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Sacral Chordoma: Report of Two Cases and Review of the Literature

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Abstract **Case Report**

Chordomas are rare, slowly growing, locally aggressive neoplasms of bone that arise from embryonic remnants of the notochord. Chordomas account for 2%–4% of all primary malignant bone tumors and are the most common primary malignant sacral tumors, with the exception of lympho-proliferative diseases**.** At imaging, a chordoma typically manifests as a large destructive sacral mass with secondary soft tissue extension**.** We report two cases of confirmed sacral chordoma; detailing their radiological aspects as well as the therapeutic management.

Key words: Chordoma, CT, MRI.

INTRODUCTION

Chordomas are the fourth most common malignant neoplasms originating from bone and have an incidence of less than 0.1 per 100,000 people per year [1]; they derive from notochordal remnants. Chordomas involve the sacro-coccygeal region in 50%–60% of cases and the clivus or the spheno-occipital region in 30%–35% of cases [2]. Chordoma is in most cases pauci- or asymptomatic, which makes its diagnosis quite late. Symptoms of sacral chordoma are indolent and include pain, numbness, constipation, weakness, and incontinence. Imaging (CT and MRI) is very suggestive of the diagnosis, which is confirmed by histological study.

FIRST CASE REPORT

A 55-year-old patient, who had chronic sacro-coccygeal pain for 2 years, with recent onset of constipation with burning while urinating, without deterioration of general condition. The sacral CT showed a well-limited unenhanced lytic bone mass, centered on the S3-S4 and S5 vertebral bodies, which extends inside the vertebral canal and the endopelvic region, and comes into close contact with the lower rectum infiltrating the left iliococcygeus muscle (Figure 1). On MRI examination this mass was lobulated arising from the sacrum, and extending posteriorly to the left gluteus maximus muscle. Anteriorly it extended to the endopelvic region coming into contact with the lower rectum with loss of the fatty separation line in places. On T1-weighted images, the signal in the mass was predominantly isointense relative to that in muscle, with scattered areas of hyperintensity; On T2-weighted images, the mass had heterogeneous high signal intensity; on the diffusion sequence, the mass was in hypersignal without significant restriction of the ADC, with moderate, heterogeneous and late enhancement after injection of Gadolinium (figure 2).

The patient underwent gross total resection of the mass with partial sacrectomy; the post-operative follow-up was simple. Post-operative radiotherapy was not indicated.

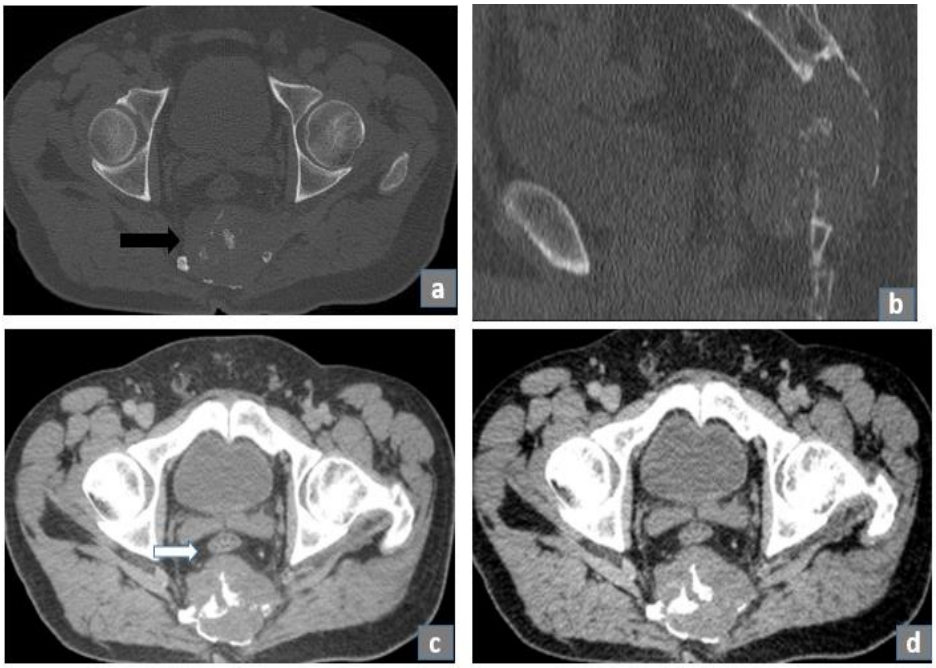

Fig-1: The sacral CT showed a well-limited lytic bone mass (a,b), centered on the S3-S4 and S5 vertebral bodies, which extends inside the vertebral canal and the endopelvic region, and comes into close contact with the low rectum (white arrow); without significant enhancement (d).

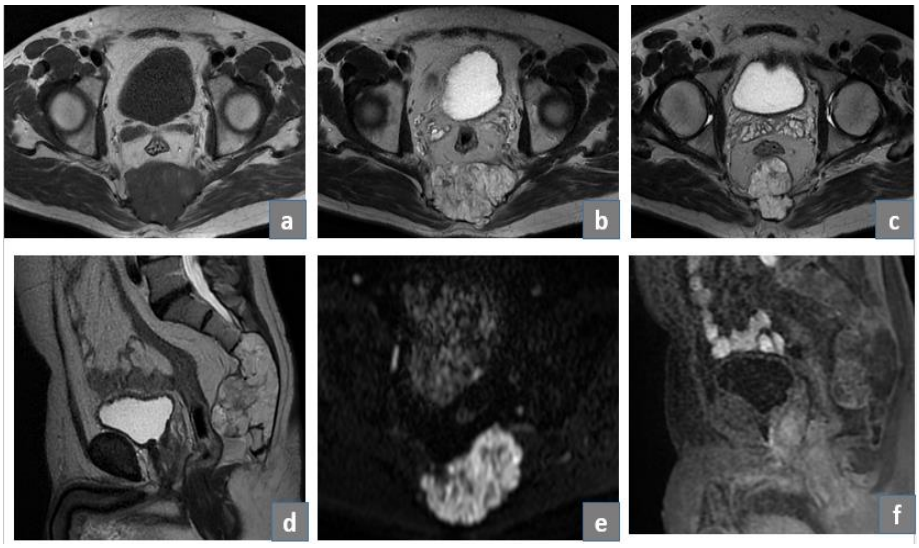

Fig-2: MRI demonstrated lobulated mass arising from the sacrum, which is isointense relative to that in muscle on T1 with scattered areas of hyperintensity (a); On T2-weighted images, the mass had heterogeneous high signal intensity (b,c,d) ; on the diffusion sequence the mass had high signal (e), with moderate, heterogeneous and late enhancement after injection of Gadolinium (f).

Second Case Report

A 77-year-old patient, who has chronic sacro-coccygeal pain recently worsening, with pain on defecation. MRI showed a voluminous sacro-coccygeal mass, fairly well limited, It was isointense relative to that in muscle, with scattered areas of hyperintensity on T1-weighted images; it had heterogeneous high signal intensity On T2-weighted images, heterogeneously enhanced after injection of the contrast product. This masse fills the spinal canal, encompasses the sacral nerve roots; extends into the endopelvic region with filling of the ischio-rectal fossa and infiltration of the levator ani muscle (Figure 3).

The patient underwent a partial resection, given the size and the infiltrating nature of the tumor, with adjuvant radiotherapy.

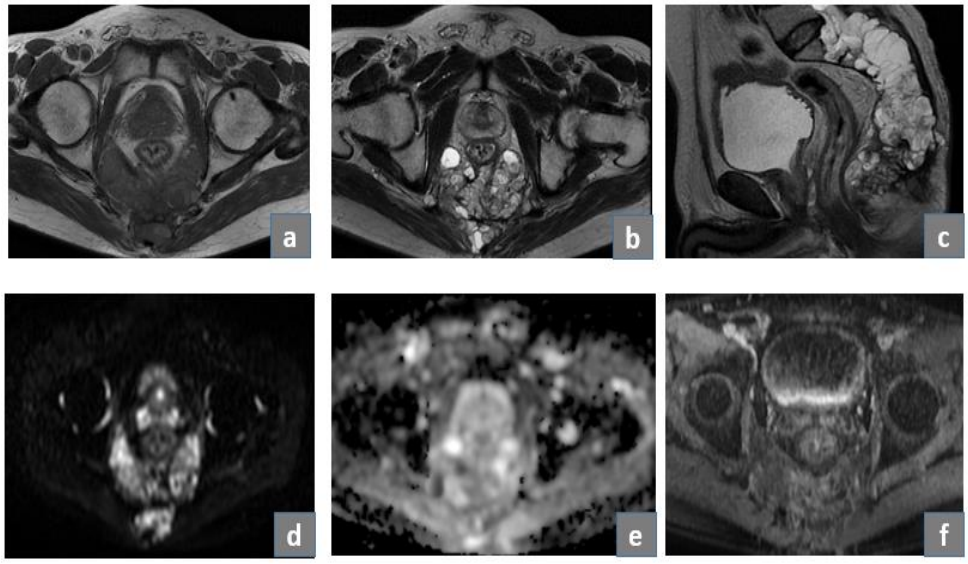

Fig-3: MRI demonstrated lobulated and infiltrative sacro-coccygeal mass with vertebral canal extension; which is isointense to the muscle on T1 with scattered areas of hyper-intensity (a); On T2-weighted images, the mass had heterogeneous high signal intensity (b,c) ; on the diffusion sequence the mass had high signal (d), with ADC restriction zones (e) heterogeneously enhanced after injection of Gadolinium (f)

DISCUSSION

Chordomas are primary bone tumors, which arise from notochord remnants along the axial skeleton. They involve the sacro-coccygeal region in 50%–60% of cases and the clivus or the spheno-occipital region in 30%–35% of cases [2]. There is a broad age distribution, with most individuals presenting in the 4th to 7th decade and a peak in the 5th decade [3]; however, presentation with skull base tumors may occur at a younger age [4]; and has been reported in children and adolescents. There is a male-to- female ratio of 3/1 [5].

Sacral chordoma can result in back pain, dysfunction of sphincter, radiculopathies, and motor weakness of lower extremities [6, 7]. The average duration of preoperative symptoms is 14 months (range, 4-24 months) [4, 8, 9, 10]. Patients often present late in the disease process due to the slow-growing tumour mass, which produces vague, non-specific symptoms in elderly patients that are not initially viewed as sinister in origin [1].

Histologically, chordomas consist of fibrous tissue separating chords of tumor from mucoid matrix. There are 3 histological subtypes: classical, chondroid and de-differentiated, with the latter being the most aggressive and affecting mostly children [11]. Immuno-histological markers are needed to establish diagnosis and to differentiate it from other tumors like chondrosarcoma.

At imaging, a chordoma typically manifests as a large destructive sacral mass with secondary soft tissue extension [12]. Radiographs can show bone erosions with irregular calcium foci along with a lytic lesion [13], It may show sacral osteolysis with an associated soft-tissue mass and calcifications [11]. CT can be very useful for defining the extent of bone involvement. It shows bone destruction with an associated lobulated midline soft-tissue mass [12]. MRI is considered superior to other imaging modalities; the most striking feature of a chordoma is the high signal intensity seen on T2-weighted images. High T2 signal intensity is a nonspecific feature; however, the combination of high T2 signal intensity and a lobulated sacral mass that contains areas of hemorrhage and calcification is strongly suggestive of a chordoma [12]; Chordomas tend to show hypointense or isointense signal relative to that in muscle on T1-weighted images, and contrast-enhanced images show a modest heterogeneous enhancement in the soft-tissue components of the tumor.

The differential diagnosis of sacral tumors with bone destruction includes chordoma, giant cell tumor, chondrosarcoma, myxopapillary ependymoma, plasmacytoma, and metastasis. In the case of Giant cell tumor and Plasmacytoma; The radiographs appear as large lytic lesions with poorly defined borders. These tumors are aggressive and expansile, and may cross the sacroiliac joint posing a radiological challenge to differentiate from chordome [14]. Giant cell tumor is the second most common primary sacral neoplasm after chordoma [12]. Metastatic disease is especially seen in the sacrum in the elderly with primaries from the prostate, breast, thyroid, lung and colon. A thorough history pertaining to symptoms and previous ailments may help in guiding towards investigations. Chronic infections caused by tuberculosis and fungus should always be considered [14].

Chordoma has been considered of low metastatic potential; however, distant metastasis to lung, bone, soft tissue, lymph node, liver, and skin has been reported in up to 43% of patients [15, 16]. Metastatic sites, however, usually occur late in the course of the disease [16].

Surgical excision forms the mainstay of treatment of sacral chordoma; Complete surgical resection with negative surgical margin in localized disease is the mainstay of care [17], sacrectomy with wide resection margins offers the best long term oncological outcomes but may lead to significant functional compromise; Chordomas are relatively radio-resistant [1]; However, radiotherapy can be useful, and it can give temporary benefit to patients who underwent inadequate surgery or in case of unresectable tumours [18]. Imaging-guided percutaneous radiofrequency ablation was recently described as a possible alternative to surgery for locally recurrent disease or for palliation [15]; Due to poor sensitivity, chemotherapy is not instituted in patients with sacral chordome [14].

The prognosis in patients with chordoma is defined by the propensity of the tumor to recur locally. Although metastases also may occur, most patients who succumb to the disease do so because of local recurrences [12].

CONCLUSION

Chordoma is the most common primary malignant tumor of the sacrum; MRI is the gold standard in imaging. At present, en bloc resection with possible radiation therapy remains the only effective treatment option; and there is significant morbidity associated with the tumor. There is optimism regarding development of standardized therapies; however that will require more funding and internal collaborations that are carried out on a larger scale.

Abbreviations

ADC: apparent diffusion coefficient
CR: conventional radiographs
CT: computed tomodensitometry
MRI: magnetic resonance imaging

Conflict of interest

All authors state that they have no conflicts of interest.

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