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Case Report

Oncology Section

Lumbar Chordoma Presenting as an Epidural Collection with Vertebral Destruction- An Unusual Cause of Lumbar Canal Stenosis

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ABSTRACT

Chordomas are locally aggressive neoplastic lesions that arise from physalipherous cell nests that originate from notochordal remnants left behind during early foetal development. Chordomas of the mobile spine (C3-L5) constitute less than 5% of the overall incidence of chordomas in the spine. They generally are osteodestructive leading to vertebral collapse and severe deficits including paraplegia and quadriplegia. Here authors presented a case of 26-year-old female presented with severe bilateral L5 radicular pain, no deficits and intact bladder and bowel control. The pain was progressive and resistant to analgesic medication. On imaging a collection in the lumbar spine emanating from the L5 vertebral body extending into the canal leading to secondary spinal canal stenosis was observed. On surgical exploration a soft friable vascular mass compressing the dural tube and the exiting nerve roots was observed and the mass was analysed and found to be a chordoma. Although, these lesions are seen to compress the vital neural elements of the spine, the presence of a collection in the spinal canal was unusual and resembled tuberculosis which is a much common lesion, or pyogenic osteomyelitis of the vertebral body. Both differentials were proven wrong. Such an approach not only detected and treated the lesion early, but good rehabilitation and adjuvant therapy was initiated enabling excellent overall quality of life to the patient.

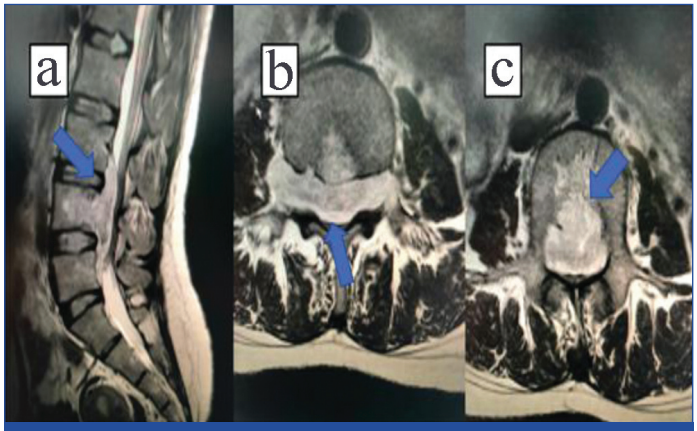
Keywords: Instability, Laminectomy, Mobile spine, Spinal bony tumours, Tuberculosis

CASE REPORT

A 26-year-old female, presented to the Neurosurgical Outpatient Department with severe radicular pain over both her lower limbs for the past three months. The pain was severe and incapacitating, and limiting her routine daily life activities. The pain worsened on standing and walking but decreased on lying down. No back pain was present nor was there any history of trauma. No spinal or epidural anaesthesia was given during the delivery of her child six months ago. On examination, she had no focal deficits, but her straight leg raising test was positive in both lower limbs at 45 degrees. Bilateral ankle jerks were present and both plantars were flexor.

Magnetic Resonance Imaging (MRI) of lumbo sacral spine showed a collection in the spinal canal extending from L3-L5 causing compression of the exiting L5 nerve root and the cauda equina. The maximum compression was over the L4 vertebra and under the L5 exiting nerve root [Table/Fig-1]. Computed Tomography (CT) scan of lumbo sacral spine showed destruction of the posterior aspect of the L4 vertebral body adjacent to the collection [Table/Fig-2], bone densitometry showed poor minerisation of the bone. Erythrocyte Sedimentation Rate (ESR) (40 mm/hr) and C-Reactive Protein Test (CRP) (2.7 mg/L) were elevated, and mantoux test was also positive. Due to the prevalence of tuberculosis, positivity of the mantoux, elevated ESR and CRP, a tentative diagnosis of spinal tuberculosis was made with the cold abscess causing secondary lumbar canal stenosis (narrowing of the lumbar spinal canal). The patient was counselled appropriately and had undergone a decompressive laminectomy of the lumbar spinal canal at L3-L5 with stabilisation of the lumbar spine with an L3-L5 lumbar pedicle screw and rod construct.

The possibility of pyogenic osteomyelitis and vertebral tumours were also entertained, and due to the fear of instability after laminectomy, stabilisation was planned if there was no pyogenic infection. A stage stabilisation was contemplated, in the event of a pyogenic infection after adequate appropriate antibiotic therapy.

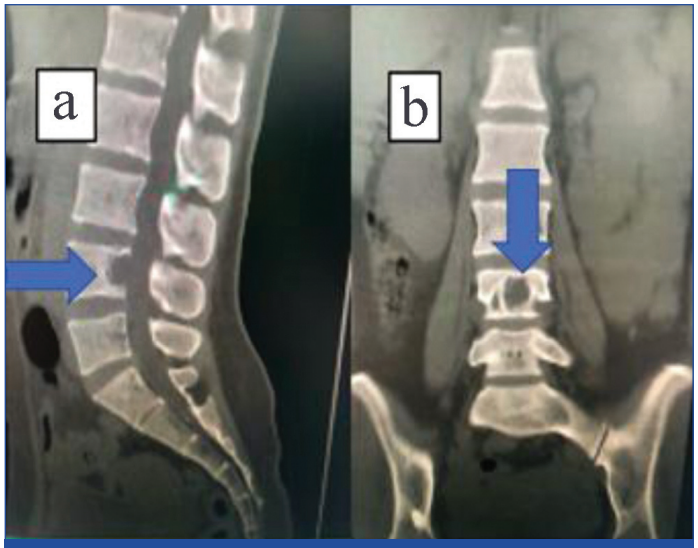


[Table/Fig-1]: Preoperative Magnetic Resonance Imaging (MRI) of the lumbosacral spine. The MRI shows, a) sagittal view of the lumbosacral spine in T2 which shows a collection in the spinal canal emanating from the L4 vertebral body compressing the cauda equina, (Blue arrow); b) axial T2 view showing the collection/pus extending along the nerve roots into the neural foramina compressing the exiting nerve roots, (Blue arrow) and c) axial T2 image showing extension of the collection/lesion into the vertebral body (Blue arrow).

Following lumbar laminectomy, a soft friable vascular mass occupying the anterior aspect of the spinal canal displacing the dura posteriorly was found. The mass was extending superiorly up to L3 and inferiorly to L5. It was arising from the L4 vertebral body with destruction of the posterior cortex. The mass was extending laterally along the exiting nerve root stretching the nerve root and compressing it at the foramen. The mass was removed and the lumbar canal was decompressed.

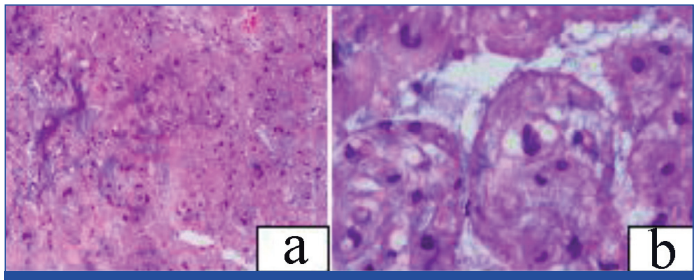
Postoperatively, the patient improved and was ambulated on postoperative day 2 without any focal deficits. She was discharged on postoperative day 4. Microbiological evaluation yielded no bacterial growth. There were no bacteria or acid-fast bacilli in smear.

Histopathological evaluation revealed neoplastic tissue composed of sheets and occasional whorling pattern of cells. The sheets consisted of large polyhedral cells with central atypical pleomorphic nuclei,

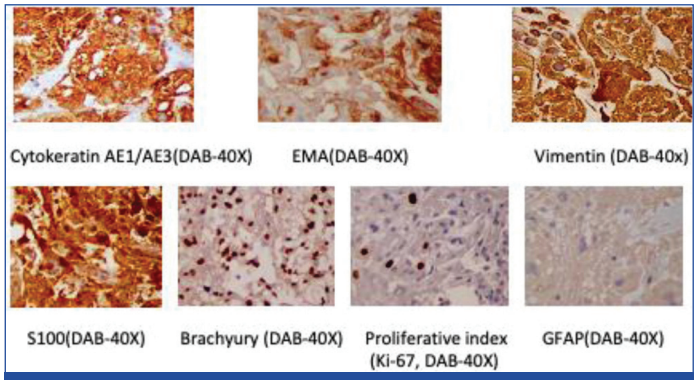


[Table/Fig-2]: Computed Tomography (CT) scan of the lumbosacral spine demonstrating the extent of body destruction over a short period of time in a) sagittal and b) coronal views (Blue arrows).

irregular nuclear contours and abundant eosinophilic cytoplasm. Occasional binucleate, multinucleate and bizarre forms were also seen. The cells had bubbly cytosol with pseudo inclusions set in a stroma showing marked myxoid change along with focal chondroid areas. These physalipherous cells were arranged in lobules and chords with some just present individually as well [Table/Fig-3]. The tumour also showed abundant cytokeratin, Epithelial Membrane Antigen (EMA), S100 and vimentin but was negative for Glial Fibrillary Acidic Protein (GFAP). Brachyury was also positive consistent with chordoma. The Ki-67 proliferative index was approximately 5% [Table/Fig-4]. The patient was counselled and advised radiation therapy. She is currently undergoing radiation therapy and under regular follow-up. She is being seen every three months for six months, then every six months for one year, after which she is suggested for yearly with an MRI and CT of her spine.



[Table/Fig-3]: Histopathology H&E section in a) low 10x and b) high 40x magnification shows large epithelioid cells with central bland nuclei and abundant eosinophilic, finely vacuolated cytoplasm.



[Table/Fig-4]: These neoplastic cells show positivity for cytokeratin, Epithelial Membrane Antigen (EMA), and S100. Glial Fibrillary Acidic Protein (GFAP) is negative. Ki67 proliferation index is low (<5%). Brachyury is strongly positive (40x).

DISCUSSION

Spinal chordomas are rare, constituting only 10-15% of the incidence of chordomas in toto. The lesions are seen most frequently in the sacrum (80%) followed by the craniocervical junction (15%). Chordomas across the rest of the mobile spine are extremely rare and present as destructive lesions leading to collapse of the affected vertebral body leading to instability and pain [1,2]. These lesions rarely cause radiculopathy and canal stenosis by themselves without vertebral body destruction and collapse. Intradural extensions are almost unheard of [2]. Chordomas develop from notochord remnants and seldom present with such a short history. They are slow growing, low grade malignant tumours. They have been known to be locally invasive [3,4].

Treatment of chordomas consists of radical excision, stabilisation, followed by adjuvant treatments (radiation and possibly chemotherapy) [4]. Based on well established principles, resection should involve obtaining a wide margin or performing en bloc resection. It is generally accepted that the resected margin should be free of tumour. It is recommend to perform en bloc excision of all spinal chordomas when feasible [3,4].

If an en bloc resection is not feasible, palliative debulking followed by radiotherapy is worthwhile [4,5]. Although chordoma is known to be relatively radioresistant, the value of radiation therapy has been stressed in several reports and series [4,5]. As most studies cover long periods, during which new techniques were developed, and as the tumour is rather rare, there is no consensus in the literature regarding the optimal radiotherapy scheme for cervical chordomas [4,5]. Proton beam and stereotactic radiotherapy are currently being investigated and recommended as treatment for these lesions However, there is no clear evidence about the efficacy of these modalities at this time [5,6]. Chemotherapy does not play a role in the treatment of chordomas, although the use of imatinib mesylate is being investigated [6].

Chordomas are well known to recur locally [5-7]. The recurrence rate seems to be related to the incompleteness of resection. Chordomas of the mobile spine metastasise more often than sacrococcygeal lesions. The reported incidence of metastases varies widely, from 3-60%. They are discovered between one and ten years after the initial diagnosis. Sites of metastases include bone, lungs, lymph nodes, soft tissues, intrathecal space and liver [6-8].

Only primary and complete resection of the tumour offers a major advantage as far as prognosis is concerned [7,8]. Clearly, the length of follow-up in the present study is short, but the extent of tumour resection that could be achieved using these techniques, as well as the limited risk of morbidity encountered and the successful stabilisation that was accomplished, has been encouraging.

Authors also reviewed recent literature and presented a look at the last 10 years of lumbar spine chordomas presenting with secondary spinal canal stenosis, to highlight the rarity of the case along with management. Only four cases of lumbar mobile spine chordomas have been reported between 2010 and 2021 and have been treated with a variety of procedures ranging from vertebroplasty to vertebrectomy [Table/Fig-5] [9-12].

Study	Procedure employed
Chatterjee S et al., [9]	Vertebroplasty
Zuccato JA et al., [10]	Laminectomy and decompression with pedicle screw fusion
Nishiguchi T et al., [11]	Vertebrectomy with stabilisation
Stambough JB et al., [12]	Laminectomy with pedicle screw stabilisation and decompression of lesion
Ganapathy S et al., (current report)	Laminectomy with stabilisation and tumour excision

[Table/Fig-5]: List of previous case reports of mobile spine chordomas along with the surgical procedure employed for their management [9-12].

Authors proceeded with a standard fusion with decompression and complete excision offering the patient an improved chance at recovery.

CONCLUSION(S)

Spinal chordomas are great mimics, resembling infectious and other neoplastic pathologies. A high index of suspicion assists in a quick diagnosis and early treatment and a longer disease free and symptom free survival.

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