Case Report

An unusual case of solitary spinal intradural plasmacytoma: The unforeseen challenge

ABSTRACT

Localized proliferation of atypical plasma cells, either at bony or extramedullary, forms a rare subset of multiple myeloma (MM) disorders. The patients usually present with intractable pain and pathological fractures and respond well to radiotherapy. The clinical presentation is variable and radiologically is nonspecific. The spinal location is rare, and the solitary plasmacytoma in the intradural extramedullary (IDEM) region is unusual. Herein, we report the second case of solitary plasmacytoma at the lumbar IDEM region. A 54-year-male patient was referred to our institute with complaints of radicular pain in bilateral lower limbs along L5 dermatome for the past 3 months. The neurological examination was normal with power 5/5 and reflexes 2+, except for bilateral straight leg raising test restricted at 30. Magnetic resonance imaging of spine showed a well-defined eccentrically placed (left Side) spherical lesion at the level of L2 vertebrae. The lesion had foraminal extension and showed minimal contrast enhancement. The underlying ventral vertebral body was irregular and hyperosteotic. Our radiological impression was primarily neurofibroma, but the features were slightly atypical. The patient underwent L1–L2 laminectomy and excision of IDEM tumor. The histopathological features were consistent with plasmacytoma. The histopathology was a surprise to us. We further evaluated the patient for MM. He received adjuvant radiotherapy and is currently asymptomatic. An index of suspicion for SP must be kept among differentials of intradural lesions, especially when adjacent bony changes are present. The diagnosis of plasmacytoma warrants further workup according to the recommendations of the International Myeloma Working Group.

Keywords: Hyperosteotic response, International Myeloma Working Group, intradural, radiotherapy, solitary plasmacytoma, surgery

INTRODUCTION

Absence of systemic plasma cell proliferation and focal accumulation of monoclonal plasma cells characterizes "solitary plasmacytoma" (SP). It is a rare subgroup of plasma cell neoplasm quantifying to nearly 5%–10% of all.[1] SP can be further subclassified into two groups, i.e., solitary bone plasmacytoma (SBP) and solitary extramedullary plasmacytoma (SEP), depending on whether the lesion originates in bones or soft tissues.[2] SBP arises from plasma cells of bone marrow, whereas SEP arises from cells in the mucosal surfaces. The incidence of SBP is nearly 40% higher than SEP, with predisposition toward vertebra and skull bones.[1] The patients usually present with intractable pain and pathological fractures and respond well to radiotherapy.[3,4] The median age of presentation in SP is 55–60 years, comparatively lower than in multiple myeloma (MM) patients, with the male-to-female ratio being 1.2:1–2:1.[3] The SP may be an isolated disease or the initial manifestation of a MM or Stage I myeloma.[4] The radiology is nonspecific and often may be misleading. Herein, we report a rare case of SBP at the lumbar level, who presented unusually with radiculopathy and masqueraded as intradural lesion on radiology. To best of our knowledge, ours is the second

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case in English literature, as single vertebral level intradural plasmacytoma.

CASE REPORT

Clinical history and examination

A 54-year-old male patient was referred to our institute with complaints of radicular pain in bilateral lower limbs along L5 dermatome for the past 3 months. The pain was progressive and not responding to conservative medical management. The patient had no other known medical comorbidities. The neurological examination was normal with power 5/5 and reflexes 2+, except for bilateral straight leg raising test restricted at 30°. There was no pint tenderness or step deformity. There was no positive finding on systemic examination.

Radiological assessment

Magnetic resonance imaging (MRI) of the spine showed a well-defined eccentrically placed (left Side) spherical lesion at the level of L2 vertebrae. The lesion had foraminal extension and showed minimal contrast enhancement. The underlying ventral vertebral body was irregular and hyperosteotic. The lesion was T1 isointense and T2 hypointense with obliteration of lateral recess [Figure 1]. In the T2 axial image, the nerve roots were seen shifted to the right side. The hypointense dura was seen intact throughout. The ventral hyperostosis was looking bony spur. Apart from this intradural lesion, there was no intervertebral disc prolapse seen. Our radiological impression was primarily neurofibroma, but the features were slightly atypical.

Surgery

The patient underwent L1–L2 laminectomy and excision of intradural extramedullary (IDEM) tumor. Intraoperatively, the tumor was well-defined, hard in consistency, capsulated, mildly vascular, and adhered to the ventral bone [Figure 2]. It was difficult to find plane of cleavage with ventral dura. The nerve roots were dissected off meticulously. The mass was coagulated, cut, and excised in piecemeal. We could not find any rootlet or root attached to tumor capsule. Once the tumor was detached from the bone, the ventral incomplete spur-like hyperostosis was curettage and excised. Our intraoperative impression was of meningioma or neurofibroma.

Histopathology

The histopathological study showed the tumor disposed in sheets of monotonous population of atypical plasma cells, with moderately pleomorphic eccentric nuclei, coarse chromatin, occasional conspicuous nucleoli, and moderate-to-abundant eosinophilic cytoplasm. Few binucleated cells were also seen. The features were consistent with plasmacytoma.

Postoperative evaluation

Postoperatively, the patient had an improvement in radiculopathy and he was discharged on the 5th day postoperative day. The histopathology was a surprise to us. The hyperosteotic bone and intradural location are rare for such pathology. We further evaluated the patient for MM. The bone marrow biopsy showed 3–4 intratrabecular spaces with overall cellularity of 30%–40%. The megakaryocytes were adequate with normal myelopoiesis in morphology and maturation. The erythropoiesis was also adequate and showed normoblastic maturation. Few interstitially scattered plasma cells were present, but no focal aggregates or excess blasts seen. Peripheral blood smear showed predominantly normocytic normochromic red cells with normal cell counts. Serum calcium was 9.7 mg/dl. Serum lambda free-light

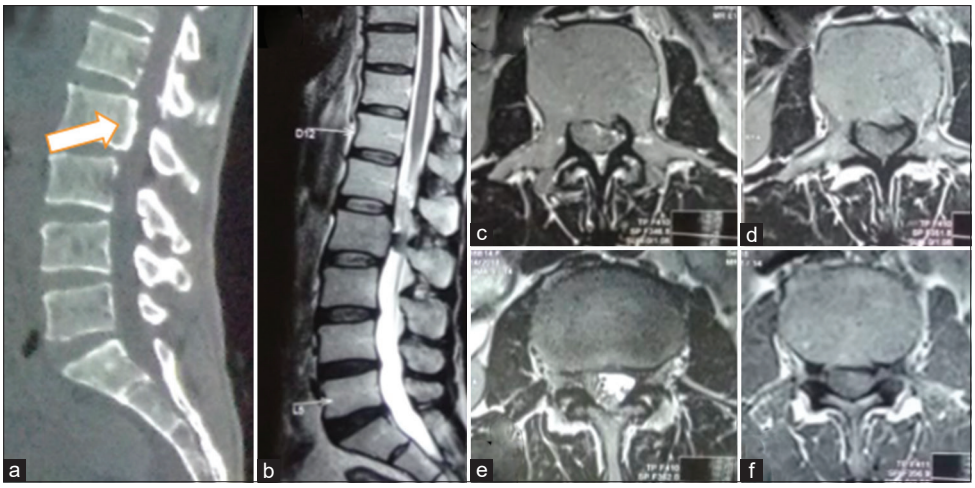

Figure 1: (a) Mid-sagittal computed tomography of lumbosacral spine suggesting hyperosteotic changes in the posterior surface of L2 vertebral body. The corresponding lumbosacral spine magnetic resonance imaging T2-weighted imaging (b) suggests a well-defined hypointense lesion at the L2 vertebral level with cerebrospinal fluid cut-off and nerve roots displaced; axial magnetic resonance imaging T1-weighted imaging at the L2 level (c, d, and f) suggests intact ventral dura (hypointense black line), with a bony spur-like hyperosteotic changes of vertebral body and well-defined mass located in the intradural extramedullary region; and (e) T2-weighted imaging axial magnetic resonance imaging below the L2 level shows displaced nerve roots to the opposite side

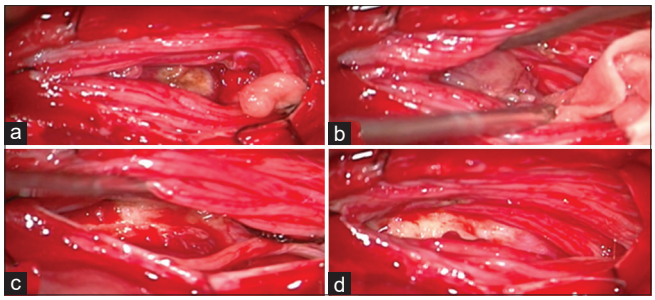

Figure 2: A well-defined lesion (a and b) seen after opening the dura and dissecting the nerve roots. The lesion was hard, pinkish, and mildly vascular. After excising the lesion, there were hyperosteotic changes in (c) adjacent vertebral body with no plane of cleavage (d)

chain and kappa free-light chain were 19.5 and 27.4 mg/l, respectively. Serum beta-2 microglobulin was 3.2 mg/l. Kappa/lambda ratio was 1:40. Neither serum immunofixation showed monoclonal protein nor serum electrophoresis showed any M-band. Bone scan showed no other uptake activity. Hence, it was a case of solitary SBP with intradural extension, and it was masquerading as IDEM lesions such as neurofibroma or meningioma.

Follow-up

The patient is under our outpatient follow-up. He received radiotherapy (30 Gy/10 fractions) and is currently asymptomatic after 12 months of surgery.

DISCUSSION

Among the differentials of intradural, the SP is an extremely rare pathology. The nonspecific radiologically and varied clinical findings compared to other differentials of intradural forge SP as a surgical challenge. SP is a discrete mass of monoclonal plasma cells and forms a part of plasma cell proliferative disorder spectrum (smoldering MM, non-IgM monoclonal gammopathy of undetermined significance (MGUS), IgM MGUS, light chain MGUS, SP, SP with minimal marrow involvement, POEMS syndrome, and systemic AL amyloidosis). It represents an "early-stage plasma cell malignancy" placed between MGUS and MM. It is further subclassified into two groups, i.e., SBP and SEP, depending on whether the lesion originates in bones or soft tissues.[2] The rare group, "intradural SP," is commonly located in the cranium, with the spinal location being extremely rare [Table 1].

The International Myeloma Working Group (IMWG) has been recently modified the diagnostic criteria for plasma cell proliferative disorders.[3] The diagnostic criteria (for SP) include (a) biopsy-proven solitary lesion of bone or soft tissue with evidence of clonal plasma cells, (b) normal bone marrow with no evidence of clonal plasma cells, (c) normal skeletal survey and MRI (or computed tomography [CT]) of the spine and pelvis (except for the primary solitary lesion), and (d) absence of end-organ damage such as hypercalcemia, renal insufficiency, anemia, and bone lesions or amyloidosis that can be attributed to the plasma cell proliferative disorder. IWMG recommends the use of low-dose whole-body CT or MRI in the workup of SP.[3,4] The finding of lytic lesions in CT warrants the initiation of treatment in an asymptomatic patient.

Sod and Wiener described the first case of intradural plasmacytoma in 1959, but their case has multiple-level involvement.[5] It was Zazpe *et al*. who reported the first case of solitary intradural plasmacytoma.[6] Their case was a 25-year-old female, presented much earlier than our case with complaints of paraparesis and sensory involvement, in contrast pure radiculopathy in our case. The thoracic level tumor in Zazpe *et al*.'s report showed homogeneous contrast enhancement. The SP seems to have nonspecific radiology and even intraoperative inferences are variable.[7] The difficulty faced by Sod and Wiener in 1957 when they wrote "It looked like a typical meningioma or neurofibroma and the fifth thoracic nerve root was resected with the tumor" continues even today.[5] This confusion persists until histopathology surprisingly comes to be plasmacytoma. The usual presenting age for plasmacytoma is around 55 years. At an early age, SP may be the initial manifestation of MM.

Other reported cases on solitary dural plasmacytoma are all from the cranial region. The radiological feature of cranial SP, however, is nearly corresponding to our case. These lesions are isointense to hyperintense on T1-weighted images and isointense to hypointense signal on T2-weighted images, with homogeneous mild-to-moderate contrast enhancement.[8,9] The characteristic moth-eaten appearance and "mini brain appearance" are not found in dural plasmacytoma, as was missing in our case. The dural involvement occurs through contiguous route from the adjacent bone or hematogenous spread.[10] One more conundrum in radiology was "non osteolytic" appearance. The hyperosteotic bone misleads us in line of meningioma. On retrospective review, we found that abnormal osteoblastic response to bone destruction may occur in plasmacytoma.[11-13] These lesions are hyperintense or isointense on T1-weighted images and hyperintense on T2-weighted images and enhance on contrast imaging.

According to a review by Lebrun *et al*., it was found that SP is a separate clinical entity with comparatively longer progression-free survival.[7] The osseous involvement has a poor prognostic sign. Radiotherapy is the standard treatment for SP, but there are no specific guidelines on the optimum dose. A general consensus is to use approximately 40 Gy for solitary bony lesion including adjacent normal vertebra.[6]

Table 1: Review of the reported cases of intradural plasmacytoma in literature

	Author	*n*	Age (years)/Gender	Clinical presentation	Location	T1WI/T2WI MRI	Contrast MRI	Surgical management	Adjuvant therapy
1	Zazpe *et al*.[6] (2007)	1	25/female	Pain with paraparesis	Intradural extramedullary at D2 D3 level	Intermediate-low homogeneous signal in both T1 and T2 sequences	homogeneous contrast enhancement without dural-tail or bone alterations	Laminectomy and gross total resection	30 Gy local radiation therapy
2	Hans FJ *et al*.[16] (2013)	1	52/male	Quadriparesis	Intramedullary at C5 C6 level	Hyperintense on T1 and T2 signal intensity	mild to moderate nodular homogeneous contrast enhancement	Hemilaminectomy with biopsy	radiotherapy in 40 Gy/20 fractions combined with systemic dexamethasonetreatment.
3	Gao B *et al*.[15] (2007)	1	31/male	Paraparesis	Intramedullary at T6-10 level	Isointense on T1- and hyperon T2weighted images	Irregular enhancement	Laminectomy with gross total tumor excision	Not mentioned
4	Present case	1	54/male	Radiculopathy	Intradural extramedullary at L2 level	Isointense on T1 and hypointense on T2	minimal homogenous enhancement	Hemi-laminectomy and gross total excision	radiotherapy (30 Gy/10 fractions)

n is number of cases; and MRI magnetic resonance imaging

We used further lower dosages because surgical excision was complete. Chemotherapy is recommended for refractory and/or recurrent dural-based SP.[10]

The prognosis of disease is guarded. There are three patterns of failure; these are the development of MM, local recurrence, and development of new bone lesions without MM. The SBP has a higher risk for progression to myeloma (nearly 65%–84% in 10 years), and with the best of treatment, the median time to progression to MM is 2–3 years. The long-term prognosis of spinal intradural plasmacytoma is not known due to scarcity of literatures. Tsang *et al*. showed that the patients of SBP, with size of lesions <5 cm, had a higher local control rate (nearly 100%).[13] Similarly, Wilder *et al*. reported that the persistence of M protein, more than 1 year after radiotherapy, was the only independent adverse prognostic factor.[14] The skeletal survey of our patient, after 9 months of radiotherapy, showed no recurrence.

CONCLUSION

SSIP is a rare clinical entity, with nonspecific radiological features. An index of suspicion for SP must be kept among differentials of intradural lesions, especially when adjacent bony changes are present. The diagnosis of plasmacytoma warrants further workup according to the guidelines of IMWG. The radiotherapy response is nearly 80% and forms the mainstay of management. Gross excision, wherever possible, followed by radiotherapy forms the best management option.

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 initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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