

# A Rare Case of Spinal Amyloidoma: Case Report and Review of Literature

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**Abstract:** Amyloidosis is accumulation of abnormal protein in tissue which can virtually affect any organ in the body. Large collection of amyloidosis in spine is called spine amyloidoma. We report a case of spine amyloidoma which cause bilateral lower limb neurological deficit and thoracic vertebral erosion. Investigations including imaging and tissue biopsy were helpful in establishing diagnosis. Localized resection and spinal stabilization surgery has good clinical outcome.

**Keywords:** spine, amyloidoma, amyloidosis, vertebral erosion

## 1. Introduction

Amyloidosis is a group of disease which is caused by abnormal accumulation of amyloid fibril in tissue or organ. The symptoms vary according to the types and the location deposition. To date, there are about 30 types of amyloidosis. However, it can be broadly divided into localized or systemic amyloidosis.

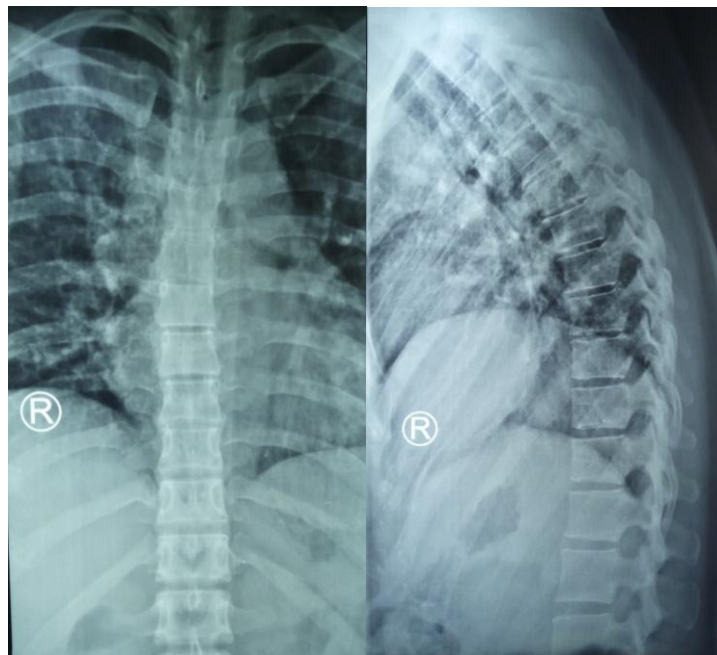
Large amyloid collections forming macroscopic lumps are called amyloidomas, these are rare and can cause mass effect.

## 2. Case Report

Mrs ZM 41 year old lady, with no known medical illness presented with sharp thoracic back pain which radiates to bilateral upper chest for 4 months. Her pain was only

relieved with analgesia and she developed gait instability and progressive weakness of bilateral legs 2 months after onset of pain. There were no associated bowel or urinary incontinence. She has no history of significant trauma, prolonged fever, family history of malignancy nor loss of weight or appetite prior to pain.

On examination, she has no back deformity but has minimal tenderness over midline midthoracic area. Her neurological examination showed normal tone but reduced power bilaterally from L2 to S1 (grade 4/5). Reflexes were brisk but no demonstrable clonus. Sensory below T8 was reduced. Anal tone and perianal sensation were intact. Other systemic examination including abdomen, pelvis, prevaginal, breast and neck were normal. Plain radiography of thoracolumbar showed bilateral T6 and left T7 pedicle erosion with paravertebral soft tissue swelling over T6-9.



**Figure 1:** Plain radiograph of thoracolumbar

MRI showed large enhancing soft tissue component within right paravertebral region with intraspinal extension through right T5/6 and T6/7 neural causing significant cord

compression at T5 and T6 level. CT Thorax, Abdomen and Pelvis showed lytic destruction of T6 vertebral with intraspinal extradural extension resulting in cord

Volume 8 Issue 8, August 2019

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compression with lytic changes to anterior body of T7 to T10 and metastatic changes to middle lobe of right lung, left adrenal and left paraaortic nodes. We worked her up for

tumor markers including CEA, Ca-125, Ca19-9, aFP but all were negative. Tuberculosis workout was also negative.

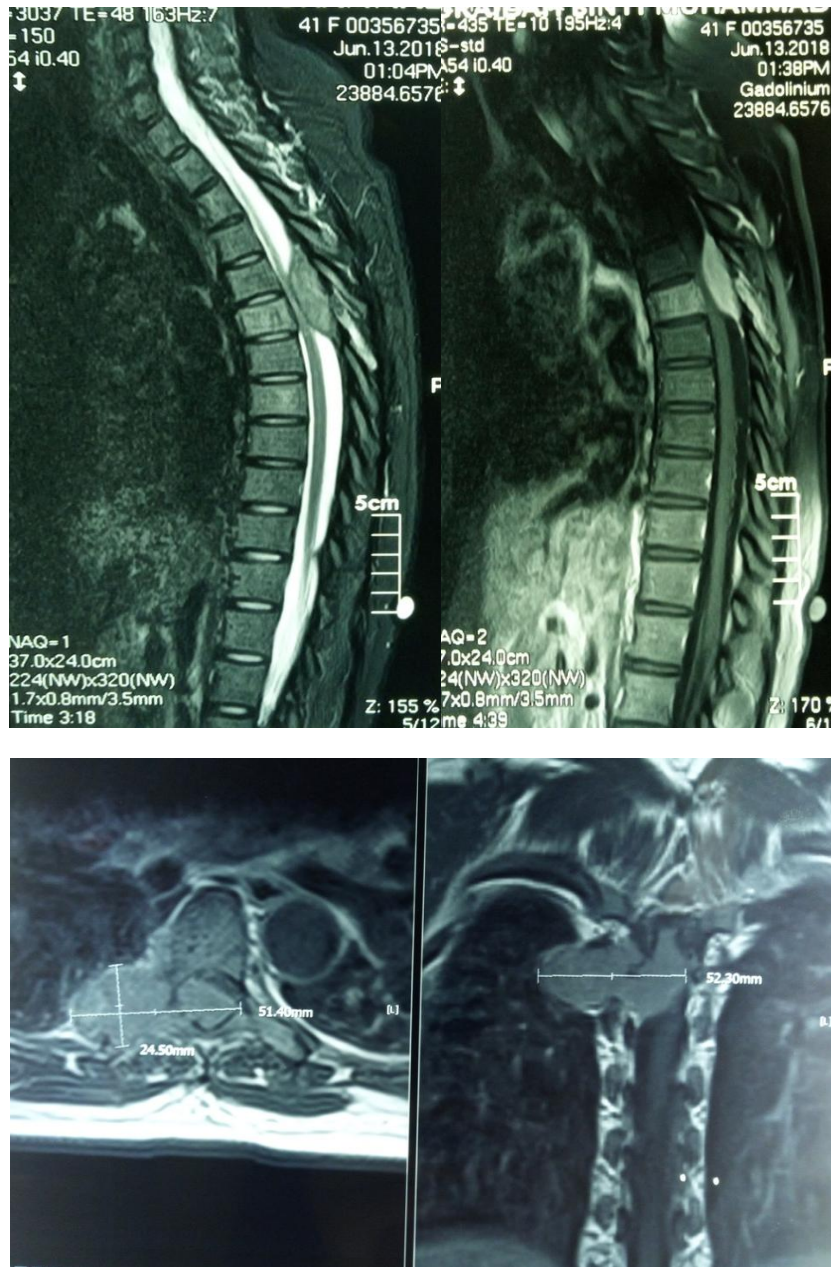
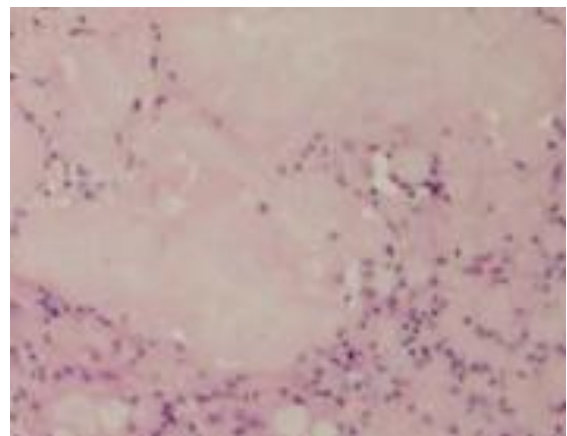
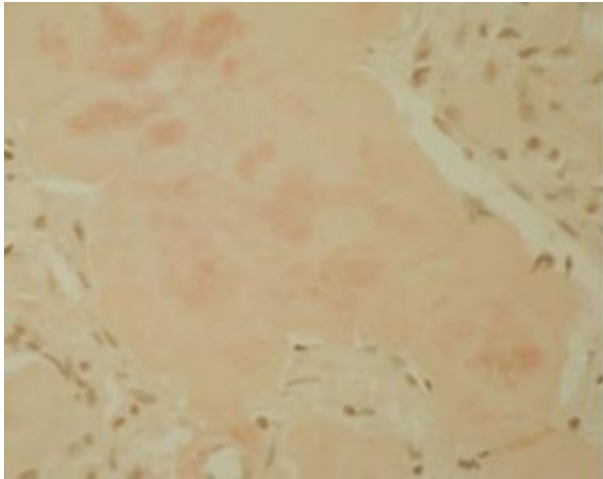


Figure 2: MRI of thoracolumbar showing the lesion in T6 and compressing on the cord

She was sent for CT guided biopsy of lesion. Histopathological examination showed pale, pink extracellular material within marrow spaces with surrounding lymphoplasmacytic cells. The deposits produce apple green birefringence under microscopy with Conge Red stain and positive with PAS stain.



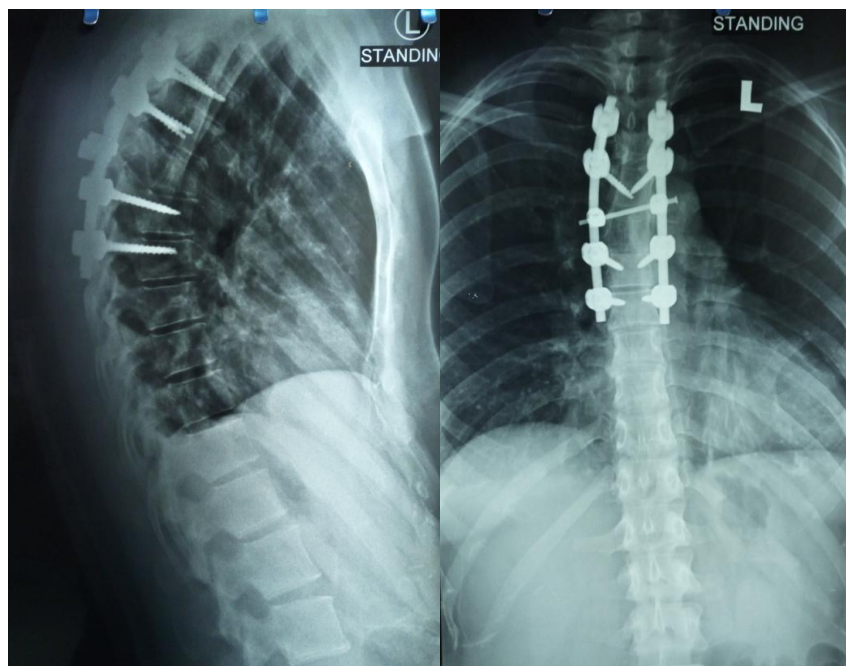
(a)



(b)

**Figure 3:** Histopathological specimen, a) normal polarized microscope. b) Congo Red Stain

Diagnosis of Spinal Amyloidoma was made and she underwent laminectomy T5-T6 with posterior spinal instrumentation and fusion of T4-T8. Intraoperatively there was yellowish gritty avascular mass located at right paravertebrae level T6-T9 with extension into spinal canal compressing the cord at T5 and T6. After performing laminectomy and excision of the m, neuromonitoring showed immediate improvement of signals.



**Figure 3:** Post operative plain radiograph of thoracolumbar

Three months post-operatively, her motor symptoms improved to grade 5, numbness symptoms improved and there is no more gait instability.

She was referred to hematologist for plasma cell dyscrasias screening and chemotherapy.

### 3. Discussion

Amyloidosis occurs as a result of misfolding of protein during synthesis, which were then clumped together because of presence of hydrophobic beta-pleated sheets. When exposed to water, the hydrophobic part tend to clump to other hydrophobic pieces, aligning themselves at the center of the clump. This arrangement is further stabilized by presence of glycosaminoglycan (GAG) and serum amyloid P (SAP) which prevent proteolytic cleavage [1].

Primary amyloidosis of spine is rare. The exact incidence is unknown as it is only described in case reports. These are

benign lesion which primarily affect thoracic vertebrae [2], followed by cervical and lumbar spine. Despite being slow growing lesion, it can cause significant local tissue destruction which may lead to pathological fracture. Most patients will present with localized back pain and neurological deficit.

Xray and MRI findings are almost indistinguishable from metastatic tumor and metabolic disease of spine, making radiological diagnosis difficult.

Due to the deceiving features in MRI, tissue biopsy is critical in making a diagnosis. Histopathological examination requires special stain, Congo Red which when combined with polarized light will produce a characteristic apple-green color in microscopy. Because of amyloidosis has many types, electron microscopy and immunohistochemistry study is helpful for further characterization of amyloid deposit [3]

Treatment of choice is surgical excision of the lesion. Patient with primary amyloidosis usually have good prognosis after excision and recurrence is rare. Reduction of 50-75% of the lesion size is sufficient for regression of the amyloid deposit [4]. These patients should also be screened for co-existing plasma cell dyscrasias such as myelomatosis as failure to detect may lead to generalized amyloidosis [5]. Chemotherapy may also be used to suppress the underlying monoclonal plasma cell dyscrasias. Currently there is no role for radiotherapy in treating spinal amyloidoma.

#### 4. Conclusion

Spinal Amyloidoma is a rare disease affecting the spine. it should be considered in differential diagnosis of suspected spinal tumors and histopathological investigation is critical in making diagnosis. Patients with spinal amyloidoma have good prognosis following surgical excision of lesion.

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