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CASE REPORT

Any Etiquette for Sacrococcygeal Chordoma's Diagnosis?

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ABSTRACT

The current paper focuses on a trial to understand the imaging manifestations in combination with the clinical presentation of the sacrococcygeal chordoma in a patient with referred back pain. Also, the steps for the final diagnosis are described and via *this procedure, the paper demonstrates the crucial role of magnetic resonance imaging, computed tomography guided biopsy and histopathological examination in order to minimize the differential diagnosis and lead to the correct diagnosis.*

Keywords: sacrococcygeal, chordoma, MRI, spine, tumor.

INTRODUCTION

Clinical information

A 75-year-old woman with a known tumoral lesion of the lumbosacral region of the spine, which was revealed on a previous magnetic resonance imaging (MRI) scan, was referred to our Radiology Department for further investigation by computed tomography (CT)-guided biopsy. In her medical history, chronic lumbar radiculopathy (sciatica pain) was included, which was deteriorated during the last two months. This kind of pain was combined with constipation, fecal incontinence and symptoms from the lower urinary tract (such as increased urinary frequency, urgency, painful urination and pain above the pubic region). After her clinical examination by an urologist, a MRI scan was recommended. No other medical conditions were referred in her personal anamnesis.

Imagistic diagnosis

The MRI scan revealed an expansile and well-defined lobular mass located between the

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middle part of the I3 sacral vertebra and the upper coccyx vertebrae, with maximum indicative dimensions of 92 mm (oblique sagittal) X 62 mm (oblique coronal) X 72 mm (oblique axial). T2-weighted MRI revealed very high signal. T1-weighted MR imaging and STIR imaging demonstrated intermediate to low-signal intensity and small foci of hyperintensity. The mass was extended to the sacral ala (ala ossis sacri) and to the minor pelvis, where bone swelling was occurred and mass effects were provoked on had adjacent structures (more accurately on the backside of the orthosigmoid and less to the dorsal surface of its subcutaneous tissue and to the intergluteal cleft) (Figure 1).

The checked-tissue from the biopsy characterized by necrotic areas, fluid and gelatinous mucoid substance, physaliphorous cells and low mitotic rate. Immunohistochemical control was positive for AE1/AE3, EMA and vimentin and partially for S100 and negative for any other control (Figure 2). All these findings were compatible with chordoma of sacral bone.

Imagery differential diagnosis

Radiologically differential diagnosis includes rarer tumors which arise from the sacrum and emerging sacral nerve roots, such as the neurilemmoma, which may have similar signal characteristics to sacral chordoma, but the lack of internal septa is usual (1). An intrasacral metastatic paraganglioma also shows heterogeneous signals on T2W and irregular intralesional signal voids (due to its vascularity) (1). Other conditions to consider in the differential diagnosis include chondrosarcoma, giant cell tumor, ependymoma, plasmacytoma, a solitary metastatic deposit and spinal lymphoma (1-3).

Final clinical comments

In the present case, the patient underwent complete en bloc resection of the tumor with clean

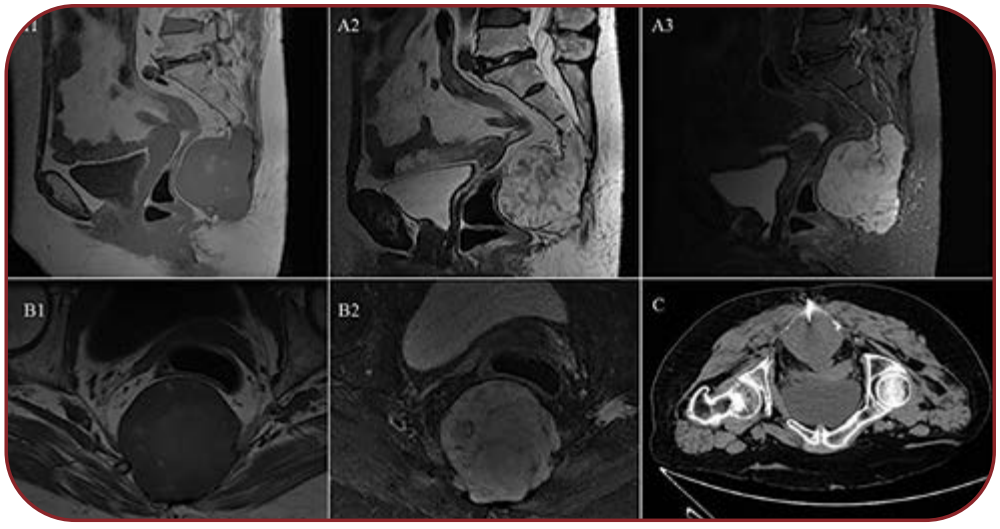

FIGURE 1. The described mass as it is showed in the MRI sagittal plain – A1) T1W; A2) T2W; A3) T2W FS (which is equal to STIR imaging) – and in MRI axial plain – B1) T1W; B2) T1W FS; and C) CT-guided core biopsy

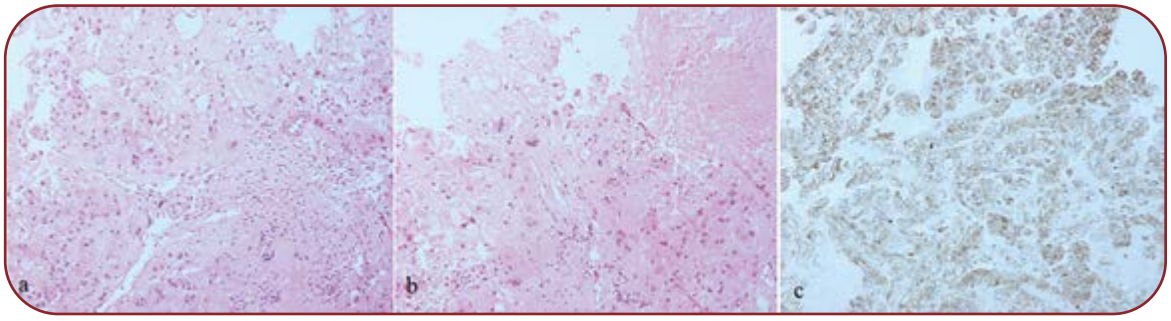

FIGURE 2. a, b) Epithelioid cells forming sheets with clear or eosinophilic cytoplasm, some of them with bubbly appearance (physaliphorous cells); nuclear pleomorphism with necrosis also noted (Hematoxylin and Eosin x 100); c) Cells diffusely expresing EMA (Immunohistochemistry x 100)

margins which seems to provide the longest survival. Pathological examination confirmed the specimen's free surgical margins. No complications were noted and no adjuvant therapy was needed by that time. Postoperatively our patient complained about fatigue and general malaise for a few days. No special medication was needed except from antibiotics and analgesics. To date, after three months of the operation the patient is symptom-free with a much better quality of life. A follow up program with total body CT screening (for detection of local recurrence and distant metastasis) after six months has been recommended. The patient is aware of possible complications (shortly or at a distance) as the operation can be associated with a high complication rate and morbidities due to extensive and time-consuming operation. The complications report includes bowel or urinary disturbances, abdominal hernias and adhesions, bladder-skin fistulas, infections, neurological impairment due to sectioned nerve roots, etc (4).

Sacrococcygeal chordoma is a rare, slow-growing bone tumor that may have large dimensions at presentation (range 3–20 cm in diameter, median 8 cm) (1). Chordoma is usually (approximately 30-50%) located at the tailbone (or either the skull base or vertebral bones) (3). It can be presented at any age, but more often between 40 and 70 years old, with an annual incidence of <0.1/100,000 people (3). Patients experience variant signs and symptoms such as back pain and neurological changes depending on where the tumor arises. Local invasiveness and destructiveness to soft tissues and skeletal muscles are common characteristics (1). The role of imaging technics is crucial, especially that of MRI, where they are usually presented with intermediate to low signal intensity in T1W images and high intensity in T2W images (1, 3). CT-guided biopsy is also a useful tool, allowing the histological evaluation. The classic cytologic features of chordomas include the physaliferous cells, small epithelioid cells and myxoid background (2, 5). Prognosis is variable (typically poor – average survival ~10 years), because chordoma is diagnosed late (6). In some cases, the tumor can be totally extracted by surgery, while adjuvant radiotherapy and and radio frequency therapy can also help to prolong survival (2). Radiotherapy can be useful and give temporary beneficial results to patients who undergo inadequate surgery or in case of unresectable tumors (7). The majority of primary sacral tumors do not respond to common radiotherapy and chemotherapy, and only extensive surgical resection represents a potential curative treatment (8). Over recent years, molecular targeted therapy based on gene mutation screening and immunohistochemistry (monotherapy or combined therapy) is also used with significant therapeutic efficacy (9). Even if total management may be therapeutic, recurrence is common (3, 6).

Conflicts of interest: none declared.
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