Follow-Up Evaluation of Resected Lumbar Vertebral Chordoma Over 11 Years

A Case Report

Ken Y. Hsu, MD,* James F. Zucherman, MD,* Norval Mortensen, MD,*
James O. Johnston, MD,† and John Gartland, MD*

Study Design. A case report with an 11-year follow-up assessment after resection and reconstruction for lumbar chordoma is given. The literature relevant to this topic is reviewed.

Objectives. To report the long-term outcome in a case of lumbar chordoma, to review the literature on vertebral chordoma, and to outline the rationale for surgical resection in such cases.

Summary of Background Data. Chordoma is a malignant bone tumor that grows slowly, often recurs locally, and metastasizes late. Although different treatment approaches exist, including radiation and surgery, the only curative treatment is early and complete surgical excision of the tumor. Immediate spinal stability must be achieved with appropriate replacement or bone graft with rigid fixation.

Methods. The 11-year follow-up evaluation of a 42-year-old woman with L3 and L4 vertebral body chordoma treated with complete removal, femoral shaft allograft replacement, fusion, and rigid metal fixation is reported. The patient was observed with serial physical examinations, radiographs, and laboratory studies over 11 years.

Results. At this writing, 11 years after the resection of the L3 and L4 chordoma, the patient is asymptomatic without evidence of recurrence or metastasis.

Conclusions. As reported, vertebral chordomas are not curable, but the authors' experience contradicts this. The surgeon should aim at a wide, or at least a marginal, excision followed by a stable reconstruction. [Key words: allograft, chordoma, fusion, lumbar spine, rigid fixation, surgical resection] Spine 2000;25:2537-2540

Chordoma is a malignant bone tumor believed to be derived from the remnants of the notochord.2,10,11 Usually, it grows slowly over a period of many years, often recurs locally, and metastasizes late in its course to the lungs, liver, brain, bone, and soft tissues.1,11 Typically, chordoma is seen in late middle-age and older patients. It may occur anywhere along the spinal column, but it is seen more commonly in the clivus and the sacrum and less often in the vertebrae.8,9,13 On gross inspection, a chordoma may be soft or firm, and often appears lobulated. Although there are variable histologic patterns, a chordoma characteristically consists of mucin-producing vacuolated physaliphorous cells.7,10,11 Although it metastasizes late, it is a difficult tumor to treat because of its location close to vital structures.11 Compression of neural tissues and invasion of the brain or spinal canal may result in serious neurologic complications.

Various treatment approaches have been attempted, including isolated low-dose or high-voltage radiation therapy, combined radiation and surgical excision, and surgical excision alone.1-4,6,8,10,12,13 Surgical treatment has become more aggressive in recent years, evolving from intraslesional partial excision to en bloc resection, as documented by Boriani et al1 over a 45-year period in their treatment of 21 cases of chordoma located in the spine above the sacrum. It is generally agreed that complete surgical excision of the tumor is the only curative procedure.4,10 When adequate excision is not possible, radiation therapy may provide at least short-term benefit.4,6 Radiation therapy also is used for recurrence. The radiation doses are delivered with megavoltage equipment.1,4,6,12

The survival rate appears to be affected more by local tumor progression than by metastasis.4,11 Chordomas lead directly to death in 63% of cases, with a 50% survival rate at 5 years3,4,13 and a 28% survival rate at 10 years.4,6 Treatment outcome is significantly influenced by the size and site of the chordoma.4,11 Chordomas found in the vertebral bodies appear to be more malignant than those arising in the clivus or the sacrum.8 Metastasis have been reported in 80% of the vertebral body chordomas, as compared with an overall rate of 43%, considering all chordomas.8

In general, vertebral chordomas are so uncommon that the authors are forced to report long-term follow-up evaluation of such tumors on a case-by-case basis. They believe that this case report on the 11-year follow-up evaluation after surgical resection of a lumbar vertebral chordoma should be documented to define further the role of such difficult treatment efforts.

Case Report

A 42-year-old woman began having increasing low back pain after a fall down the stairs in April of 1986. Her back pain progressed for the next 4 months. Lumbar spine radiographs, Computed tomography (CT) myelogram, and bone scans were unremarkable. Laboratory studies showed only an abnormal Westergren sedimentation rate of 30. By October of 1987, the patient had begun to have bilateral anterior thigh pain.

From *St. Mary's Hospital and Medical Center, San Francisco, California, and the †Department of Orthopaedic Surgery, University of California San Francisco.
Acknowledgment date: October 5, 1999.
First revision date: December 13, 1999.
Acceptance date: January 18, 2000.
Device status category: 1.
Conflict of interest category: 12.
By March of 1988, the pain had become disabling, despite prolonged bed rest. Magnetic resonance imaging (MRI) was performed at this time, showing soft tissue lesions in the L3 and L4 vertebral bodies (Figure 1). An extensive search for metastasis was negative. Vertebral body needle biopsies of L3 and L4 demonstrated only necrotic bone, and repeat needle biopsy 1 month later showed no tumor. Finally, an open biopsy in May of 1988 demonstrated vacuolated cells with intracytoplasmic mucous droplets, consistent with physaliphorous cells of chordoma (Figures 3A and 3B).

In July 1988, surgical resection of the L3 vertebral body and the superior 75% of the L4 vertebral body was performed (Figure 2). The vertebral bodies were removed anteriorly and replaced with a femoral shaft allograft 14 cm long, which was fixed with an AO plate and Steinmann pin anteriorly. This was followed by posterior resection of the posterior elements of L3 and L4, and posterolateral fusion with autogenous iliac bone graft using variable spinal plate and pedicle screw fixation from L2 to S1 bilaterally (Figures 4A and 4B). The diagnosis of chordoma in L3 and L4 was confirmed with permanent section analysis.

After surgery, the patient was found to have weakness of the left foot dorsiflexion. She had a complicated postoperative course, including difficult fluid volume management, metabolic acidosis, and a brief episode of pulmonary edema, which resolved with diuresis. Oxycodeone was used for pain control for 1 year after surgery, and hydrocodeone for 5 years subsequently.

By 1991, she was able to walk several miles at one stretch. The patient eventually was able to wean herself from all pain medications by 1994. She had no back or lower extremity pain. Follow-up radiographs in August 1999 (Figures 5A and 5B) showed solid fusion from L2 to S1 with incorporation of the femoral shaft allograft. Fractures of one L2 pedicle screw, one anterior AO screw at L5, and one Steinmann pin were noted, but did not affect stability or the final surgical outcome. At this writing, 11 years after surgery, there is no radiologic evidence of degeneration above the solid four-level fusion, nor evidence of local recurrence or metastasis.

**Discussion**

It is very uncommon to have chordoma of two neighboring vertebrae at presentation as in the reported case. The question arises whether this was the result of hematogenous spread, multifocal development of chordoma, or direct spread across the disc. Nevertheless, this is a case report on removal of chordoma involving two adjacent lumbar vertebrae.

The authors believe that the only curative treatment for chordoma is an early and complete surgical excision of the tumor. Aggressive and thorough resection is important because survival is related more to local progression of the tumor than to metastasis. However, in most cases, chordomas are very difficult to excise completely. The size of these tumors and the challenge of operating around the spine without injuring the adjacent vital neural and vascular tissues often result in oncologically unsatisfactory treatment. With a chordoma confined to the vertebral body of the lumbar spine, such as that found in the reported case, surgical excision is possible using anterior and posterior approaches.

**En bloc** resection of the tumor is achieved anteriorly by freeing the vertebral body from the pedicles. The remainder of the pedicle is then removed posteriorly, along with the other posterior elements that have known or suspected tumor involvement. If extraosseous soft tissues are involved, wider resection must be considered. If the psoas muscles are involved, they may need to be resected.
Figure 3. A, Photomicrograph of the L3 lesion, with bubbly physaliphorous cells characteristic of chordoma (magnification ×250). B, Higher-power (×500) view showing the photomicrograph of the L3 lesion in which the physaliphorous cells can be seen.

Risk–benefit assessment and careful planning must be performed before such surgical decision preceding surgery.

The second major problem encountered in the surgical management of vertebral chordoma is the reconstructive surgery. Immediate spinal stability must be provided using appropriate replacements or bone graft with internal fixation. The authors elected to use a femoral shaft allograft, which has adequate strength and size in the lumbar spine. This was augmented with posterolateral autograft fusion using rigid metal fixation. The stainless steel pedicle screw system was used posteriorly and an AO plate anteriorly. Although radiation therapy was considered, this adjuvant treatment was not used after an adequate surgical resection. The recommended high-voltage radiation may have interfered with the spinal fusion and incorporation of the allograft. The postoper-

Figure 4. A, Radiographs immediately after surgery with anteroposterior view. B, Lateral radiograph showing the 14-cm-long femoral shaft allograft from L2 to L4 fixed with an AO plate and Steinmann pin anteriorly, and with variable spine plates and pedicle screws posteriorly from L2 to S1 with posterolateral fusion.
ative infection rate also may have been affected by radiation treatment.

References


Address reprint requests to
Ken Y. Hsu, MD
St. Mary's Spine Center
One Shadrer Street
San Francisco, CA 94117