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Solitary Plasmacytoma of Spinal Cord: A Case Series

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ABSTRACT
Solitary plasmacytoma is a rare entity with an incidence of about 5% of all plasma cell dyscrasias. It is the monoclonal proliferation of plasma cells in either bone or soft tissue without systemic involvement. Depending upon location, these can be of two types: solitary bone plasmacytoma (SBP) and solitary extramedullary plasmacytoma (SEP). The most common site of involvement by SBP is axial skeleton, such as the vertebra, but intervertebral disc involvement is very rare. We have reported the involvement of disc with SBP in one of the cases of our case series. Diagnosis of plasmacytoma was confirmed by histopathological examination of biopsy tissue in all three cases. A bone marrow examination was done to rule out systemic involvement by the disease. An immunohistochemistry correlation was obtained in one case. The risk of progression to multiple myeloma (MM) is more in the case of SBP, and it is important to distinguish between the two as treatment and prognosis are different. We have presented this case series to highlight the occurrence of SBP with such clinical presentation and also to report the involvement of intervertebral disc with SBP, which is very rare, and only a few such cases are reported in the literature.

KEYWORDS: bone; intervertebral disc; plasmacytoma; solitary; spine

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1. Introduction

Plasma cell dyscrasias present with a varied clinical spectrum ranging from the indolent Monoclonal Gammopathy of Undetermined Significance (MGUS) to a more aggressive presentation of multiple myeloma (MM). Plasma cell dyscrasias are rare malignancies accounting for around 10% of all hematological neoplasms, and solitary plasmacytoma is even rarer, with an incidence of 1-5% [1]. Solitary plasmacytoma can be of two types: solitary plasmacytoma of bone (SPB) and solitary extramedullary plasmacytoma (SEP) [2]. Although SBP can involve any bone, like the skull, ribs, spine, and scapula, the vertebra is the most common. SEP occurs most commonly in the head and neck, with the nasal cavity being the most common site. SBP is more common in males, with M:F ratio being 2:1. Median age of the patients with either SBP or SEP is 55 years [3]. The criteria to diagnose SBP or SEP include single lytic bony lesion (for SBP) or extramedullary lesion (for SEP) histologically comprising of clonal plasma cell

proliferation, normal bone marrow aspiration and biopsy, no other bony involvement or end-organ damage [1]. Intervertebral disc involvement is infrequent in SBP, and only a few cases are reported in the literature [4].

2. Case Presentation

Case 1: A 57-year-old male was admitted to the neurosurgery department with a complaint of back pain for one month and weakness in bilateral lower limbs for 15 days. His medical history was unremarkable. On clinical examination, the power of lower limbs was 4/5, and deep tendon reflex was present. His routine blood and urine investigations were within normal limits, including creatinine, hemoglobin, calcium, and 24-hour urine protein electrophoresis. MRI dorsal spine showed T2 hypointense lesion involving D8 and D12 vertebrae with disc space destruction without spinal stenosis (Figure 1). Bone marrow examination was normal. Corpectomy was done with tumor resection. Intraoperative squash cytology revealed non-Hodgkin's lymphoma, possibly a plasma cell tumor (Figure 2). The biopsy sample showed a monotonous population of neoplastic plasma cells along with amyloid material. Complete skeletal radiographs revealed no evidence of other osteolytic lesions. The patient received radiotherapy postoperatively, and there was no evidence of disease during the six-month follow-up period.

Case 2: A 50-year-old female patient came to the neurosurgery department with bilateral lower limb weakness for two months. On examination, power was 1/5. Medical history was unremarkable, and hematological investigations were within normal limits. CT scan showed a lytic lesion at the L2 vertebra, and MRI showed L2-L5 cord stenosis with intervertebral disc showing bulging and cystic tumor mass involving the right posterior element of the L2 vertebral body encroaching on the spinal canal (Figure 3). No other lytic bony lesion was seen on radiographs. Tuberculin test was negative. L2-L5 laminectomy was done. Biopsy revealed sheets of neoplastic plasma cells, amyloid deposits, and intervertebral disc involvement (Figure 4). Bone marrow was within normal limits. Radiotherapy was given to the patient, who was disease free on a six-month follow-up period.

Case 3: A 60-year-old male patient presented to the neurosurgery department complaining of back pain and paraparesis with bladder involvement for one month. Medical history was unremarkable, and hematological parameters were normal. On CT scan, a lytic lesion was seen at the D8 level with the destruction of the vertebral body (Figure 5). MRI showed degenerative spondylotic changes and heterogenous marrow-replacing lesions (Figure 6). Biopsy showed a plasma cell tumor, further confirmed by immunohistochemistry (IHC). IHC studies showed diffusely positive CD138, kappa positivity, and lambda were negative with Ki 67 of 25-30%. Bone marrow aspiration and biopsy were within normal limits. The case was diagnosed as solitary plasmacytoma and received radiotherapy postoperatively. The patient had no recurrence of the disease and no transformation to multiple myeloma on a one-year follow-up period.

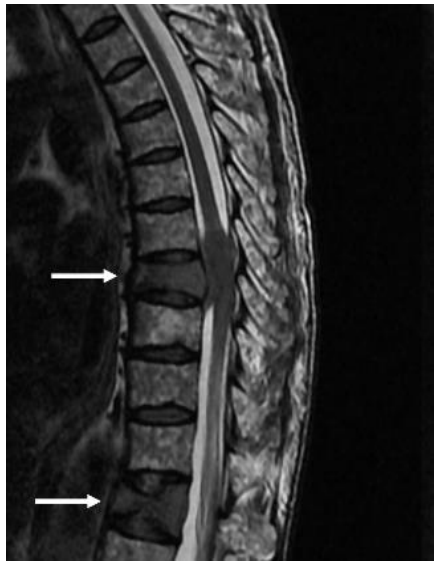

Figure 1. T2 sagittal MR image shows T2 hypointense lesion in D8 and D12 vertebra (arrows) with decreased height of D12 vertebra and posterior bulging of soft tissue at D8 level causing indentation of thecal sac.

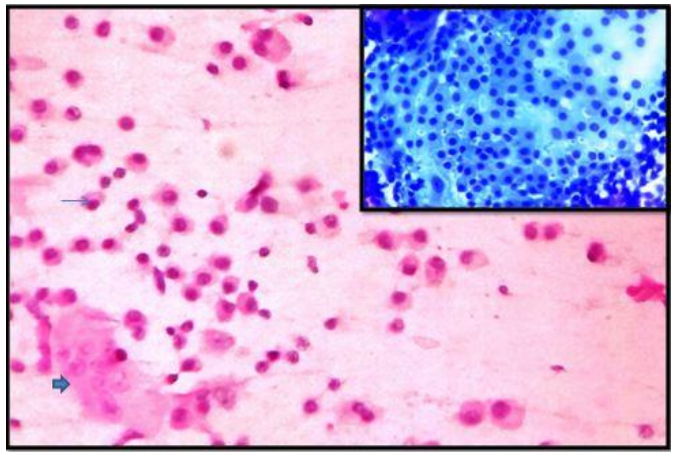

Figure 2. Smear shows monotonous population of neoplastic cells with round eccentric nuclei and perinuclear hof (arrow), along with a multinucleated giant cell (arrow head) (HE stain, x40); inset shows sheet of plasma cells (PAP stain, x40).

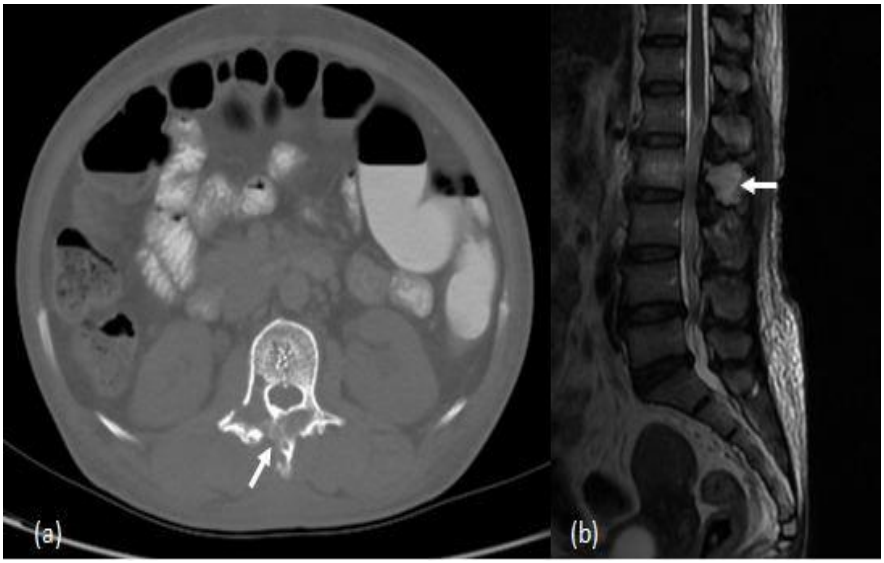

Figure 3. Axial CT image shows lytic lesion (arrow) involving right pedicle, right transverse process, lamina and spinous process of L2 vertebra (a); T2 sagittal MR image shows T2 hyperintense lesion (arrow) involving posterior elements of L2 vertebra (b).

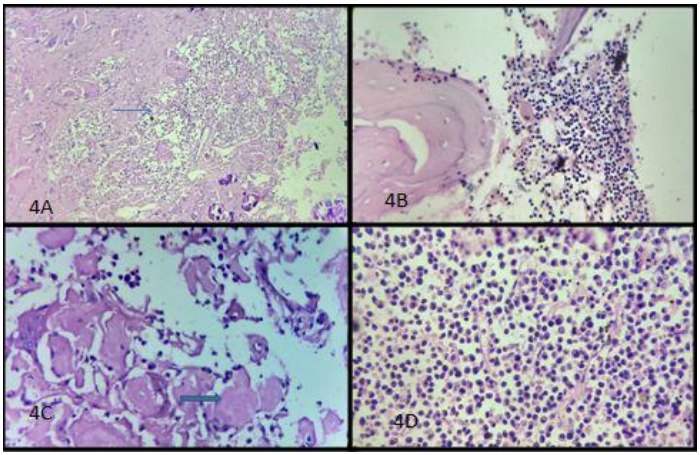

Figure 4. Section shows sheets of neoplastic plasma cells (arrow) infiltrating the bone (4A, HE stain, x4); section shows involvement of adjacent intervertebral disc tissue (4B, HE stain, x10); section shows pink amorphous material (amyloid) (arrow) seen surrounded by multi-nucleated giant cells (4C, HE, stain x40); uniformly distributed monotonous population of neoplastic plasma cells with round eccentric nuclei and perinuclear hof (4D, HE stain, x40).

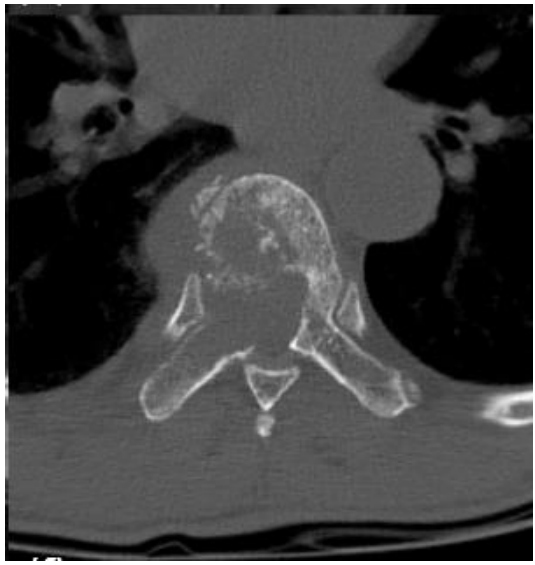

Figure 5. Axial CT image at D8 shows lytic lesion with associated paraspinal soft tissue.

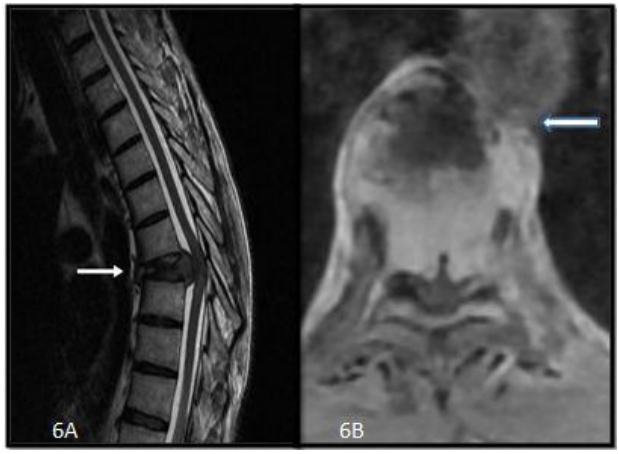

Figure 6. T2 hypointense lesion in D8 vertebra with collapse of vertebra and posterior bulging of soft tissue causing indentation of thecal sac with cord compression (6A); post-contrast axial image shows enhancement and associated soft tissue extending in anterior epidural space with spinal canal stenosis and cord compression (6B).

3. Discussion

Plasmacytoma is a rare hematological malignancy first described by Schridde in 1905, characterized by a mass of monoclonal plasma cell proliferation without systemic involvement [5]. It presents as a solitary lesion for years but can transform into multiple myeloma eventually. The most common sites of SBP are marrow-containing bones like the thoracic vertebra while that of SEP is the site with rich lymphatic drainage like the nasal cavity [6]. Median age of presentation is 55 years, and males are affected more commonly. The diagnostic criteria for SBP are biopsy-proven solitary lesion without bone marrow involvement, end organ damage, and no other lytic bony lesion on skeletal radiographs [1]. All three cases in our series fulfilled these diagnostic criteria. On imaging, SBP may show a "mini-brain sign" characterized by curved coarse trabeculae with hypertrophic sclerosis. One of the cases in our series showed involvement of intervertebral disc, which is a very rare occurrence. Usually, vertebral lesions involving two consecutive vertebral bodies and disc and paravertebral abscess involvement are suggestive of spinal infection. Unlike in our case where vertebral body collapse, disc involvement, and cord compression showed neoplastic plasma cell proliferation on biopsy. The adjacent disc destruction may be due to aggressive traits of plasmacytoma infiltrating and destroying adjacent bones, muscles, and fat [6]. So, in spinal lesions with adjacent disc involvement, neoplasia should also be considered in differential diagnoses along with infection. Bone marrow evaluation is important in such cases to diagnose and rule out other bone tumors and lymphomas [1]. Presently, regional radiotherapy is the recommended treatment for SBP at a dose of 40 to 50 Gy over four weeks, and its response rate is around 94% [7].

4. Conclusion

Due to the limited literature available about the exact incidence of SBP in India and keeping in mind its rarity, a complete histopathological workup of such spinal lesions is necessary, along with bone marrow evaluation. Complete workup is important to rule out the systemic nature of the disease, and periodic surveillance is needed to assess for transformation into multiple myeloma. Histopathological diagnosis was further confirmed by immunohistochemistry (IHC) in one of the cases of our series. The study has limitations, as IHC was not available in the remaining two cases. Our case series highlights the rare occurrence of SBP with disc involvement and will help to avoid misdiagnosis.

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Conflict of Interest Statement

The authors declare no conflict of interest.

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analysis, and review of draft, M.S.; Data Collection, data analysis, and review of draft, J.B. All authors have read and agreed to the published version of the manuscript.

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