

Epidural Chordoma of the Cervical Spine with Secondary Bone Involvement

Grace Kalish, Brian P. Rubin, Felix S. Chew, Michael L. Richardson

A 53-year-old man presented with cervical myelopathy. magnetic resonance imaging (MRI) revealed a predominantly extraskelatal, extradural lesion extending along the posterior aspects of the C2 to C5 vertebral bodies, with greater than 60% spinal canal compromise and severe cord compression. Bone involvement was present, but was thought to be secondary. Based on histopathology and immunohistochemical stains, the final pathologic diagnosis was chordoma. The lesion was treated with embolization, surgical resection, and proton beam radiotherapy, and there was no evidence of recurrence or metastasis after five years.

Case Report

A 53-year-old man, in prior good health, presented with right lower extremity weakness and difficulty walking. One year earlier, the patient began noticing bilateral hand numbness and finger paresthesias. Additional complaints included inability to abduct his right arm and paresthesias along the skin overlying the spine. Physical exam was significant for profound deltoid weakness, diminished deep tendon reflexes in the right leg, and bilateral clonus with dorsiflexion of the ankles.

MRI of the cervical spine revealed a homogeneous T1 hypointense and T2 hyperintense lesion. The lesion was predominantly extraskelatal and extradural, and extended along the posterior aspect of the vertebral bodies from C2 to C5 with at least 60% spinal canal compromise and severe cord compression (Fig. 1). There was heterogeneous

enhancement noted within the lesion following intravenous gadolinium. There was some involvement of the posterior aspects of the C3 and C4 vertebral bodies along the right side. The right neural foramina were involved from C3-C4 to C5-C6 and there was stenosis of the right transverse foramina from C2 to C4. There was no apparent invasion of the spinal cord.

The patient underwent angiographic evaluation and neuroembolization of the right anterior cervical artery and right deep cervical artery. The patient then underwent a C2-C6 laminectomy and tumor resection. The mass was extradural with a capsular component encasing a gelatinous tumor. The tumor and its capsule were resected with sparing of the ventral portions surrounding the vertebral artery at the C4-5 level. Frozen sections at the time of surgery suggested a benign lesion of unknown etiology, possibly a nerve sheath tumor.

At pathology, the lesion infiltrated bone and soft tissue and was composed of islands of epithelioid cells embedded in a prominent myxoid matrix (Fig. 2A). The lesional cells contained abundant eosinophilic cytoplasm, round to oval shaped nuclei, fine chromatin, and small, inconspicuous nucleoli (Fig. 2B). Focally, the cytoplasm of the cells was extremely vacuolated and there were occasional multivacuolated physaliferous cells (large vacuolated, glycogen or mucin-containing cells) (Fig. 2C & 2D). There was occasional nuclear pleomorphism but mitotic activity was minimal. Necrosis was not present. Immunohistochemical studies revealed the lesion to be positive for cytokeratins and S-100 protein (Fig. 2E). The final pathologic diagnosis was chordoma. Because of the predominance of the extraosseous extradural component and the limited, superficial involvement of bone, we believe that this case repre-

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Abbreviations: MRI, magnetic resonance imaging, STIR, short tau inversion recovery

Grace Kalish (Email: kalish@u.washington.edu), Brian Rubin, Felix Chew, and Michael Richardson are from the Department of Radiology, University of Washington School of Medicine, Seattle, WA, United States of America.

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Epidural Chordoma of the Cervical Spine with Secondary Bone Involvement



Figure 1. Extraskeletal chordoma of the cervical spine.

A. Sagittal T2-weighted MRI in the midline shows a lobulated mass along the posterior aspect of C1-C4 with high signal. The mass does not appear to invade the vertebral bodies. The cervical cord is severely compromised. There is also incidental degenerative disc disease with loss of height at C3-C4, and mild retrolisthesis of C3 over C4.

B. Sagittal T1-weighted post-gadolinium MRI shows mild enhancement within the mass.

C. Axial short tau inversion recovery (STIR) MRI following gadolinium injection shows the large epidural mass occupying the right side of the spinal canal, compressing the cervical cord to the left. The lesion extends into the right neural foramen.

Epidural Chordoma of the Cervical Spine with Secondary Bone Involvement

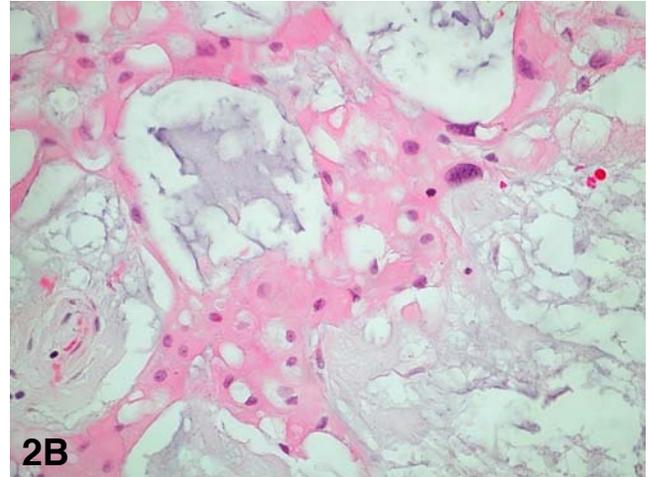
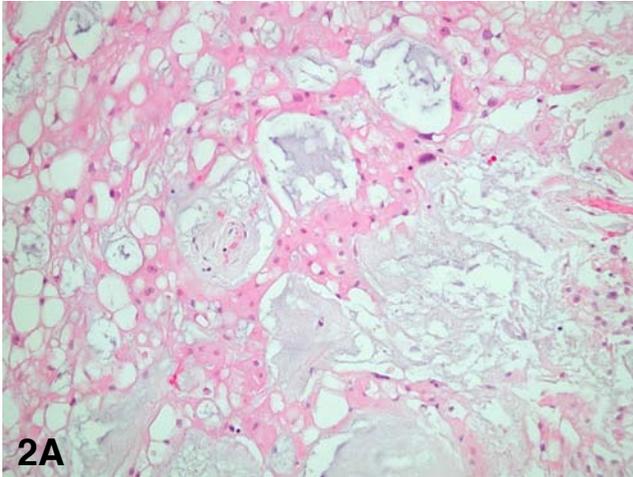


Figure 2. Extraskelatal chordoma of the cervical spine. **A.** Photomicrograph (200x, hematoxylin and eosin) - Some areas of the neoplasm revealed sheets of epithelioid cells admixed with pools of mucin. Some of the cells contain vacuolated cytoplasm. **B.** Photomicrograph (400x, hematoxylin and eosin) - Higher power of the area from part A showing sheets of cells with brightly eosinophilic cytoplasm and some vacuolization. Note the focal cytologic pleomorphism.

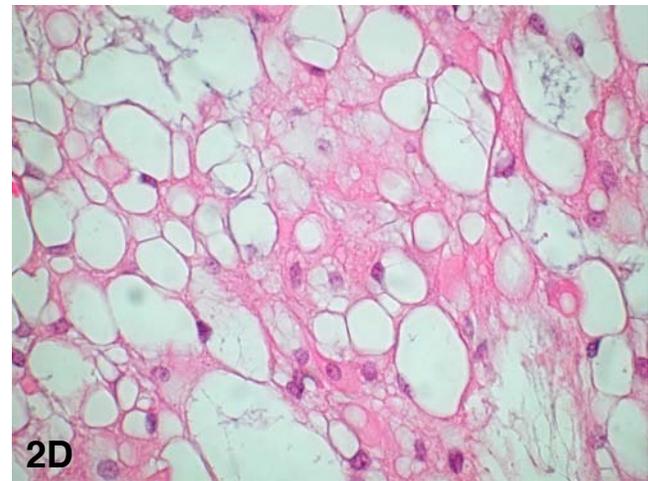
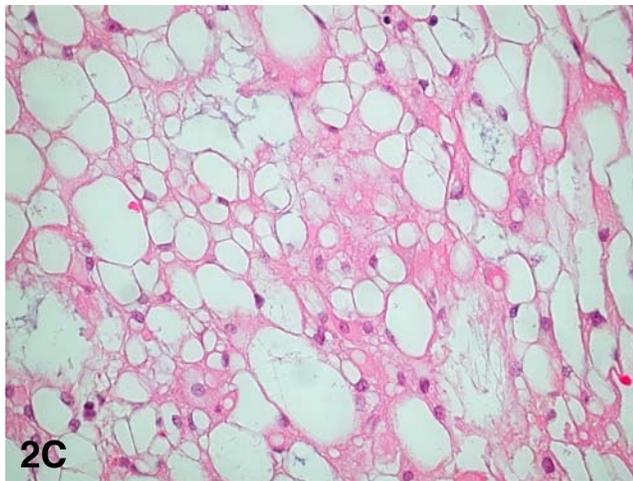


Figure 2. Extraskelatal chordoma of the cervical spine. **C.** Photomicrograph (200x, hematoxylin and eosin) - Other areas of the neoplasm contained cells with extensive cytoplasmic vacuolization. **D.** Photomicrograph (400x, hematoxylin and eosin) - Higher power view of the area from part C revealing multivacuolated physaliferous cells in areas with extensive cytoplasmic vacuolization.

Epidural Chordoma of the Cervical Spine with Secondary Bone Involvement

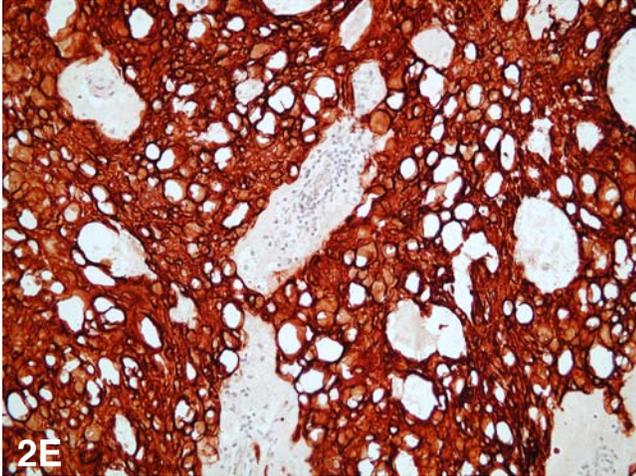


Figure 2E. Extraskelatal chordoma of the cervical spine. Photomicrograph (200x) - Immunohistochemical study for cytokeratin revealed the cells to be extensively positive for cytokeratin. The lesional cells were also positive for S-100 protein (not shown).

sents a chordoma of extraosseous extradural origin with secondary direct extension to bone.

The patient subsequently underwent adjuvant proton beam radiotherapy. There has been no evidence of recurrence or metastasis in five years of follow-up.

Discussion

Chordomas are rare tumors that arise from the embryologic remnants of the notochord. Most chordomas occur in the sacrococcygeal (50%) and sphenoccipital regions (35%), with a much smaller percentage occurring in the cervical, thoracic, and lumbar spine (15%) (1). Of those that arise in the mobile segments of the spine, approximately 50% will be in the cervical region (2). Chordomas comprise 1 to 5% of all primary malignant bone tumors (3). Most patients with chordomas present in middle adulthood, but chordomas have been described in a wide age range from childhood to senescence (3).

The notochord represents the embryological origin of the axial skeleton. It is surrounded by cartilage which eventually ossifies into the clivus, vertebral bodies, and sacrum (4). With ossification of the surrounding cartilage, extrusion of the notochord takes place, eventually forming the nucleus pulposus. Occasionally, extrusion can occur in aberrant locations, and smaller remnants of the notocord (notocord rests) may persist in other locations along the cranio-spinal axis. It is believed that most notochordal rests occur within osseous structures, but complete extrusion into the extradural or intradural spaces would allow for growth without bony involvement.

Histologically, chordomas are composed of mucinous fatty material, which is surrounded by a fibrous pseudocapsule that results in a lobulated radiographic appearance.

They are well demarcated from adjacent soft tissue structures, but may be associated with local bone destruction (3-4).

MRI features of chordomas include hypointense signal in T1 weighted images, with moderate-to-high signal intensity on T2 weighted images, owing to high fluid content. Heterogeneous appearance is frequent on T1 and T2 weighted images, explained by high protein content or high ferritin from old blood products, respectively. Enhancement patterns are variable (4-6). Vascular encasement is a common finding, both intra and extra-cranially. However, chordomas rarely cause arterial stenosis in the spine (7).

Chordomas arising in the cervical spine have been reported in very small series (8) or as individual cases. Many of them arise within bone or involve bone (9-17). Multiple-level involvement is common, as is an extraosseous component (9-17). Unusual presentations include morphologic resemblance to nerve sheath tumors (18-21), Horner's syndrome (22), and oral cavity mass (23). Involvement of the upper levels may be more common than the lower levels (8-23). The challenge of surgical resection with adequate margins has been emphasized by many authors (24-25).

There have been a few case reports of extraosseous, intradural cervical chordomas (26-30). Intradural notochordal remnants may occur anywhere in the spinal axis, but are more common in the retroclival space, and have been reported to occur in up to 2% of autopsy specimens (31). Usually, these remnants are associated with a thin stalk that penetrates the dura. These were initially described by Virchow as "eccordosis physaliphora." Eccordosis physaliphora are currently thought to be distinct from chordomas, in that they are clinically silent, and represent developmental vestiges rather than tumors (32). Histologically, however, chordomas and eccordosis physaliphora are almost identical. Whether chordomas arise from eccordosis physaliphora is still under debate.

There have been a few case reports of extraosseous, extradural cervical chordomas (33-35). In these cases, the lesions were located in the loose areolar tissue of the spinal epidural space. Jallo et al. have developed a classification of spinal chordomas, based on location and osseous involvement (35). Type I are osseous and extradural and comprise the majority of chordomas. Type II are extraosseous and extradural, as may have been this case. Type III chordomas are osseous and intradural, and Type IV are extraosseous and intradural (Table 1). Type III lesions would presumably arise in bone and involve the dura secondarily by direct extension. Jallo hypothesizes that the classification serves as a prognostic indicator, because extraosseous location (Type II and Type IV) is associated with favorable resectability and therefore lower rates of recurrence. The extreme rarity of these cases makes this hypothesis difficult to test.

Epidural Chordoma of the Cervical Spine with Secondary Bone Involvement

TABLE 1. Classification of Spinal Chordomas Based on Anatomic Location

	Extradural	Intradural
Osseous	Type I	Type III
Extraosseous	Type II	Type IV

Source: [Jallo et al.](#) (35)

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Epidural Chordoma of the Cervical Spine with Secondary Bone Involvement

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