reach the underlying diagnosis. POEMS syndrome is one of such disorders, and requires high degree of suspicion, and polyneuropathy gives a way to proceed in nearly all cases. Osteosclerosis, endocrine abnormalities and organomegaly are usually encountered in routine investigations but ignored initially by treating physicians. Moreover, serum protein electrophoresis is recommended in cryptogenic neuropathies, especially with radiological evidence of bone mineralisation defects.

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