Rib Plasmacytoma Presenting as Progressive Muscle Weakness

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Abstract

We report the case of a young male who gradually developed a subacute demyelinating polyradiculoneuropathy. Routine examination and extensive biochemical investigations revealed demyelinating neuropathy, M-Band in electrophoresis. The patient referred for the whole-body ^{18}F-*fluorodeoxyglucose*-positron emission tomography/computed tomography to look for skeletal and bone marrow lesions. The patient was found to have rib plasmacytoma. The case is rare because of infrequent association with the site, age, and symptoms.

Keywords: *Demyelinating neuropathy, fluorodeoxyglucose positron emission tomography/computed tomography, paraneoplastic syndrome, plasmacytoma, polyneuropathy, organomegaly, endocrinopathy, M-protein, skin changes syndrome*

Introduction

Solitary plasmacytoma is a type of plasma cell dyscrasia that presents as a single mass of monoclonal plasma cells that may be located either extramedullary or intraosseously. It mainly occurs primarily in vertebrae, femurs, pelvis, and ribs, but its extraosseous variant can involve any site or organ, with the most common being the head and neck region, gastrointestinal tract, and lungs.

The occurrence of a solitary plasmacytoma along with peripheral neuropathy is a rare finding. The neuropathy presents quite early in the natural history of this malignancy before the patient becomes symptomatic for myeloma. Such findings can also occur in the polyneuropathy, organomegaly, endocrinopathy, M-protein, skin changes (POEMS) syndrome. If not suspected, the diagnosis may be entirely missed, as myeloma can be detected only by performing certain specific investigations.

Current treatment strategies all target the underlying plasma cell clone, with the exception of bevacizumab, a monoclonal antibody targeting vascular endothelial growth factor. Management can be complicated, and can be fatal. With the right treatment, however, prognosis in many patients can be very good.

Case Report

A 43-year-old male patient presented with complaints of gradual onset progressive difficulty in getting up from squatting posture for 3 months, difficulty in lifting heavy objects above head, clumsiness while buttoning his shirt, imbalance while walking, and tingling along with numbness in his feet for 15 days. There was a history of ~5% weight loss in the last 3 months. He is a known diabetic, hypertensive, and chronic alcoholic.

On examination, his higher mental functions were intact, and cranial nerves examination unremarkable. Motor system examination revealed normal bulk, tone, and power in all limbs. There was a weakness in the dorsiflexion of the ankle. Deep-tendon reflexes were absent in all four limbs. Superficial reflexes were normal. The sensory system examination was unremarkable. Patients had positive signs of cerebellar involvement. Nerve conduction velocity showed decreased amplitude in bilateral common peroneal nerve with unrecordable sural sensory nerve action potentials in B/L sural nerves. However, there was no conduction block in any of the four limbs - S/O demyelinating neuropathy. The skeletal survey was unremarkable. Bone marrow aspirate showed 10% of plasma cells. Serum electrophoresis with absolute quantification revealed M-band in the gamma region (quantity 0.73 g/dl).

How to cite this article: Kalimuthu LM, Singh P, Ora M, Gambhir S. Rib plasmacytoma presenting as progressive muscle weakness. Indian J Nucl Med 2021;36:66-8.

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Received: 15-06-2020
Revised: 27-07-2020
Accepted: 03-08-2020
Published: 04-03-2021

Access this article online

Website: www.ijnm.in

DOI: 10.4103/ijnm.IJNM_133_20

Quick Response Code:

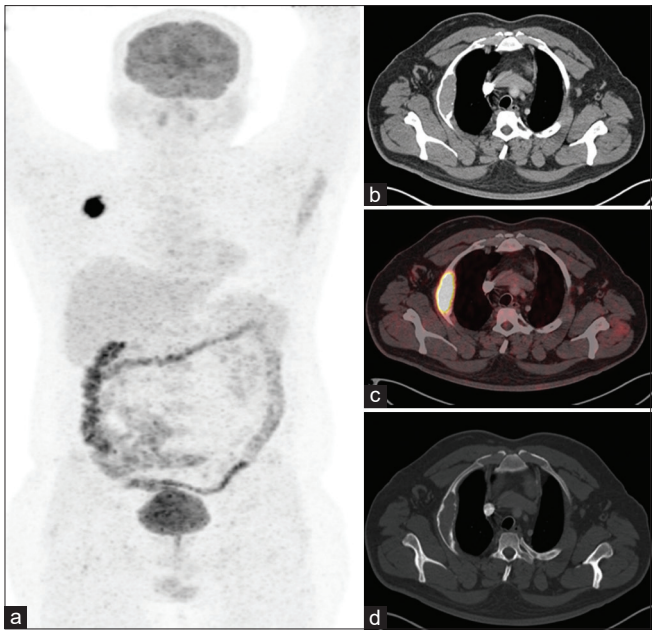

Figure 1: (a) Maximum intensity projection of 18F-fluorodeoxyglucose positron emission tomography/computed tomography, (b) Lesion in computed tomography, (c) Lesion in positron emission tomography/computed tomography and (d), Lesion in computed tomography (bone window)

The antibody panel analysis was negative. Bone marrow biopsy revealed increased vascularity (~70%) with trilineage hematopoiesis. Serum immunofixation showed IgG (lambda) type.

Fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET-CT) was performed, showed an FDG avid (SUVmax 20.9) lytic, expansile lesion with soft-tissue component in the right 2nd rib anterolaterally [Figure 1]. No apparent extra or intrathoracic soft-tissue component is noted. CT-guided biopsy from the lytic lesion was performed and suggestive of plasmacytoma. The patient was started on RVD (lenalidomide, bortezomib, and dexamethasone) regimen. The patient has significant improvement in the symptoms in follow up.

Discussion

Solitary plasmacytomas of the bone are defined as clonal proliferations of plasma cells which manifests as a localized osseous growth. Plasmacytomas can be divided into multiple, solitary osseous, and solitary extraosseous or extramedullary plasmacytomas and rare as compared with multiple myeloma.[1] Solitary plasmacytoma of the rib is rare and commonly involves fourth and fifth ribs. Most of the patients present with the mass or local pain. Rib solitary plasmacytoma presenting with the peripheral neuropathy is extremely rare, and only few cases have been reported.[2] Solitary bone plasmacytoma has an indolent course of the disease, with a median survival time of 10.7 years and 5-, 10-, and 20-year survival rates of 75, 52, and 37%, respectively.[3]

In these patients, neuropathy manifests earlier than other clinical manifestations of the malignancy, diagnosis rests on detection of a monoclonal protein or solitary bone lesion. Therefore immuno-electrophoresis and detailed radiological survey should be done in all patients with predominantly motor neuropathy of undetermined aetiology. Solitary plasmacytoma tends to involve the axial skeleton i.e., ribs, scapula, vertebrae and pelvis and spares the skull and long bones.[4]

Such presentation can also occur in POEMS syndrome. POEMS syndrome, also known as Takatsuki syndrome or osteosclerotic myeloma, is a rare paraneoplastic syndrome arising from an underlying plasma cell disorder. Most commonly, it presents in the fifth to sixth decade.[5] The diagnosis of the syndrome is based on the criteria by the International Myeloma Working Group, requiring a combination of clinical and diagnostic tests.[6] Only 88% of the patients have a monoclonal protein in the serum and/or urine in a very minimal amount. Serum protein electrophoresis is normal in one-fourth of the patients, and some patients can have a pattern suggesting the presence of a polyclonal gammopathy. Bone marrow examination is often nondiagnostic. Less than 15% of the patients have a bone marrow plasmacytosis (>10%).[7] Clinical features involve neuropathy, thrombocytosis or erythrocytosis, organomegaly, hypogonadism hyperpigmentation, hypertrichosis, or lipodystrophy.[8] Around 55%–60% of the patient present with sclerotic bone lesions and lytic lesions being rare.[9]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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