Intraforaminal schwannoma of the sacrum

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Clinical information

A 53-year-old man presented with a 6-month history of back pain, with left leg discomfort and paresthesia of recent onset. No history of lower extremity numbness or weakness, bowel or bladder complaints, or trauma was elicited. Physical examination disclosed midline tenderness over the lumbar and sacral region. Although the neurological examination revealed normal muscle strength, reflexes and sensation, left-sided radiating leg pain could be invoked with straight leg raising. The clinical signs and symptoms were consistent with a herniated nucleus pulposus.

Plain radiographs of the lumbar spine were normal. An anteroposterior view of the pelvis demonstrated a well-demarcated radiolucent lesion near the midline of the sacrum, which was outlined with a thin sclerotic border (Fig. 1). Computerized axial tomography (CT) revealed a lucent lesion (Fig. 2).

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sion adjacent to the left second sacral nerve root foramina. The lesion was homogenous in appearance without evidence of calcification. While the mass did not involve the cortex anteriorly, it did extend beyond the cortex posteriorly to abut the paraspinal musculature (Fig. 2). Magnetic resonance (MR) images identified a solitary soft tissue mass which appeared to arise from the left second sacral nerve root. On T1 and T2 images, the mass was isointense with the nerve. The ovoid lesions was well defined with smooth margins. On T1 images the mass was low signal and isointense with the surrounding muscles. It enhanced moderately with T2 parameters. Utilization of intravenous Magnevist contrast better defined tumor margins and resulted in marked enhancement. Some intratumoral inhomogeneity was seen (Fig. 3). The patient underwent a posterior needle biopsy under CT guidance.

Fine-needle aspiration biopsy demonstrated clusters of bland spindled cells with palisading nuclei (Antoni A) and areas of diminishing cellularity containing microcystic degenerated foamy macrophages (Antoni B), diagnostic of a benign schwannoma (neurilemoma). The patient underwent a marginal resection through a posterior approach. Intraoperatively the tumor originated from and compressed the second sacral dorsal nerve root ganglion near its origin from the lower limit of the dural sheath. Grossly the tissue measured 6.0x4.0x1.2 cm in aggregate and was soft and gray-white in appearance. Microscopic examination revealed a predominance of Antoni A areas, characterized by bland spindled cells with prominent nuclear palisading (Fig. 4). Other areas were less cellular and showed evidence of both recent and remote microscopic hemorrhage with both hemosiderin-laden macrophages and microcystic degeneration with foamy macrophages (Antoni B). In some areas, the tumor invaded the surrounding lamellar bone, replacing some of the marrow spaces. A tiny portion of well-myelinated nerve and dorsal root ganglion was present. These histologic features were most consistent with a benign schwannoma, and intraforaminal schwannoma of the sacrum was diagnosed.

The preoperative differential diagnosis included bony intrasacral neoplasms such as chordoma and giant cell tumor. In addition, peripheral nerve tumors were considered, such as neurofibroma, schwannoma, and ependymoma.

Discussion

Sacral chordomas are primary malignant tumors of the axial skeleton that are characteristically slow growing and locally invasive. The majority (40–50%) originate in the sacrum, with the remainder presenting at the base of the skull and spinal vertebrae [1, 2]. The symptoms and signs of sacral chordomas are often late to present and relate primarily to nerve root involvement. Sacral nerve root compression can result in bladder, bowel, and lower extremity paraplegia, especially with high sacral involvement. Plain radiographs are often difficult to assess because of the overlying pelvic organs. Anteroposterior projection usually demonstrates central sacral destruction, the lateral view revealing a presacral soft tissue mass. CT imaging is the study of choice and usually reveals midline sacral involvement, with peripheral calcification being a common feature (40–50% of cases) [2]. MR imaging usually complements the results of CT, with soft tissue extension being better appreciated with MRI. The microscopic appearance is fairly typical, consisting of physaliferous cells in cords, clusters, or sheets. Occasionally cartilage is present, giving rise to the term “chondroid chordoma,” which can resemble chordosarcoma. Aggressive surgical resection is recommended to prevent local recurrence, with radiation therapy reserved for residual gross or microscopic disease [1, 2].

Giant cell tumors characteristically present in the long bones, primarily in the distal femur or proximal tibia. They are the second most common sacral tumor, with approximately 4% occurring in this region [3, 4]. Though benign, these lesions are locally aggressive. Treatment of sacral giant cell tumors can be difficult, radical resection being recommended by some authors. Radiation therapy has been used, but is of questionable efficacy [5].

Soft tissue tumors involving the sacrum secondarily include neurofibromas, schwannomas, and ependymomas. Patients with neurofibromas generally present with symptoms referable to local soft tissue compression. Radicular symptoms, similar to those associated with a herniated intravertebral disc, can occur with tumors located in the spinal nerve root foramina. MR imaging often demonstrates a central fusiform mass on T1 images and a hyperintense signal on T2 with normal nerve entering and exiting the lesion [6]. Histologically, neurofibromas contain interlacing bundles of spindled cells with wavy, elongated, darkly staining nuclei. Varying amounts of mucoid material are interspersed between cells and collagen strands. In addition, small neurites can often be identified throughout these tumors. Surgical excision leads to relief of symptoms in most cases [7].