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Solitary Plasmacytoma of the Spine: A Rare Case Managed Successfully with Surgery

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ABSTRACT: Solitary Plasmacytoma (SP)of the spine accounts for 5% of plasmacytoma cases and is considered a rare condition. Many of the patients suffer from myelopathy, approximately 42–71%, where lesions are located in the dorsal spine, more than 25–60%. The ultimate choice between stabilising and irradiating locally or directly is debatable according to the therapy and diagnosis. Relying on institutional expertise and patient results, advocate for different therapies through several centres. **Clinical presentation:** A 55-year-old patient presented with upper back pain diagnosed with D3-4 SP and was managed successfully with surgery. **Conclusion:** SP must be kept in mind as a differential diagnosis despite their location. Proteinemia abnormal levels might not always be present but are often seen in clinical settings. Patients, who are mostly misdiagnosed as having spinal Tuberculosis (TB), begin with Anti-tubercular therapy (ATT), which is harmful as it extends the conventional period of therapy. Patients advocate radiotherapy as it shows a decrease in recurrence occurrence.

Keywords: Plasmacytoma, Dorsal Spine, Surgery.

INTRODUCTION

SP is a condition where neoplastic monoclonal plasma cells aggregate in bone or soft tissue, commonly found in men, with a 2:1 ratio with a mean age of 55 years and an exponentially increasing incidence rate with advancing age [1]. SP is an uncommon disorder that has a global incidence rate of 0.191/100.000 for males and 0.090/100.000 for females, according to a recent Swedish population study [2]. SP primarily affects axial skeleton bones like the vertebra and skull. Computer Tomography (CT) and Magnetic Resonance Imaging (MRI) can detect bone destruction extent and multiple vertebral lesions, or bone marrow disease, respectively, aiding in diagnosis and treatment. In Solitary Bone Plasmacytoma (SBP), radiation therapy is considered the standard therapy, and in cases where bone instability or symptoms of the neurological disease worsen, surgical interventions are suggested. According to the extramedullary lesion resection here, radiation along with surgery may be appropriate. And in the case of adjuvant chemotherapy, it basically fails to enhance disease management and is unable to prevent it from developing into Multiple Myeloma (MM).

Plasmacytoma is a rare plasma cell dyscrasia that affects the axial skeleton and soft tissue without systemic symptoms. It is a precursor to plasma cell malignancies like multiple myeloma. It can be presented as solitary or multiple lesions anywhere in the body. The International Myeloma Working Group divides plasmacytoma into two types: solitary plasmacytoma of the bone (SPB) and extramedullary plasmacytoma (EMP). Multiple solitary plasmacytoma is even rarer and consists of monoclonal cell infiltrates in one or more lytic bone lesions or extramedullary tissue. Solitary plasmacytoma is more common in men and the median age is 55 years. Management includes surgery, radiotherapy, and chemotherapy [3].

Solitary bone plasmacytoma is a rare, immunoproliferative disease primarily affecting men, primarily affecting the axial skeleton. It presents a decade younger than myeloma (MM) and typically involves the head and neck region. Both SBP and EMP show absence of CRAB, with SBP characterized by confirmed solitary bone lesions and biopsy-proven plasma cell infiltration. Plasma cell dyscrasias are neoplastic B-cell proliferations, classified as multiple myeloma (MM) and plasmacytoma (localized extramedullary type). Solitary bone plasmacytoma (SBP) occurs in bone and extramedullary plasmacytoma (EMP) in soft tissue. SBP is infrequent, representing 3-7% of all plasmacytoma, and has a high risk of progression to MM. The present case of SBP is being reported for its unusual clinical and morphological findings and the role of fine-needle aspiration cytology in its early diagnosis [4].

Case Presentation

A 55-year-old female patient came to the hospital with complaints of bilateral lower limb pain, weakness for last 3 months, fever onset was on and off continuously and patient was able to stand with support. The patient had a history of type 2 Diabetes Mellitus (DM) with regular Blood sugar level (BSL) monitoring. Patient has no history of trauma, significant weight loss, tuberculosis or any previous surgery. On examination, power in bilateral leg was 3/5 and power in thigh and hamstring was 4/5. The patient was initially diagnosed with MM. On hematological examination, there was evidence of rouleaux formation, initially Bence jones protein was done which resulted negative and peripheral blood smear indicated mild rouleaux formation.

Laboratory Assessment

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Table (1): Chemical and Cerebrospinal fluid (CSF) examination.

Parameters	Observed Values	Normal Values
Sugar	111 mg/dl	50-80 mg/dl.
Protein	414 mg/dl	Up to 45 mg/dl
LDH (Lactate Dehydrogenase)	42 U/L	140 to 280 U/L
ADA (Adenosine Deaminase)	5.8 U/L	< 10 U/L

(mg/dl - milligrams per deciliter, U/L - units per liter).

MRI Spine was done for patient which revealed localized lesion around the right pedicle near the L4 vertebrae, the inferior endplate of the L3, and the sacrum. L3 right nerve compression indicated positive results due to sacrum pathological fractures observed in the L3 foramen right side. Left S1 nerve root compression was also seen due to the disc bulges at L3-L4, L4-L5, and L5-S1. [Fig. 1].

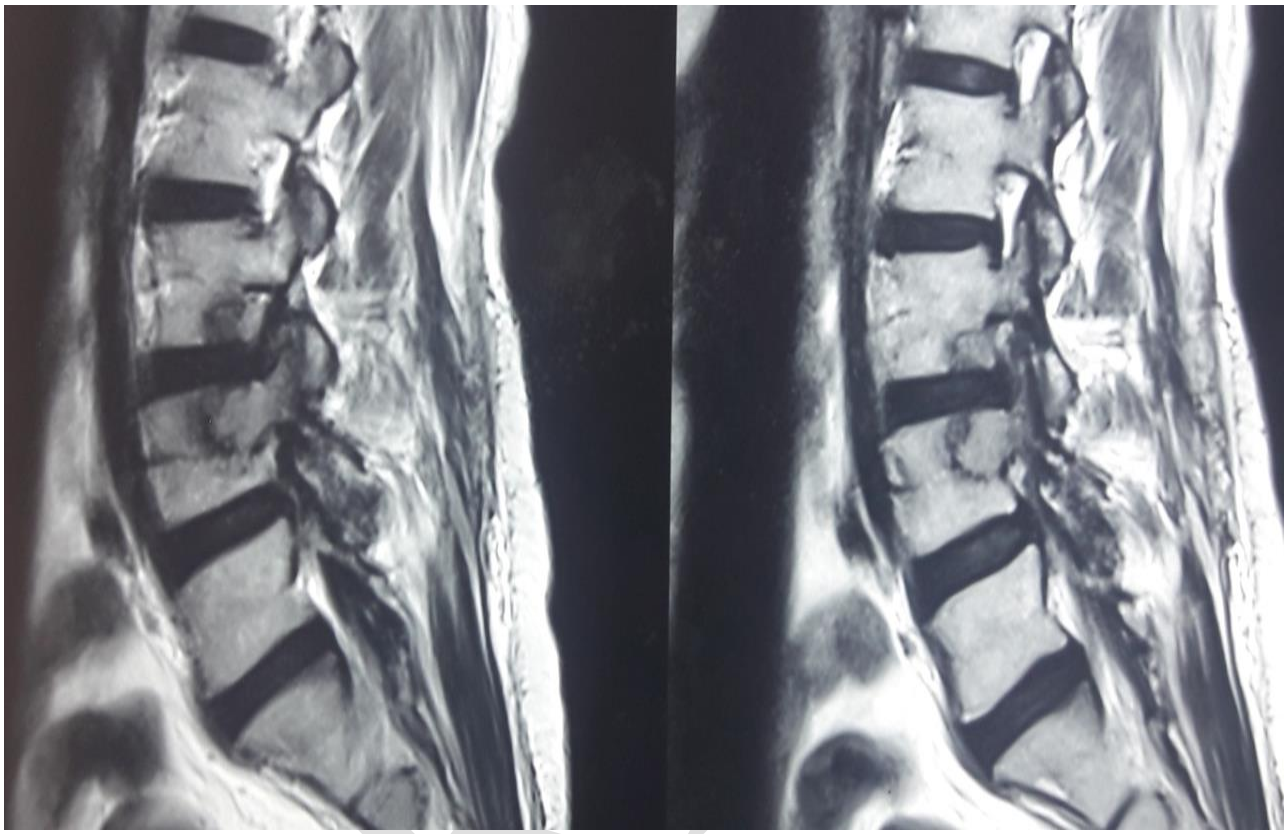

Figure (1): MRI dorsal spine sagittal view.

Since the lesion was showing a pathological fracture to look for spine metastasis Positron emission tomography (PET) Scan was done which revealed L4 vertebrae had metabolically active lesion. In the right sacrum, along with the L4 vertebrae, a lesion was seen that was metabolically active, with no other metabolically active lesion was found to suggest primary malignancy (Fig. 2). It was decided to treat the patient surgically. Thinking of plasmacytoma in mind and keeping a differential diagnosis of a solitary spinal lesion, the diagnosis was plasmacytoma (Fig. 3).

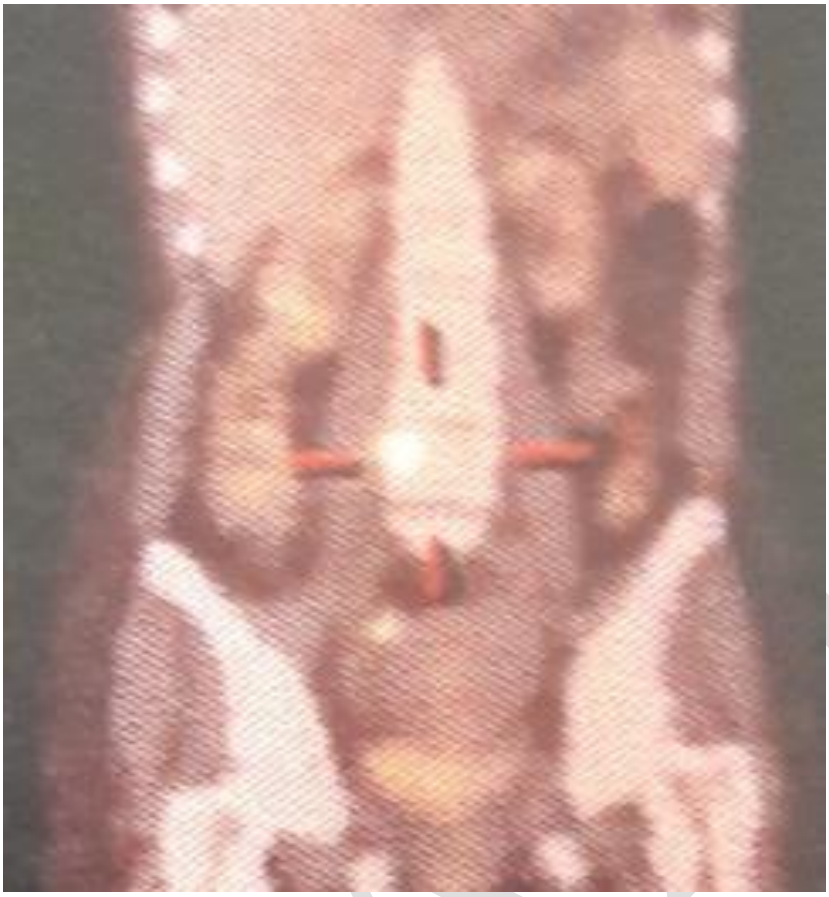

Figure (2): – PET Scan image shows FDG avid metabolically active lesion in L4 vertebrae.

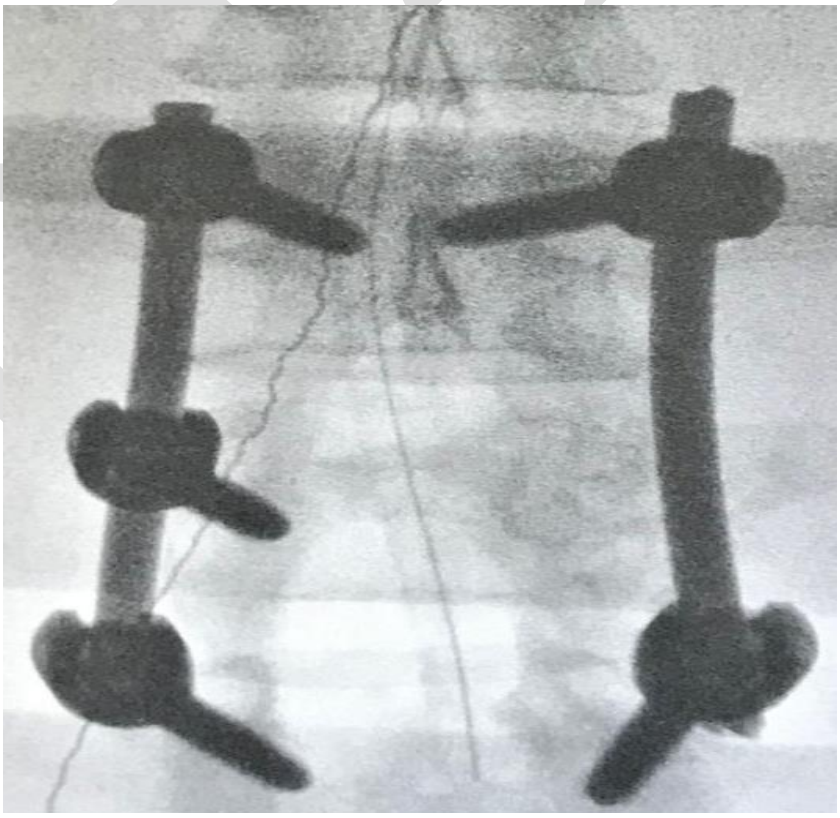

Figure (3): - Shows surgical intervention in the form of Decompression with screw fixation.

Histological examination on the specimen send was performed which showed CD 138 focal positivity more than 30% (Fig. 4).

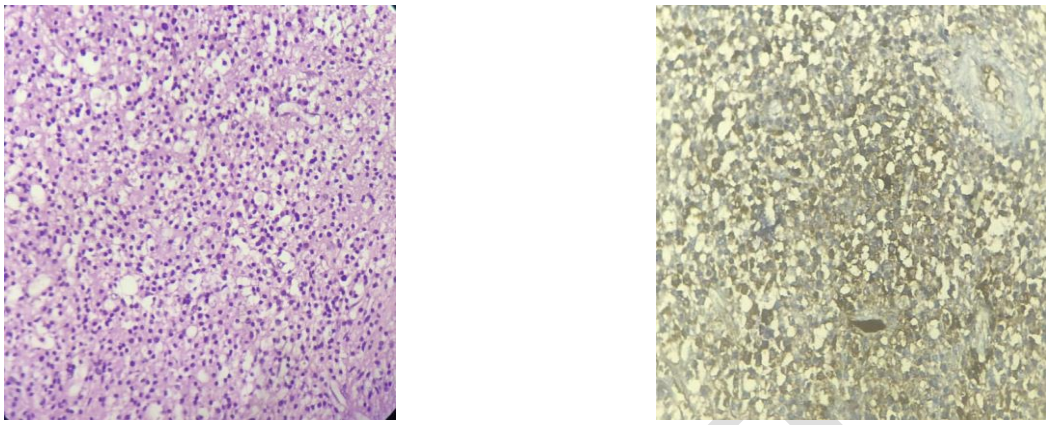

Figure (4): Histopathological examination (HPE) Specimen.

DISCUSSION

SP makes up around 5–10% of all dyscrasias, which is an uncommon malignant tumor of plasma cells. It is mainly diagnosed when the lesion is made up of monoclonal plasma cells and there is basically no systemic illness sign denoted. Radiation therapy is one of the modes of treatment [5].

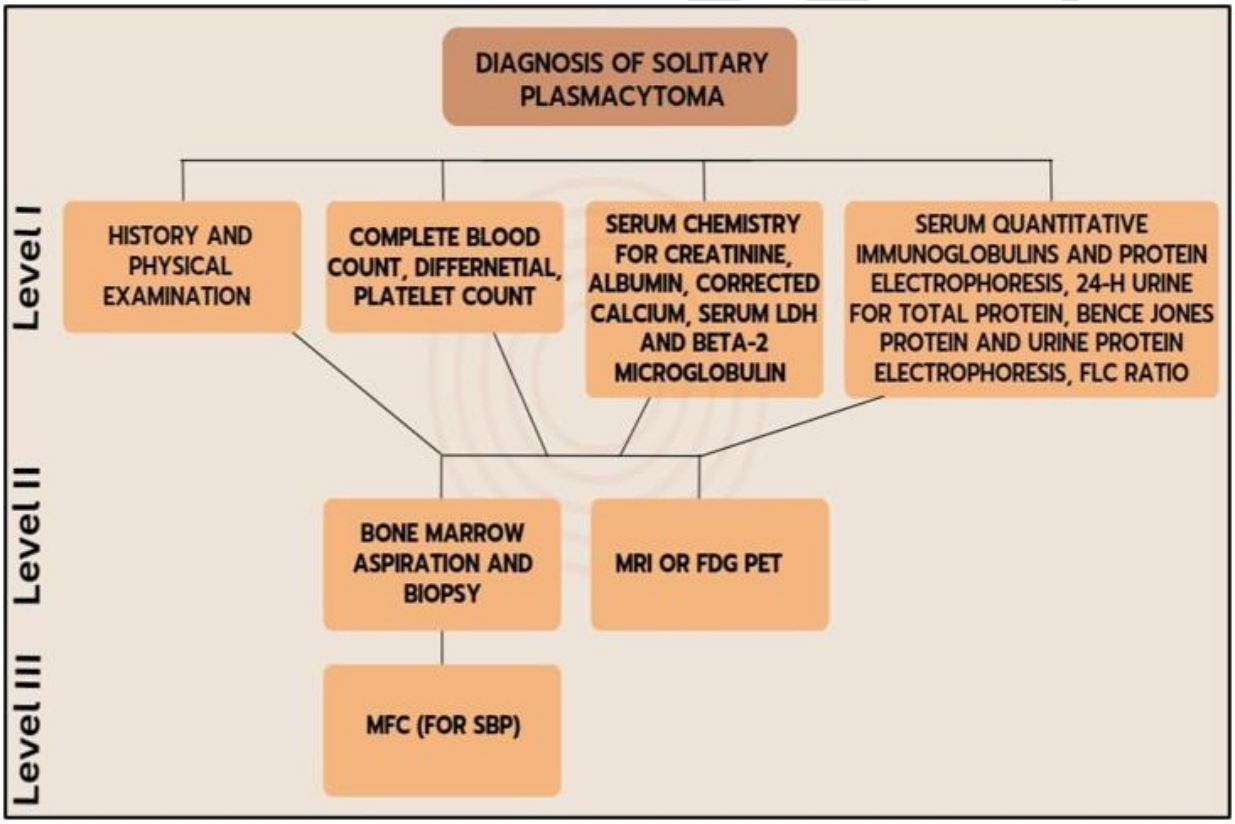

Figure (5): Diagnosis of Solitary plasmacytoma [6].

Solitary bone plasmacytoma affects roughly 2–5% of myeloma patients. Patients had a single bone lesion, no more than 5% of plasma cells in bone marrow, and received intensive radiation, except for one who had the lesion excised [7].

Nearly MM is developed within the initial 10 years from the diagnosis, and there are 50% chances for those with SBP and a 30% chance for patients with Extramedullary plasmacytoma (EMP) to develop MM [8].

SP is a disease classified as having less than 10% involvement in the bone marrow of a small plasma cell clone. The involvement of bone marrow is proven by flow cytometry, which is a sensitive test that shows that there is a higher incidence rate that reveals the progression of the disease to MM (56%–70%) with a shorter time period of 2–3 years.

The International Myeloma Working Group (1) has established diagnostic criteria for solitary plasmacytoma. Solitary plasmacytomas are diagnosed after ruling out systemic plasma cell diseases.

Table (2): Diagnostic Criteria.

Plasma Cell Disorder	Diagnostic Criteria	References
Solitary bone plasmacytoma/ solitary extramedullary plasmacytoma.	Biopsy-proven solitary destructive lesion of bone/ soft tissue mass of clonal plasma cells Absence of clonal plasma cells in bone marrow biopsy, aspirate. Normal skeletal survey, magnetic resonance imaging/ (computed tomography) of spine, pelvis (except for primary solitary lesion) If available positron emission tomography/ computed tomography showing solitary lesion. Absence of end-organ damage such as hypercalcemia, renal insufficiency, anemia or bone lesions (CRAB) attributed to plasma cell proliferative disorder.	Salgado LR, Wang S, Adler A, Chang S, Ru M, Moshier E, Dharmarajan K, Cho JH, Bakst R. The Safety Profile of Concurrent Therapy for Multiple Myeloma in the Modern Era. Advances in Radiation Oncology.2018 Sep 27.
Solitary plasmacytoma with minimal marrow involvement	As above but: Clonal bone marrow plasma cell detected but quantified to be <10%.	

5% of the plasmacytoma comprises solitary plasmacytoma along the spine, which is a rare entity [10]. SP of the spine shares similar features of bone, accounting for 25% to 60% of all cases. Spinal cord compression is common in SP of the spine, accounting for 71% of cases, compared to 7.4% to 16% in MM[11].

The choice of radiation vs. spine stabilisation remains a matter of debate. Radiation therapy plays a crucial role in addressing localized manifestations, particularly(SPs), in mature B-cell malignancies, primarily MM. The International Lymphoma Radiation Oncology Grouppresents guidelines for comprehensive evaluation and management of patients with SPs, offering insights into optimal imaging assessments, precise target volume definitions, and meticulous treatment planning.

SPs require a multidisciplinary team with the involvement of a surgical oncologist, a medical oncologist, a radiation oncologist, and a spine surgeon for effective treatment. The standard approach involves definitive local radiotherapy, with surgical interventions considered in specific scenarios. The use of advanced imaging modalities like MRI and PET/CT enhances diagnostic accuracy and treatment planning.

Naveen et al. studied Plasmacytomas arising in bone marrow or mucosal cells, sharing traits with plasma disorders like IL-6 dependence, genetic anomalies, and high-grade cytology/angiogenesis. Solitary Plasmacytoma of Bone (SPB) typically occurs in the axial skeleton with high red bone marrow involvement. Diagnosis involves a solitary bone lesion confirmed by CT or MRI, biopsy showing plasma cell infiltration, normal bone marrow biopsy, and no myeloma-related organ dysfunction. High-risk cases may benefit from Autologous Stem Cell Transplantation (ASCT), showing promise in recurrent cases. Systemic therapies, notably lenalidomide as second-line, have improved survival rates. Plasmacytoma requires differentiation from diseases like Multiple Myeloma (MM), non-Hodgkin lymphoma, reactive plasmacytosis, and plasmablastic lymphoma. Solitary plasmacytoma, though rare, necessitates lifelong follow-up due to potential recurrence. The case study underscores demographic and clinical diversity, challenging assumptions of elderly predominance and advocating for enhanced diagnostic methods and long-term management strategies [3].

Annapurna et al. reported a 40-year-old male patient presented with fractures left arm and left leg following a trivial trauma. He had progressively increased and painful swellings at both the sites. Histopathological analyses alone are not enough to diagnose MM, and FNAC can help in early provisional diagnosis. Extracellular and intracytoplasmic CI in plasma cells, composed of monoclonal immunoglobulins, have been reported in MM and lymphoproliferative disorders. The relationship between these inclusions and tumor progression, prognosis, and disease recurrence remains uncertain. Recent advancements in skeletal survey and molecular detection may reveal plasma cell monoclonality. SBP, a benign tumor, has a poor prognosis compared to EMP. It is more likely to progress to myeloma (MM) in patients with lesion size >5 cm, age ≥40, spine lesions, high 'M' protein levels, and persistence of 'M' protein after treatment. Treatment involves radiotherapy, surgical intervention, and adjuvant CT. The median time of progression to MM is 2-3 years, with a rate of 65-84% in 10 years. Rare appendicular skeleton SBP in young adults, with intracytoplasmic and extracellular CI in plasma cells, aids early diagnosis using FNAC in clinico-radiologically unsuspected cases [4].

CONCLUSION

When dealing with the solitary lesion in the spine, the differential diagnosis of bone plasmacytoma should be kept in mind. Radiation has a role in surgery, which lowers the risk of recurrence. It is not necessarily required to have abnormal proteinemia, also known as proteinuria, yet it is prevalent in healthcare settings. After the surgical decompression, patients have improved neurological as well as clinical benefits that were either with or without stabilization. After surgery, it is recommended to go for radiotherapy because it lowers the risk of recurrence. In an Indian scenario Starting ATT, which is frequently misinterpreted as spinal TB, can be hazardous to the patient because it delays the typical course of therapy.

Consent for publication

We declare that all authors have read and approved the paper. The paper has not been published previously, nor is it considered by any other journal.

Availability of data and materials

All data generated for this study are included in the article.

Author's contribution

Saurabh Boralkar: ideation, conceptualization, data curation, writing - original draft, writing - review & editing, visualization, validation. **Karishma Rathi:** ideation, visualization, validation, supervision, writing - review & editing, writing - original draft. **Rushika Mhaske:** writing - review & editing, visualization, validation.

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Conflict of interest

The authors declare no conflict of interest related to this work.

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