ORIGINAL RESEARCH PAPER

Medical Science

POEMS SYNDROME- A REHABILITATION CHALLENGE

KEY WORDS: .

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Introduction

POEMS Synchrome also known as crow-fukase syndrome was first reported in 1956.It is characterized by association of polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy ,skin changes.POEMS syndrome is "a rare variety of plasma cell dyscrasia"(including gynecomastia,impotence, amenorrhea,diabetes mellitus and hypothyroidism),elevated M protein level, and skin changes(such as hyperpigmentation, hypertrichosis and thickening).Other major findings include papilledema,peripheral edema, ascites and clubbing)

Background

POEMS is an rare but unique multisystem disorder characterized by polyneuropathy(predominantly sensorimotor),organomegaly (hepatosplenomegaly,lymphadenopathy),endocrinopathy.Affect ed patients may not exhibit all these features, and sometimes presentations are atypical. Most patients have been of Japanese descent.Very few reports of this unique presentation has come across and literature from India that compels us to bring this case in notice.

Case report

A 34-year old man was admitted to D.P.M.R ,K.G.M.U,Lucknow with complain with ascending progressive peripheral neuropathy in all four limbs of 12 years duration and multiple contractures in all four limbs of 5 years duration . He noted no change in urinary or bowel habits, but he had anorexia with about 15 kg weight loss in last five years.He had no fever or night sweat and no cardiac,respiratory or gastrointestinal symptoms. He was followed intermittently by physician for hypertention.He had a history of cerebrovascular accident in the form of intracranial haemorrhage 3 years back.He had no high risk behavior for HIV infection.

Physical examination revealed a distal and limb girdle weakness of both upper and lower limbs with bilateral foot drop and multiple contractures in bilateral wrist and finger joints, and at ankle joint, proprioception and vibration were impaired,hyperaesthesia at ankle and soles bilaterally,hepatosplenomegaly,bilateral axillary lymphadenopathy, and bilateral papilledema, wasting of muscles. Thickness of skin at bilateral soles.Gynaecomastia was present,a non tender firm mobile round mass (3*3 cm)was palpable in abdomen near umbilicus.Mild ascites was present, bilateral pitting pedal edema was present, touch and pain was impaired in both upper and lower limbs, cranial nerves examination was normal except optic nerve where diminished vision was seen.Bladder and bowel was not involved.Knee and ankle jerks were absent with absent plantar and hypotonia bilateral upper and lower limbs,pallor present.Clubbing present bilaterally.Organomegaly present, skin dry, hyperpigmented, shiny wasting of muscles in all four limbs,more severe distally,flexion contractures in both upper and lower limbs and severe compromised activities of daily living and virtually bed ridden.

Investigations

Hb and white blood cell level were normal,platelets counts were elevated.GBP,MCH, MCHC, MCV, HCT, retic count were normal.ESR was raised. Urine examination shows monoclonal light chains.His renal functions were normal,SGOT,SGPT were normal, total proteins 6.2gm % albumin 3.5gm/dl and lactate dehydrogenase 1200 u/l. CK-23, CK-MB-03, CKMM-20.Serum sodium and potassium were normal,serum calcium was low,serum alkaline phosphatase normal.Serum glucose (random) was normal.Bilirubin level normal.HIV HBsAg,HCV negative.Serum TSH level raised,testosterone level low.

Chest radiograph and Magnetic resonance imaging (MRI) of the spine are demonstrated no significant finding.Ultrasonography showed hepatosplenomegaly.

Computed tomography(CT) of the chest showed generalized lymphadenopathy,a moderate left pleural effusion,hepato splenomegaly,the testosterone level (1.67 ng /mL). Electromyography (ENG) was consistent with a demyelinating polyneuropathy,elevated thyroid stimulating hormone level.A myelogram obtained elsewhere was reported to be normal.A cranial CT scan was normal.Biopsy of the skin , muscles,and lymph nodes failed to estabilish a uniform diagnosis.

On admission, white blood cell count was 13,400/mm3.Platelet count was 6,25,000 / mm3.Proteingria and Bence-Jones protein were not noted.Serum protein was 8.2g(albumin-4.5g and globulin-3.7g) and IgG was 3095mg/dl (800-1700).A CSF examination disclosed 36.5mg/dl of IgG and 128mg/dl of protein but no cells. Serum and CSF protein electrophoresis and immunoelectrophoresis showed a monoclonal spike of IgG-lambda. Serum testosterone was 2.84 ng/ml (0.15-1.1) but ACTH,cortisol, growth hormone, prolactin , LH and FSH were normal.Liver scan showed hepatosplenomagaly without dysfunction of reticuloendothelial cells.MR showed discrete low signal intensity lesions in T1 and T2 weighted images indicating osteosclerotic myeloma.Examination revealed bone marrow was normal with less than 2% plasma cells.Nerve conduction studies revealed a marked reduction in amplitude and velocity of the median and ulnar nerves.But an electrical response could not be obtained in the lower extremities.A sural nerve biopsy specimen showed scattered myelin digestion chambers and mild fibrosis in perineurial and endoneurial tissue.Endoneurial mononuclear cell infiltration with focal vasculitis was also noted.Biopsies of two cutaneous nodules disclosed endothelial proliferation compatible with cherry hemangioma .

Nerve conduction studies revealed evidence of a demyelinating polyneuropathy with marked conduction slowing in peroneal,median and ulnar nerve bilaterally.Sural responses were absent and tibial nerves inexcitable.The cerebrospinal fluid was acellular with a raised protein level of 1 giL (reference range-0.1-0.4).He underwent endoscopic evaluation of this gasto-intestinal tract.There was macroscopic evidence of mild colitis affecting the rectosigmoid and descending colon,and histological examination of the affected tissue showed moderately extensive lymphoplasmacytic infiltrate of the lamina propria.There were bilateral pleural effusions and ascites on computed.

Tomography (CT) but no hepatosplenomagaly or evidence of intra-abdominal malignancy.Full blood count,renal function and calcium level were normal. MRI of thoracic and lumbpsacral spine showed that Hormonal profile testing revealed decreased testosterone level of 2.46 nmol/L (reference range -9.9-27.80) with a normal study of the pituitary axis.In view of the constellation of a demyelinating polyneuropathy,osteosclerotic plasmacytoma,extra-vascular volume overload and endocrinopathy,the patient was diagnosed with POEMS Syndrome.

Prior to this diagnosis,the patient had been diagnosed with chronic inflammatory demyelinating polyneuropathy,He had been

treated with a course oral steroid therapy.There had been a subjective initial partial response before his symptoms progressed. Following the diagnosis of POEMS,a course of radiotherapy to the bone lesions was completed . There was no objective improvement and he progressed to develop severe global weakness in his upper and lower limbs.Passive range of motion (PROM) exercise of all involved extremities.Orthosis and splint for prevention of joint contractures.

Differential Diagnosis

Castleman's Syndrome,GBS,Hansen's multiple myloma,Vasculitic neuropathy,Paraprotein associated neuropathy.

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