

Back Pain: One Year Since Onset

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Clinical Case

A 30-year old male without any history of interest attended our outpatient consult due to back pain that had its onset 1 year before, was of increasing intensity and had responded poorly to treatment with anti-inflammatory drugs. The physical examination did not find anything of interest. A simple x-ray of the lumbo-sacral spine (Figure 1) showed a lesion with a predominantly lytic component in the sacrococcygeal region and an evident mass-effect at a presacral level. The patient was hospitalized to complete a study protocol that included a magnetic resonance of the pelvis (Figure 2), that confirmed a tumor that had a lobulated appearance in the sacrococcygeal region, measuring 14 × 9 cm, associated to a voluminous soft-tissue mass. A computed tomography guided biopsy, visualizing small, amorphous, intratumoral calcifications.

Diagnosis and Evolution

The sacral mass biopsy showed mixoid material and multivacuolated cells (chordoid cells) (Figure 3), positive to the S-100 protein and cytokeratin in the immunohistochemical staining, findings that are compatible with sacral chordoma. After being informed of the diagnosis and the therapeutic options that included sacrectomy and radiotherapy, the patient opted to be transferred to a hospital in the United States for treatment.

Discussion

The chordoma is the most frequent malignant tumor of the sacrum,¹ representing approximately 4% of bone tumors, with an estimated yearly incidence of 0.5 cases/



Figure 1. Simple x-ray of the lumbosacral spine. Lateral Projection. Lesion with a mass-effect in the posterior zone of the minor pelvis (arrows) and alterations in the sacro-coccyx bone texture with some lytic component.

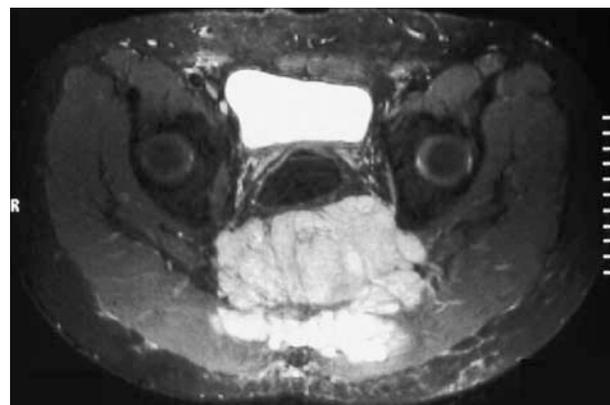


Figure 2. Magnetic resonance of the pelvis. STIR sequence. Multi-lobulated mass in the sacral-coccyx region with preferential presacrum extension associated with an anterior displacement of the rectum.

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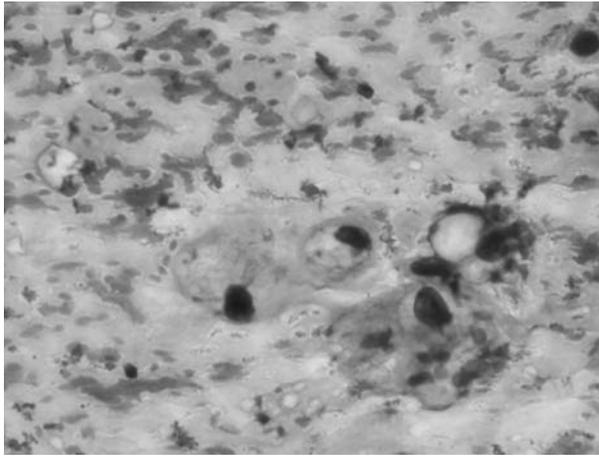


Figure 3. Biopsy of the sacrum mass. To be noted is the presence of multivacuolated cell elements that form plates (fisalipherous cells) on a background of metachromatic mixoid material.

1 000 000 persons/year.² It can present itself at any age, though generally it occurs between the fourth and seventh decades of life and is more frequent in men (2:1).¹ It is believed to originate from remains of the notochordae. It affects the axial skeleton, with a predilection for the sacrum (50%) and the base of the skull, and occurs only rarely in other localizations. Clinical manifestations depend on the localization of the tumor but are generally non-specific which accounts for the delay between the onset of symptoms and the diagnosis, estimated around a mean of 12 months.³ Among its histopathological characteristics the presence of fisalipherous cells stands out, as well as immunoreactivity to S-100 protein and several cytokeratins.⁴ Some karyotypification studies show that chordomas present complex chromosomal reordering⁵ and there have even been family cases described.⁶ Recently,⁷ its pathogeny has been linked to modifications on chromosome 7. The gold-standard for treatment is surgical block resection, but in many cases this is not possible.⁸ Therefore, the tumors found on the base of the skull are small insize but

are difficult to extract due to their vicinity to vital neurological structures. On the other hand, sacrococcygeal chordomas are usually large and adhered to surrounding tissue, making it impossible to completely extract the lesion.⁹ This fact is fundamental because if the margins of the resection are not free of the lesion a relapse is almost guaranteed.² Therefore, the rate of local relapse and metastasis reaches 60% in sacrovertebral chondromas. Relapses occur in the soft tissue around the sacrum, rectum, gluteus, and perianal region.² Metastasis can occur in advanced stages of important local disease.² Mean 5-year survival rates are estimated at 50%-80%.⁹ Although theoretically chordomas are radioresistant, published series have proven it effective, attenuating local relapse in those cases with incomplete resections^{2,9,10} and increases survival in a significant manner.⁹

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