

Chordoma of Lumbar Vertebra: Case Report

Lomber Vertebra Kordoması

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ABSTRACT Chordomas are rare, slow-growing but locally aggressive tumors. These tumors arise from remnants of the notochord and can be found from the skull base to the coccyx. Most chordomas arise from the clivus or the sacrococcygeal regions. In the spine, the sacrum is the most common site of disease, followed by the lumbar spine and then the cervical spine. Pain in the lower back and sacrum is the most common presenting symptom. The ideal treatment of chordomas consists of wide or marginal en bloc resection. Adjuvant radiation therapy can be used for the treatment of residual or recurrent tumors. We reported a lumbar chordomas which was seen at the level of L-1.

Key Words: Chordoma; lumbar vertebrae

ÖZET Kordomalar, nadir görülen, yavaş büyüyen fakat lokal olarak agresif tümörlerdir. Kordomalar, notokord artıklarından kafa tabanından koksikse kadar herhangi bir yerden kaynaklanabilirler. En sık klivus ve sakrokoksigeal bölgede yerleşirler. Omurgada ise en sık sakrumda, sonra lomber vertebralarda en nadir de servikal vertebralarda görülürler. Sırt bölgesindeki ağrı kordomalarda en sık görülen semptomdur. Kordomalarda ideal tedavi geniş ve tümör sınırlarının tam çıkarımı ile sağlanır. Rezidüel veya rekürren tümörlerin tedavisinde adjuvan radyoterapi kullanılabilir. Biz olgumuzda L-1 seviyesine yerleşmiş lomber kordomayı sunduk.

Anahtar Kelimeler: Kordoma; lumbar vertebra

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Chordomas are rare, slowly growing, locally aggressive tumors arising from embryonic remnants of the notochord. The majority of tumors are in the sacrococcygeal region (48%) or spheno-occipital region (39%), where notochord rests also exit.^{1,2} Of the tumors that do not arise in the sacrum or clivus, half occur in the cervical region, with the remainder found in the lumbar or thoracic region, in descending order of frequency. Plain films demonstrate bone destruction, with areas of amorphous calcification. A large soft tissue mass and involvement of two or more adjacent vertebral bodies and the intervening disk are frequently seen.^{3,4} Computerized tomography (CT) is best to delineate areas of osteolytic, osteosclerotic, or mixed areas of bone destruction. Magnetic resonance imaging (MRI) with gadolinium enhancement provides the best imaging of epidural tumors seventy-five percent of chordomas are isoin-

tense to the cord on T1-weighted images, and 25% are hypointense. The lesions have high signal intensity on T2-weighted images.⁵ Prominent enhancement is seen, as well as internal septations, cystic changes, and areas of hemorrhage. We report this case because chordomas seldomly occur in upper lumbar region.

CASE REPORT

A previously healthy 62-year old male patient presented with a 6 months history of progressive low back pain. He reported no history of trauma. Physical and neurologic examination did not reveal abnormalities. He had an unremarkable medical history. Radiographies of lumbar spine showed L1 compression fracture (Figure 1). MRI revealed an epidural contrast enhanced tumor and determining fracture of the vertebral L1 body and compression of the spinal cord at this level (Figure 2). A general analysis by CT revealed no tumor activity

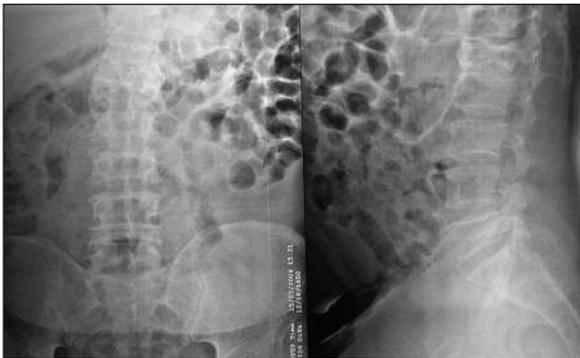


FIGURE 1: Pre-operative x-ray showing lumbar 1 compression fracture.

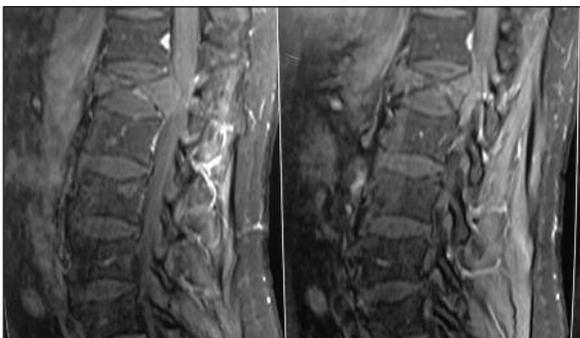


FIGURE 2: Pre-operative contrast enhancement MRI showing lumbar 1 pathological fracture and epidural mass.

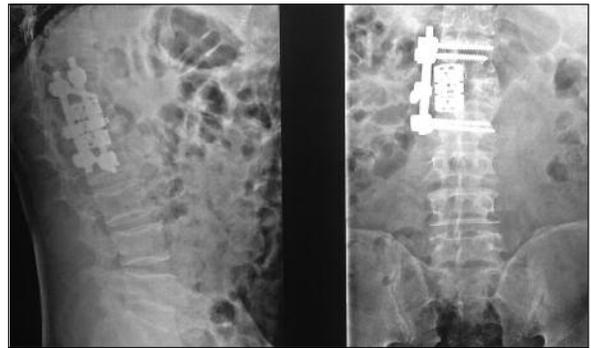


FIGURE 3: Post-operative x-ray showing thoracal 12- lumbar 2 interbody fusion and stabilization after resection of the tumor.

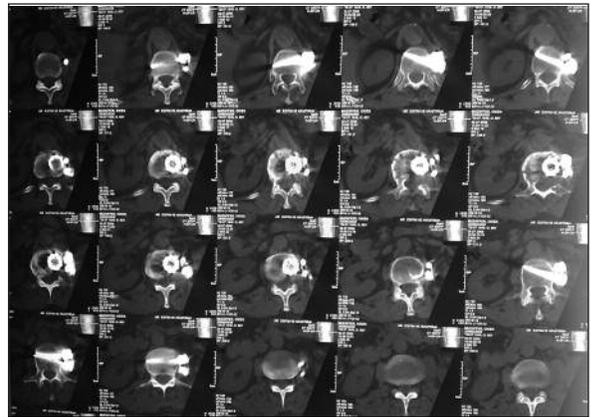


FIGURE 4: Post-operative computer tomography scans showing decompressed spinal canal and resection of the tumor.

elsewhere in the body. The patient underwent surgery using anterior approach. L1 vertebral corpectomy and total tumor excision with interbody fusion and stabilization procedure were performed (Figure 3, 4). Macroscopically the involved bone was grayish white with cheesy consistency. The patient tolerated the procedure without difficulty and his low back pain resolved. Microscopic examination of the tumor showed small, oval or round, eccentric nuclei with dense chromatin pattern, multiple vacuoles of different sizes and abundant eosinophilic cytoplasm. Immunohistochemical analysis showed expression on S100 protein, epithelial membrane antigen, cytokeratin, vimentin but no expression on CK7, CK20 and CD10. The initial analysis most likely look like renal cell carcinoma. Because of the lack of a primary tumor, re-examination of the specimen changed the diagnosis into vertebral chordoma.

DISCUSSION

Chordomas constitute less than 4% of all primary bone tumors. These tumors are slowly growing, locally invasive neoplasms; pain is the most common symptom. There is a definitive 2:1 male-female ratio. In adults there is a peak incidence in the sixth decade.³

Lumbar spine is an uncommon localization for chordoma. Lumbar chordoma may easily be confused with more common lesions in the lumbar spine such as metastasis, bone-base neoplasms, meningioma, hemangioma, schwannoma, chronic infections such as tuberculosis and preoperative diagnosis can be difficult.

Radiographic features and intraoperative macroscopic appearance of the chordoma and full body investigation can be helpful in preoperative diagnosis. The radiological features of chordomas do not depend on the anatomic location of the lesion. Soft tissue mass of the involved vertebra is the most important radiological finding. Bone destruction, prominent enhancement, internal septations, cystic changes, areas of calcifications and hemorrhage can be seen in chordomas. Macroscopically, chordomas are soft and gelatinous, with a smooth or lobulated grayish white outer surface. The cut surface is usually homogenous, but calcification and occasional hemorrhage are not unusual.⁶ Full body scanning can be helpful in differential diagnosis for metastasis. The diagnosis can be difficult especially in atypic localization of the tumor. Von Kollenberg reported a similar of our case.⁷ Their initial microscopic evaluation was suggestive of renal cell carcinoma like ours but lack of primary tumor changed the diagnosis into chordoma.

The differential diagnosis of chordoma in a small biopsy, when entire tumor is not available for pathologic examination, includes metastatic renal cell carcinoma and other similar appearing neoplasms, mucinous adenocarcinoma of colorectal and

myxopapillary ependymoma. Coffin et al compared immunohistochemical features between chordoma and renal cell carcinoma.⁸ In their study all chordomas were immunoreactive for vimentin and cytokeratin, 83% for epithelial membrane antigen and 83% for S100 protein. Renal cell carcinomas were uniformly reactive for cytokeratin and variably reactive for S100 protein (5%) and vimentin (25%). The biopsy in our patient had a positive express on vimentin, S100 protein and epithelial membrane antigen adequate to this study.

Signs and symptoms often appear to be result of discogenic or non-specific pathology, resulting in delayed diagnosis.⁹ Surgical resection, radiation therapy, and chemotherapy are the current proposed therapy modalities for the spinal chordomas. The main problem in the treatment of chordoma is local recurrence and occurs as soon as 2 to 4 years after initial treatment. Chordomas invade adjacent structures, and may metastasize in 3-60% of cases.^{1,5,10-13} Lumbar chordomas have higher metastatic rates than in any other location,^{9,14-16} however, local aggressiveness rather than metastases is more likely to cause death and disability.¹⁶ Aggressive surgical resection and stabilization with rigid fixation if necessary affords the best opportunity for prolonged survival.¹⁴ These tumors are highly resistant to radiotherapy or chemotherapy. The effectiveness of radiotherapy is related to dose applied. These tumors are radio-resistant to the standard dose of radiation which is less than 70 Gy.² A much higher dose of radiation can be delivered by proton and photon radiation has been reported to achieve a significant success rate in terms of local disease control.¹⁷

Lumbar spine is an uncommon localization for chordoma. Modern imaging technique, that make possible diagnosis of spinal chordoma preoperatively. In patients with radiographic appearance of a lytic lesion with paravertebral mass of the mobile spine, chordoma should be in mind in the differential diagnosis.

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