Extraosseous Spinal Chordoma: Radiographic Appearance

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Summary: The authors describe a chordoma arising within the soft tissues of the spinal canal (rather than from a vertebral body) in a 6-year-old boy. The lesion was entirely extradural as shown by CT and MR; this was confirmed at surgery.

Index terms: Spine, neoplasms; Chordoma; Spine, magnetic resonance; Spine, computed tomography

Chordomas are rare neoplasms that arise from remnants of the primitive notochord. Chordomas are uncommon in the pediatric age group, with only 56 cases cited in the literature (1).

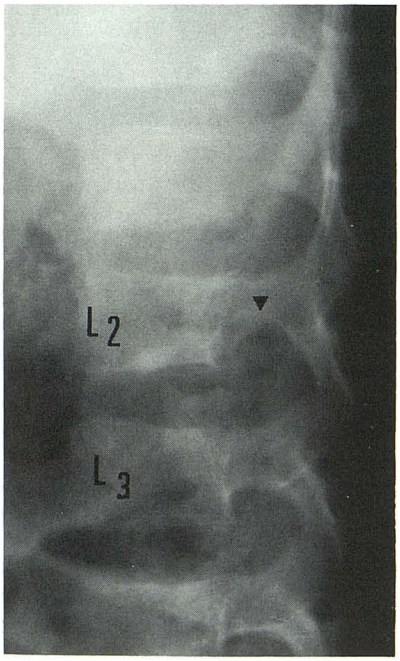
Only 15% of all chordomas arise in a vertebral location (2), and of those, all have an osseous origin (3). We report a case of extraosseous spinal chordoma in a 6½-year-old boy, and discuss its unusual appearance on plain radiography, computed tomography (CT), and magnetic resonance (MR) imaging.

Case Report

A 6½-year-old boy presented with a 2-month history of isolated right buttock pain and spinal rigidity of the dorsolumbar area. Prior medical history was unremarkable and the neurologic examination was normal.

Spinal radiographs revealed an enlargement of the right interpediculate space at the L2-L3 level due to scalloping of the inferior margin of the right pedicle of the L2 vertebra (Fig. 1).

CT demonstrated an extradural mass that filled the right side of the spinal canal at the L2-L3 level, extrinsically



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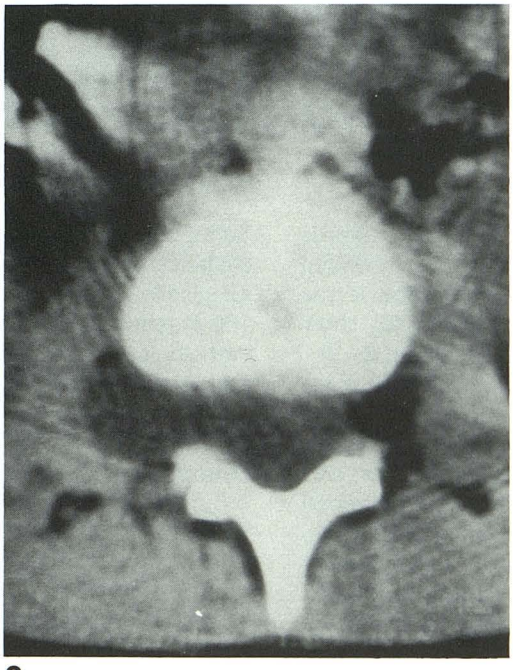

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Fig. 1. Spinal radiograph demonstrates enlargement of interpediculate space at L2-L3 (*arrowhead*) and scalloping of inferior marging of L2 pedicle.

Fig. 2. CT with contrast injection demonstrates the nonenhancing mass.

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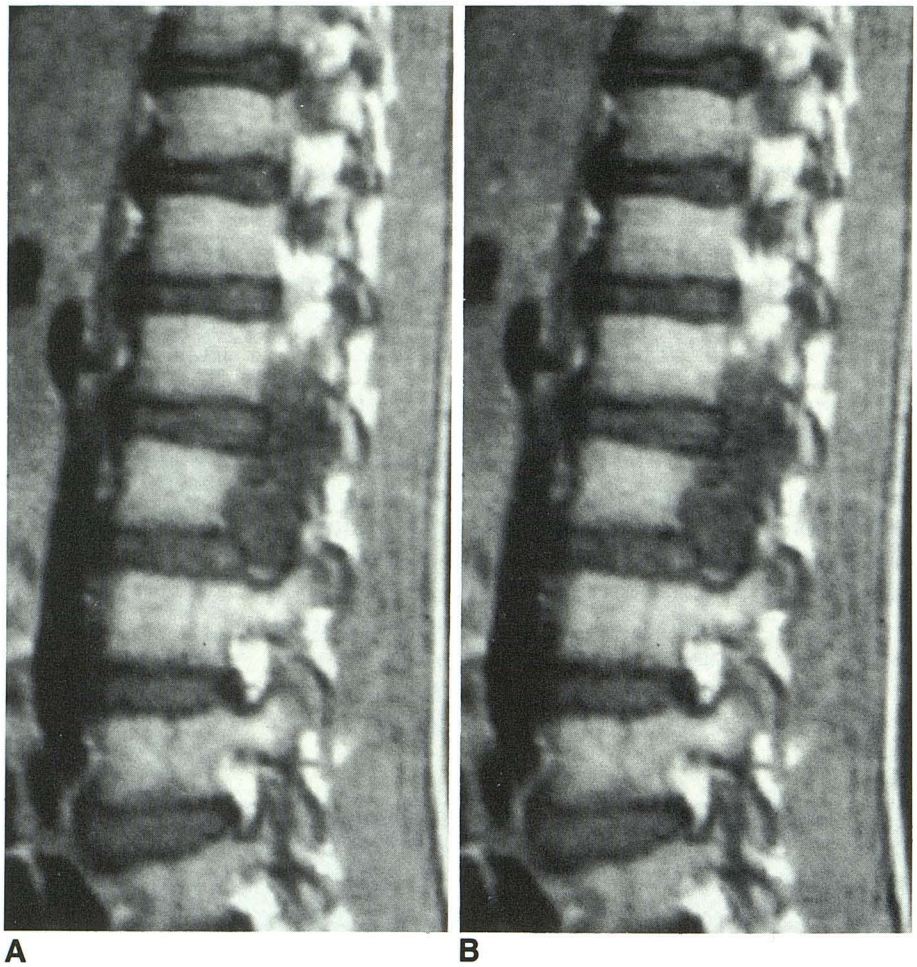

Fig. 3. *A*, Sagittal MR of the spine, T1-weighted images 480/26. A mass is filling the spinal canal at L2-L3 with extension into superior aspect of L3-L4 space.

B, T1-weighted image 480/26 with gadolinium shows absence of contrast enhancement of the lesion after gadolinium injection.

compressing and markedly displacing the dural sac towards the left. The lesion had a dumbbell appearance, extended laterally through the enlarged interpediculate space. It was homogeneous and hypodense (25 HU) and did not show evidence of enhancement after contrast injection (Fig. 2).

MR imaging (0.5 T) demonstrated an extradural mass filling the spinal canal at the L2-L3 level and extending into the superior aspect of the L3-L4 space with evidence of posterior scalloping of the L2 pedicle. The mass was homogeneous in signal and isointense to the intervertebral disks on both the T1 480/26/2 (TR/TE/excitations) (Fig. 3A) and T2 1800/120. There was no contrast enhancement following gadolinium injection (Fig. 3B). A cystic neurinoma or arachnoid cyst were thought to be responsible for the findings.

At surgery, a completely extradural, extraosseous, well-defined tumor with poor vascularization was found.

Pathologic examination was diagnostic of chordoma. Grossly, the tumor was gelatinous and did not demonstrate macroscopic evidence of septations or cartilaginous foci. Microscopically, the tumor was extremely well encapsulated by a thick fibrous capsule and lobulated by fibrous strands. The lobules were composed of vacuolated physaliphorous cells containing variable amounts of intracytoplasmic mucin, embedded in lakes of extracellular mucin. Some cells had a signet ring appearance, but there was no evidence of mitotic activity or pleomorphism. Immunohistologically, there was a positive staining reaction with S 100 protein, as well as with cytokeratin (KL1) and vimentin. Pathologically, all the features were diagnostic of a typical (rather than chondroid) chordoma (4).

Discussion

Chordoma is a rare tumor that accounts for only 1%–2% of primary malignant bone tumors (3, 4). The peak incidence for these neoplasms is in the sixth decade of life (2) and, as such, they have been uncommon in the pediatric age group. Overall, the tumor tends to occur at either end of the axial skeleton with pediatric chordomas arising most frequently from the clivus in the region of the spheno-occipital synchondrosis (2).

Spinal chordomas are thought to develop from ectopic notochordal elements within the bodies, or, more rarely, from within the posterior elements of the vertebrae (3). Given their osseous origin, it is not surprising that their radiographic

hallmark has classically been one of bone destruction (5), and, more specifically, sclerotic vertebral changes (3, 5, 6). Chordomas have been separated into two pathologic subsets, typical chordomas and chondroid chordomas, each with a different MR appearance. Chondroid chordomas have shorter T1 and T2 relaxation times than typical chordomas because in the chondroid type, watery, gelatinous matrix is replaced by cartilaginous foci (4).

To our knowledge, there have been no reports of completely extradural extraosseous vertebral chordomas. However, two cases of intradural extraosseous chordomas (pontine and clival) have been reported in adults (4, 7). In both cases, the chordoma was postulated to arise from ectopic intradural notochordal nodules known as ecchondrosis physaliphora. Given the embryologic intimacy of the notochord and the vertebral column, and the documented presence of ectopic notochordal elements throughout the spine, one may postulate an ectopic extradural extraosseous notochordal remnant as the etiology for this unusual chordoma.

The radiographic differential diagnosis for extraosseous soft-tissue mass in the pediatric age group is quite narrow. Neurofibromas, neuroblastomas, and schwannomas may all potentially present as a dumbbell tumor of the spine with erosive bony changes and, as such, are the most difficult to differentiate from this atypical chordoma. The relative avascularity of the lesion on contrast studies tend to favor the diagnosis of chordoma, inasmuch as the other lesions tend to enhance on contrast studies.

Meningoceles are easily distinguished by myelography. Dermoid cysts are diagnosed when the CT and/or MR studies demonstrate a fatty content. Although, normally, arachnoid cysts are also easily distinguished from chordomas on the basis of their purely extraosseous appearance, this was clearly more difficult in this unusual case of extraosseous chordoma.

In summary, this pediatric patient had a unique presentation for chordoma in the vertebral region. This tumor was completely extradural with both intra- and extraforaminal components, but did not involve the bone.

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