Case Report

A 53-year-old man presented with cervical myelopathy. Magnetic resonance imaging (MRI) revealed a predominantly extraskeletal, 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.

Epidural Chordoma of the Cervical Spine with Secondary Bone Involvement

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

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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.
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Figure 2. Extraskeletal 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. Extraskeletal 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.
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 sphenooccipital 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 notochord (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.
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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