

## Case Report

# A CASE REPORT OF POEMS SYNDROME WITH OSTEOLYTIC LESIONS

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## ABSTRACT

POEMS syndrome is a paraneoplastic multisystem disorder associated with underlying plasma cell neoplasm. It includes polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell proliferative disorder, skin changes and several other clinical features. Polyneuropathy is a dominant clinical feature and there is a definitive criterion for its diagnosis.

**Keywords:** Poems Syndrome

## INTRODUCTION

POEMS syndrome is a multisystem disorder. POEMS syndrome is an acronym which includes polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell proliferative disorder, skin changes. And several other features are also included in the syndrome, which are not in the acronym. Polyneuropathy is the dominant clinical feature in this syndrome. Here we present a case of POEMS syndrome with osteolytic lesions.

## CASES

A 34 yrs old female patient presented with complaints of tingling, numbness of both upper & lower limbs since 6 months, weakness of both upper & lower limbs, shortness of breath, lowback ache since 4 months, pedal edema since 2 months and abdominal discomfort since 1week. The illness is of insidious onset, continuous course & deteriorating progression.

No significant past and family history. General examination revealed pedal edema, hyperpigmentation of skin. Cardio vascular examination is normal. On respiratory system examination breath sounds are decreased in bilateral infra axillary & infra scapular regions. Per abdomen examination shows distended abdomen, shifting dullness, hepatosplenomegaly.

CNS examination performed which revealed decreased power, bulk of muscles, deep tendon reflexes and on sensory examination light touch, vibration, joint sense, pain & temperature sense are also decreased in both upper & lower limbs, but more in lower limbs. Routine lab investigations are unremarkable except for elevated TSH.

Chest radiograph shows cardiomegaly (likely pericardial effusion), bilateral pleural effusions which are confirmed on CT chest. USG of abdomen revealed mild hepatosplenomegaly, mild ascites, bilateral moderate pleural effusions. Radiograph of pelvis showed well defined lytic lesions in left ischium and pubis which is further evaluated with CT scan revealed well defined lytic lesions in left ischial bone, acetabulum and pubis.

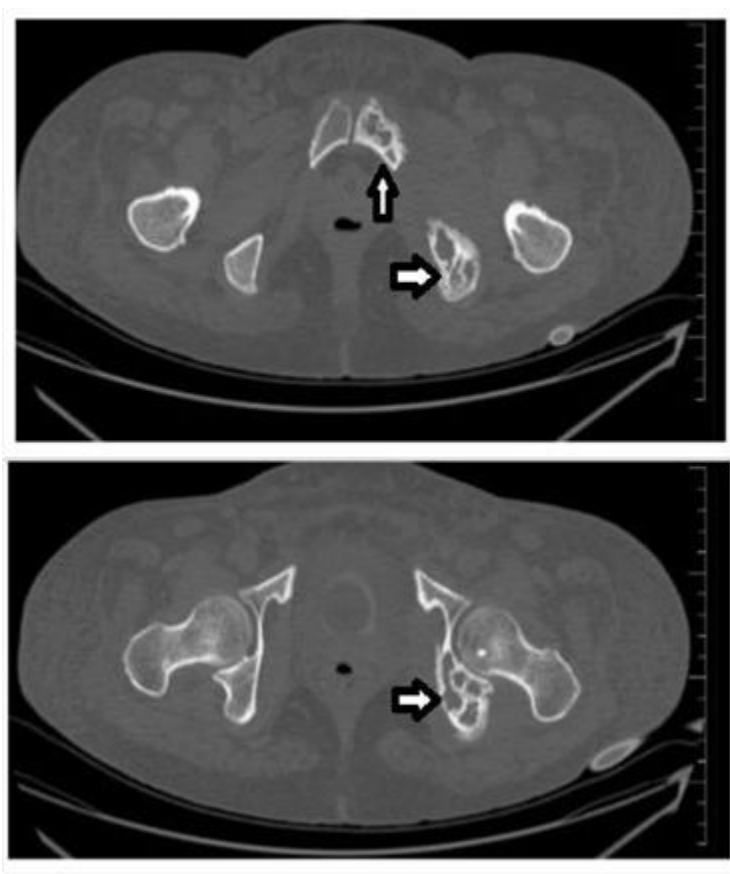
CT guided biopsy of the lesion is performed and histopathological sections of the lesion show round to plasmacytoid cells in sheets. On immunohistochemistry, the tumor cells diffusely express CD138. Expression of lambda light chains is more diffuse and stronger than kappa light chains. Bone marrow biopsy shows Cellular reactive marrow with plasma cells and with CD138 positivity suggesting plasma cell neoplasm.

Nerve conduction studies of both upper & lower limbs shows diffuse symmetrical sensory motor neuropathy more in lower limbs. Considering peripheral neuropathy, monoclonal plasma cell proliferation, hepatosplenomegaly, hyperpigmentation and osteolytic bone lesions diagnosis of POEMS syndrome is made.

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**Figure 1: Radiograph Pelvis Shows Lytic Lesions Noted in Left Ischium and Left Pubis Bones**

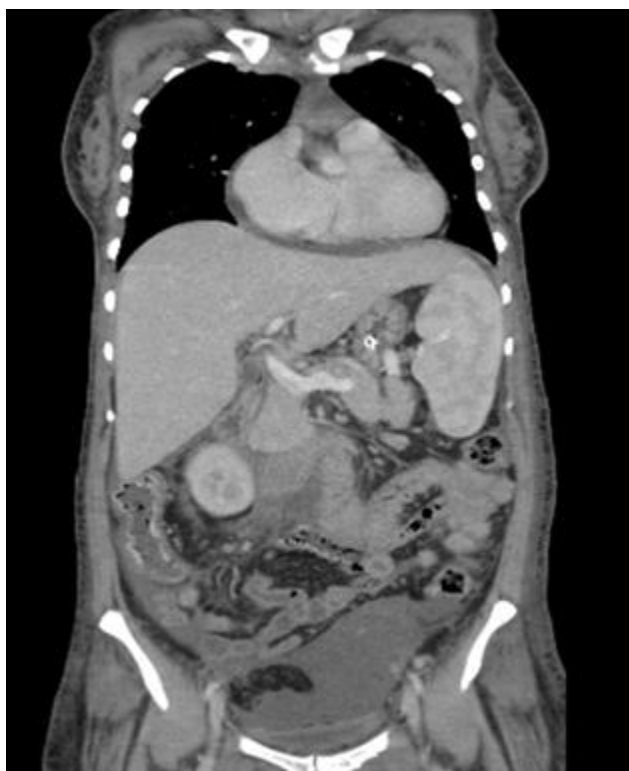


**Figure 2: Axial CT Sections Shows Well Defined Lytic Lesions in Left Ischium and Left Pubis Bones**

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**Figure 3: Axial Sections of CT Chest Shows \* Bilateral Pleural Effusions and → Pericardial Effusion**



**Figure 4: Coronal Sections of CT Abdomen Shows Free Fluid in Peritoneal Cavity (Ascites)**

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### **DISCUSSION**

POEMS syndrome consists of polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes. Many other important features which are not included in the acronym are sclerotic bone lesions, castleman disease, papilledema, pleural effusion, edema, ascites and thrombocytosis. The main pathogenesis involved in POEMS syndrome is imbalance between proinflammatory cytokines. The main pathogenic factor involved in POEMS is vascular endothelial growth factor (VEGF).

POEMS is a paraneoplastic syndrome with underlying plasma cell neoplasm. The major criteria for diagnosis of POEMS syndrome includes polyneuropathy, monoclonal plasma cell proliferative disease, castleman disease, sclerotic bone lesions, VEGF elevation out of which first two lesions are mandatory. The minor criteria includes organomegaly (hepatomegaly, splenomegaly or lymphadenopathy), extra vascular volume overload (edema, pleural effusion, ascites), endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid and pancreatic), skin changes (hyperpigmentation, hypertrichosis, plethora, acrocyanosis), papilledema, thrombocytosis / polycythemia. And the other symptoms & signs include clubbing, weight loss, hyperhidrosis, pulmonary hypertension / restrictive lung disease, thrombotic diathesis, diarrhoea.

The diagnosis of POEMS is definitive when polyneuropathy, monoclonal plasma cell proliferation are present with another one major & minor criteria (Dispenzieri, 2014). The present case shows typical involvement of four systems at the time of presentation. There is no need of involvement of all the systems included in the acronym to make a diagnosis of POEMS syndrome.

POEMS syndrome usually present during 5<sup>th</sup> & 6<sup>th</sup> decades of life. Peripheral neuropathy is the dominant symptom in POEMS syndrome. Sensory neuropathy is affected initially followed by motor involvement. Bilateral symmetrical distal involvement with gradual proximal progression. Our case also presented in similar way. The disease is progressive, more than half of the patients become weak and unable to climb stairs, arise from chair. Cranial nerves are not involved except for papilledema. Hepatomegaly is most common followed by splenomegaly, lymphadenopathy. Histopathology of these lymphnodes shows angiofollicular lymphnode hyperplasia (castleman's disease). Endocrinopathy is the poorly understood feature of POEMS syndrome (Dispenzieri, 2007).

Hypogonadism, thyroid abnormalities, glucose metabolism abnormalities and adrenal insufficiency are some of the abnormalities. Multiple endocrinopathies is also present in majority of patients. Hyperpigmentation is the most common skin manifestation. Skin thickening, rapid accumulation of glomeroid angiomata, flushing and dependent rubor or acrocyanosis are the some other skin changes. Extra vascular volume overload (ascites, pleural effusion) occurs in 1/3<sup>rd</sup> of patients. Almost all patients have underlying monoclonal plasma proliferative disorder. 95% of bone lesions are osteosclerotic but some are osteolytic with sclerotic rim and mixed soapbubble appearance. CT scan is the best imaging tool to define these lesions (Dispenzieri, 2012).

Monoclonal plasma cell proliferation with osteolytic lesions in the left ischium and pubis as seen our case are consistent with imaging diagnosis of plasmacytoma. Management of POEMS syndrome includes radiation therapy for bone lesions, systemic corticosteroids and high dose chemotherapy with peripheral blood stem cell transplantation.

Diagnosis of POEMS syndrome is difficult but a good clinical history, physical examination & lab tests, radiographic assessment of bones, measurement of VEGF and bone marrow biopsy will help in diagnosis by differentiating POEMS syndrome from chronic inflammatory polyradiculoneuropathy, monoclonal gammopathy of undetermined significance, neuropathy immunoglobulin light chain amyloid neuropathy, castleman's variant of POEMS syndrome (Dispenzieri, 2012)

### **Conclusion**

POEMS syndrome is a rare paraneoplastic multi system disorder with underlying plasma cell neoplasm. The criteria for diagnosis includes osteosclerotic lesions. But if a patient present with osteolytic lesions as in the present case and if other features are in favour of POEMS syndrome, then clinician should consider POEMS syndrome as diagnostic possibility.

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