

Case Report

Combined Anterior and Posterior En Bloc Vertebrectomy for Lumbar Chordoma

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Chordoma is a rare bone tumor derived from remnants of the notochord. The majority of chordomas involve the sacrum or skull base. We report a rare case of a L4 vertebral body chordoma treated with anterior en bloc vertebrectomy and posterior stabilization. No tumor recurrence was observed at the 5 year follow-up examination.

KEY WORDS : Lumbar chordoma · Anterior and posterior en bloc vertebrectomy.

INTRODUCTION

Chordoma is a rare bone tumor that is believed to be derived from remnants of the notochord^{1,4,6}. Chordomas usually grow slowly over a period of many years, often recur locally, and metastasize late in their course. Thus, chordoma is thought to be a clinically malignant bone tumor⁹. Chordoma may occur anywhere along the spinal column, but it usually involves the sacrum or skull base presenting in late middle-age and older adults^{3,7}. Chordomas of the vertebrae are known to be more locally aggressive, more likely to metastasize and have a poorer 5-year survival rate than sacral and clival lesions⁸. The rationale of en bloc resection for this tumor is to reduce local recurrence rate and improve survival rate. We report a rare case of solitary lumbar chordoma that was treated with staged anterior and posterior en bloc vertebrectomy with a review of the literature.

CASE REPORT

A 43-year-old woman presented with a 3-month history of pain in her left anterior thigh and calf. Neurologic examination revealed an absent left knee jerk, with reduced

sensation in the left L4 dermatome. The straight-leg-raising test showed restriction of motion on the left side (90°/30°). However, there was no motor weakness or sphincter disturbance. Blood tests, including inflammatory markers, were within normal limits. Simple lateral radiographs showed bony destruction of the anterior part of the L4 body and osteoblastic change (Fig. 1A). Magnetic resonance imaging (MRI) revealed an epidural mass compressing the dural sac and left L4 nerve root (Fig. 1B). Heterogeneous enhancement was noted with gadolinium administration. (Fig. 1C, D). Lumbar spine computed tomography (CT) revealed both bony destruction and osteoblastic change in the L4 body (Fig. 1E). Technetium bone scanning showed focally increased tracer uptake in the isolated L4 body (Fig. 1F). With the diagnosis of metastasizing tumor of unknown origin or primary bone tumor, transpedicular biopsy was performed at L4, and histological examination revealed vacuolated cells with intracytoplasmic mucous droplets, consistent with physaliphorous cells of chordoma (Fig. 2). Immunohistochemistry was positive for cytokeratin staining and EMA staining (Fig. 3). Based on the histological findings of chordoma, staged anterior and posterior en bloc vertebrectomy with pedicle screw fixation and fusion was planned. The L4 vertebral body compressing the dural sac and nerve root was removed anteriorly and was replaced with mesh packed with iliac bone chips. Anterior approach was followed by posterior resection of the total posterior element of L4. Pedicle screw instrumentation and posterolateral

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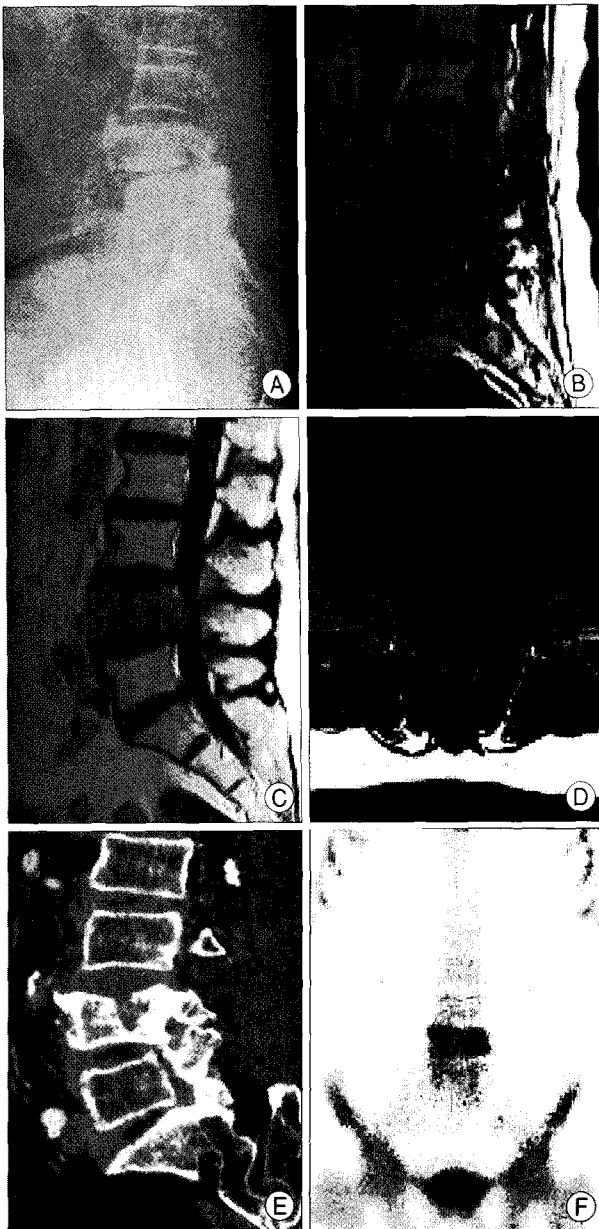


Fig. 1. Forty-three-year-old woman with chordoma. A : Simple lateral radiograph revealing an osteolytic lesion in the anterior part of the L4 body. B : T1-weighted magnetic resonance image shows low signal intensity involving the L4 body. C and D : Gadolinium-enhanced magnetic resonance images reveal a heterogeneously enhanced mass compressing the dural sac and nerve root. E : Computed tomography shows both bony destruction and osteoblastic change at the L4 body. F : Isotope bone scan shows increased tracer uptake in the isolated L4 body.

fusion with autogenous iliac bone and local bone chips were performed bilaterally at L3 and L5 level at 2 weeks after anterior approach (Fig. 4). After surgery, the patient regained normal sensation, and the pain was significantly relieved. There was no restriction on the straight-leg-raising test. At the 5 year follow-up examination, her neurological status had not changed, and she was able to keep her job without any neurologic deficits or pain.

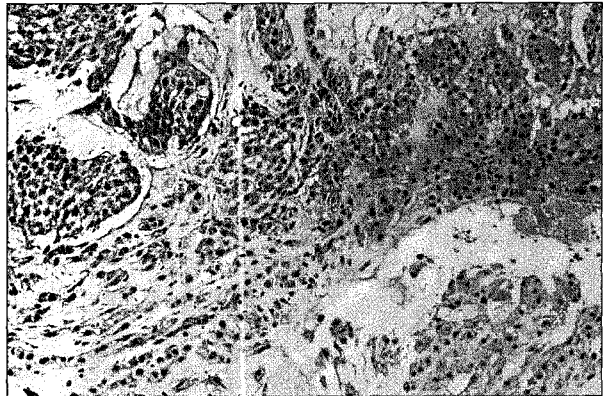


Fig. 2. Histologic examination showing myxoid tissue containing strips and ribbons of cohesive, vacuolated cells (H&E $\times 100$).

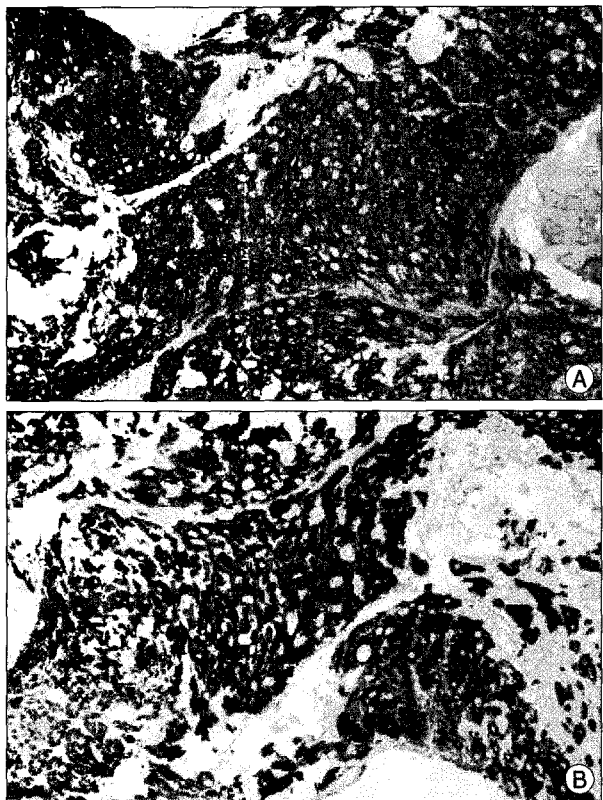


Fig. 3. Immunohistochemistry is positive for cytokeratin staining and EMA staining. A : EMA staining. B : Cytokeratin staining.

DISCUSSION

Chordoma is thought to be a primary bone tumor that occurs with a predilection for the clivus and sacrum^{1,3,6,9}. This anatomical distribution supports the generally accepted theory that it arises from notochordal remnants. As mentioned above, it is usually found in the clivus and sacrum and infrequently in the lumbar spine¹¹. When adequate excision is not possible, radiation therapy may provide at least a short-term benefit. Radiation therapy is also used for recurrence^{2,10}. So, it is generally agreed that

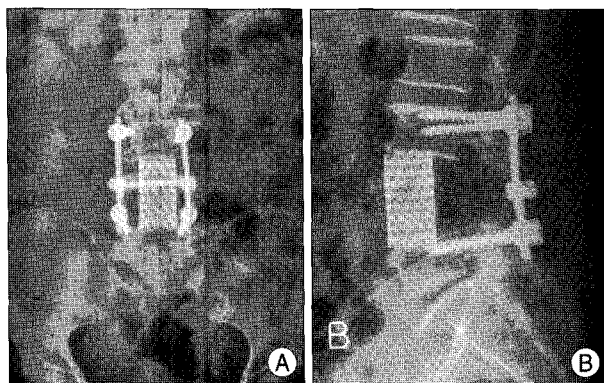


Fig. 4. Anterior posterior (A) lateral (B) radiographs 5 years after surgery show mesh packed by iliac bone chips and pedicle screws insertion at L3 and L5. There is no evidence of local recurrence or metastasis.

complete surgical excision of the tumor is the only curative procedure⁶). As has been recognized so far, recurrence rates after any treatments of chordomas have been high and unsatisfactory^{1,3,4}). Surgical resection has become more aggressive in recent years evolving from intralesional debulking to total resection to reduce local recurrence rate⁴). However, in most cases, chordomas are very difficult to excise completely⁵). The adjacent vital neural and vascular structures often result in oncologically incomplete and unsatisfactory treatment. A combined anterior and posterior procedure allows for an extralesional, marginal resection of the tumor and involved vertebra. En Bloc resection of primary malignant bone tumors including chordoma increases the survival rate and is a crucial part of the treatment regimen. Adjuvant radiation and chemotherapy have not required after this en bloc resection. Tomita et al.¹²) described an extensive experience with en bloc spondylectomy from a single posterior approach for patients with solitary spinal metastatic lesions. However, this aggressive posterior operation carries the risk of injury to the major ventrally located vascular structures, which are not well visualized using a purely posterior approach. Treatment outcome is also significantly influenced by the size and site of the chordoma. Chordomas in the vertebral bodies appear to be more aggressive than those arising from the clivus or the sacrum. Metastasis has been reported in 80% of vertebral body chordomas and in 43% of all chordomas⁷). Mobile segment chordomas have a 50% survival rate at 5 years and a 28% survival rate at 10 years. On the other hand, sacral chordomas are reported to have a 5-year survival rate of 86%⁸). A combined anterior and posterior procedure allows for an extralesional, marginal

resection of the tumor and involved vertebra. En bloc spondylectomy by combined anterior and posterior approach enables wide or marginal resection of malignant lesions of the spine that have known or suspected tumor involvement. Needless to say, preoperative careful planning should be considered adequately. The other major point after en bloc resection is reconstructive surgery. Immediate spinal stability must be provided using appropriate replacements or bone grafts with internal fixation.

CONCLUSION

In summary, we describe a rare case of lumbar chordoma. Despite the poor published results of mobile segment chordomas, it can be potentially curative using an "en-bloc" resection. Extensive wide removal using a staged anterior and posterior approach followed by a stable reconstruction may achieve an excellent clinical course.

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