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Case Report

Myxopapillary Ependymoma with Extensive Sacral Destruction: CT and MR Findings

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Abstract: There have been few reports documenting primary myxopapillary ependymomas in the sacrococcygeal region that result in extensive involvement of the sacrum. We present a 21-year-old man whose CT and MR findings showed massive bony destruction of the sacrum and a large lobulated soft tissue mass. Myxopapillary ependymoma should be included along with giant cell tumor, chordoma, and aneurysmal bone cyst in the differential diagnosis of a destructive osteolytic sacral lesion. Index Terms: Ependymoma—Spine, neoplasms—Magnetic resonance imaging—Computed tomography.

Ependymomas account for 60% of the tumors of glial origin in the spinal cord (1,2), and comprise 90% of primary tumors in the filum terminale and cauda equina (2–4). The myxopapillary subtype is the most common (5). Myxopapillary ependymoma was first described as a separate subtype of ependymoma in 1932 when Kernohan recognized four categories of this neoplasm (myxopapillary, epithelial, cellular, ependymoblastoma, or papillary choroiodeum) (1). Histologically, myxopapillary ependymoma consists of regions of mixed solid cellularity and acellular myxoid degeneration arranged in papillary projections with epithelioid cells surrounding the vasculature (1). Many reports have documented this tumor intraspinally in the cauda equina/filum terminale region (6–14) and extraspinaly, including cases with presacral and postsacral, and even subcutaneous involvement (2,4,5,13,15–23).

The myxopapillary ependymoma has been known to metastasize (5–7,14,17–19,24). Few reports, however, have shown a primary myxopapillary ependymoma of the sacrococcygeal area without systemic metastases that result in extensive bony destruction. We describe the CT and MR findings of a large myxopapillary ependymoma that presented as a primary lesion of the sacrum.

CASE REPORT

A previously healthy 21-year-old man sustained a twisting injury of his back while playing handball 3 months prior to presentation. He subsequently noted pain and a "knot" in his lower back; however, he did not seek medical attention at that time. After reinjury while playing rugby, he was evaluated for increasing pain and size of the mass in his lower back. He denied bowel, bladder, or sexual dysfunction. Radiography revealed a well-defined lobulated lytic lesion in the sacrum (Fig. 1). Subsequent CT showed a destructive lesion of the distal sacrum with soft tissue mass filling and expanding the sacral canal and extending anteriorly (Fig. 2). Open biopsy was performed, and the diagnosis of myxopapillary ependymoma was made. Postbiopsy MR (0.3 T) again showed expansion of the central canal and destruction of the sacrum from the level of the S2 intervertebral foramina through most of the sacrum inferiorly (Fig. 3). A large lobulated soft tissue mass was present within the canal and anterior to the sacrum, displacing the rectum. The lesion uniformly enhanced with gadolinium. Sacral laminectomy and subtotal excision of the large myxopapillary ependymoma was subsequently performed followed by the administration of 59.9 GY of radiation therapy to the tumor. At 2 years post-treatment there was no clinical evidence of persistent tumor and the patient remained neurologically intact, including bowel, bladder, and sexual function.

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**FIG. 1.** Anteroposterior (a) and lateral (b) radiographs show a well-defined lytic lesion of the sacrum.

**FIG. 2.** Postcontrast CT scan: The sacral canal is expanded and filled with soft tissue density, and the tumor has replaced most of the sacrum. The anterior mass displaces the rectum.

**FIG. 3.** (a and b): Sagittal MR images: [SE 600/30 (a), SE 2,000/85 (b)]. The mass appears centered in the sacrum. Soft tissue is again shown to fill the sacral canal and to project anteriorly. c: MR image following Gd-DTPA administration (SE 650/22). The tumor shows homogeneous enhancement.

**DISCUSSION**

Bony destruction does not commonly occur with myxopapillary ependymomas (25). Although erosions of the sacrum have been reported (4, 18, 23), these are usually difficult to detect on plain radiography (2). Large destructive lesions of the sacrum due to myxopapillary ependymomas are rare (13), and extensive bony involvement of the sacrum either is generally found with tumors that have metastasized systemically (6, 14) or are recurrent (2, 11).

In this case it was not possible to discern the exact origin of the lesion because destruction of the sacrum was widespread and the lobulated mass extended both anteriorly and posteriorly. It is possible that the tumor originated in the filum terminale, as most myxopapillary ependymomas do; however, a pre- or intrasacral extraspinal origin with subsequent extension into the canal could not be excluded.

Myxopapillary ependymomas have been described outside the central nervous system as direct
extension or metastasis from a primary tumor in the filum terminale, cauda equina, or lower spinal cord (2) or as soft tissue seeding following surgical excision (5). A presacral primary tumor (18) or a subcutaneous myxopapillary ependymoma of the sacrococcygeal area (5) may be present without communication with the spinal cord.

Several pathogeneses have been proposed for primary extraspinal myxopapillary ependymomas. The tumor may arise from the intradural filum terminale extending through the dura. Anderson has suggested that heterotopic ependymal cells might account for extraspinal ependymomas (2), including extradural remnants of the filum terminale (3,26) or the coccygeal medullary vestige (2,5,18), a portion of neural tube lined by ependymal cells.

Aneurysmal bone cyst, chordoma, and giant cell tumor are the most commonly encountered sacral tumors that cause sacral destruction. Intrasacral cysts with large osteolytic areas encompassing most of the sacrum have also been described (27,28). Additional differential possibilities include metastases, plasmacytoma, teratoma, meningeal cyst or diverticulum, neurofibroma, and infection. Ependymomas, especially the myxopapillary subtype, should be considered in the differential diagnosis of a lytic lesion in the sacrum.

REFERENCES