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**Radiology**

# A Mass in the Buttock: The Sacral Chordoma, a Rare Primary Bone Tumor. About Two Cases and Review of the Literature

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**Abstract** **Case Report**

Chordomas are rare bone tumors. They represent 2 to 3% of primary bone tumors, affecting the sacral parts in the majority of cases. They are characterized by their insidious evolution. Imaging plays an important role in the diagnosis, particularly with standard radiography and CT scans, allowing an optimal evaluation of bone involvement, while MRI specifies the degree of invasion of soft tissue and neighboring structures. Ultrasound is also used to guide biopsy procedures, and interventional radiology, through the combination of angiography and radiological embolization, can limit the risk of bleeding during surgery. We report two observations illustrating the role of radiology at the diagnostic and therapeutic level in two patients diagnosed with chordoma.

**Keywords:** Chordoma - Sacro-coccygeal - Imaging - Embolization.



## INTRODUCTION

Chordomas are rare primary bone tumors representing only 2-3% of primary malignant bone tumors [1]. They develop from the embryonic remnants of the notochord of the axial skeleton. The sacral parts are the most affected, with peaks in frequency between the 5th and 6th[e] decade [2]. Like most malignant tumors, these tumors are usually revealed late because of their clinical symptoms, which are not very specific and since they evolve progressively with little fanfare. Surgical excision in healthy margins is currently the gold standard for operable sacral chordomas [3]. However, interventional radiology remains an option to consider in the management of these sacral tumors. We report two observations illustrating the role of radiology in the diagnostic and therapeutic management of two patients with a chordoma.

## OBSERVATIONS

**Case 1:**

A 53-year-old patient consulted for a swelling of the sacral region associated with perineal neuralgia. The clinical examination revealed a swelling of the sacral region. On CT scan, a solid mass centered on the sacrum was found, suggesting a plasmacytoma or a chordoma with polycyclic osteolysis involving the sacral vertebrae (Figure 1). On MRI, the mass was centered on the second and third sacral pieces, with heterogeneous T2 hyper signal and T1 hypo signal, intensely enhanced after injection of gadolinium, with intra-lesional necrotic areas and erosion of the bone corticals of S2 and S3, he retro sacral soft tissues and the right erector spinae muscle were invaded. There was also involvement of the right piriformis muscle and edematous infiltration of the homolateral gluteus maximus muscle, suggesting the infiltrative aspect of the tumor locally (Figure 2). An ultrasound-guided biopsy revealed a morphological and immunohistochemical appearance suggestive of a chordoma.

The indication of an adjuvant radiological pre-surgery embolization in order to limit the hemorrhagic risk was suggested and then carried out. An embolization of the left lateral sacral artery was performed. On the right, due to a minimal tumor blush during the injection of the posterior trunk of the right internal iliac artery, the microcatheterization of the right lateral sacral artery was unstable and embolization was not performed (Figure 3).

Surgical treatment was subsequently performed with tumor resection (Figure 4).

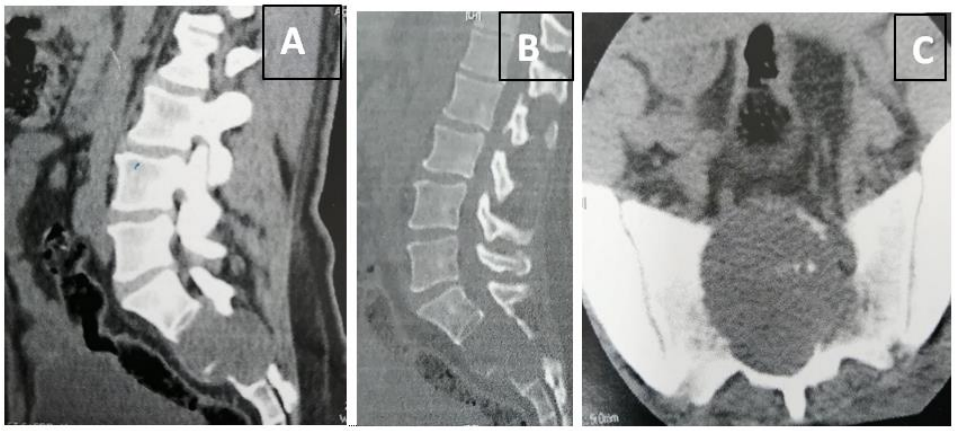


**Figure 1: CT scan of the lumbosacral spine in sagittal (A and B) and axial (C) sections. Polycyclic osteolysis involving the sacral vertebrae with a tissue process centered on the sacrum suggesting a chordoma**

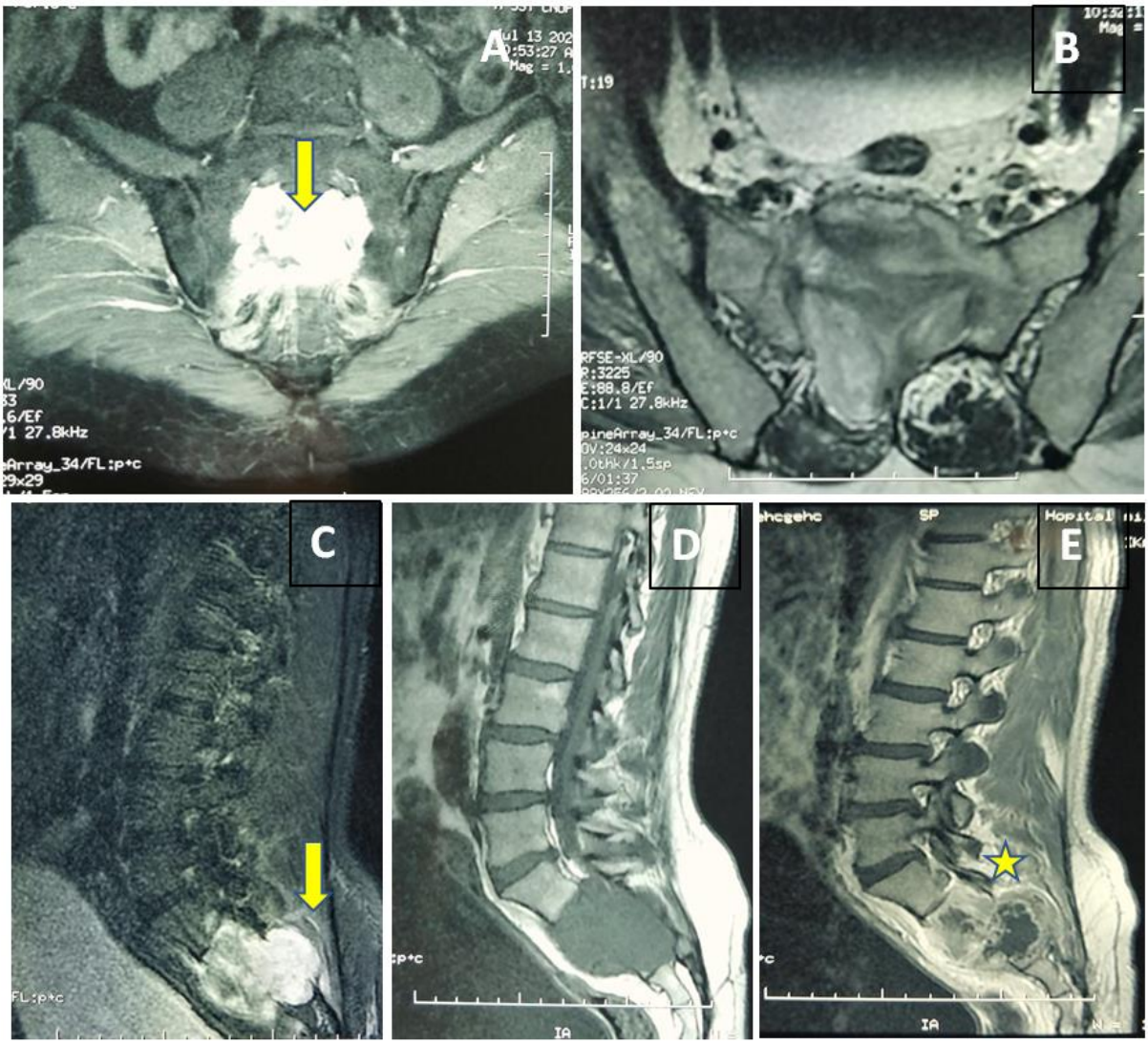


**Figure 2: Lumbosacral MRI in T2 axial (A and B) and sagittal sections without injection of gadolinium (C and D) and after injection of Gadolinium (E). Chondroma centered on the second and third sacral pieces (Arrow), in heterogeneous T2 hyper signal intensely enhanced after injection of gadolinium, seat of intra-lesional necrotic zones (star)**

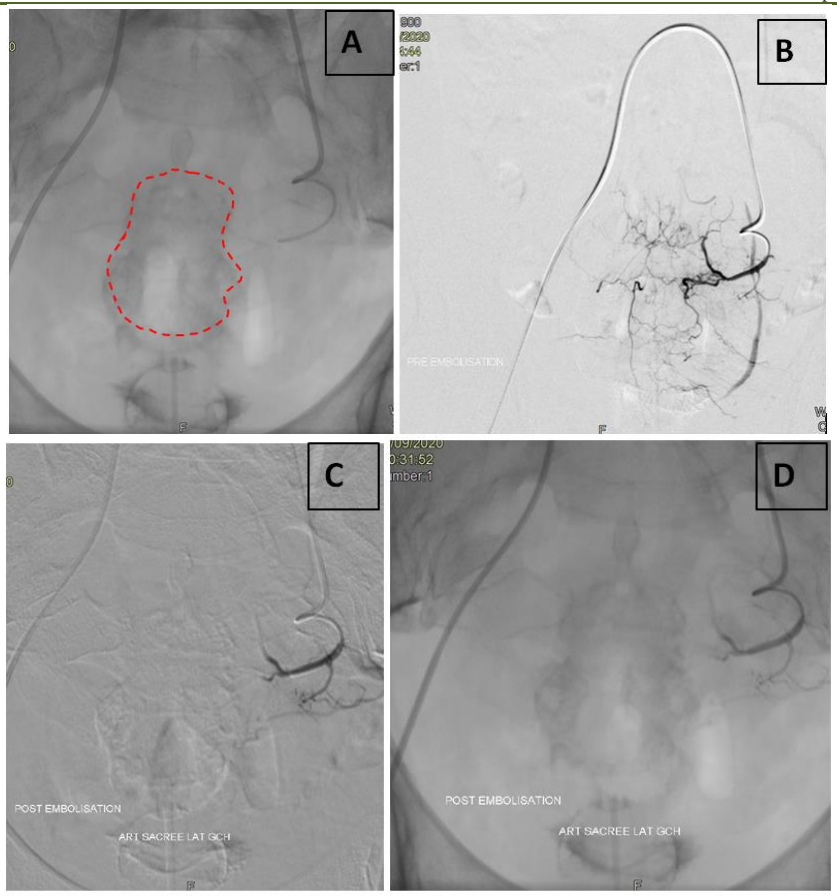


**Figure 3: Pre (A and B) and post-embolization (C and D) angiography of sacral chordoma (Case 1). Note the projection of the sacral tumor outlined in red (A)**

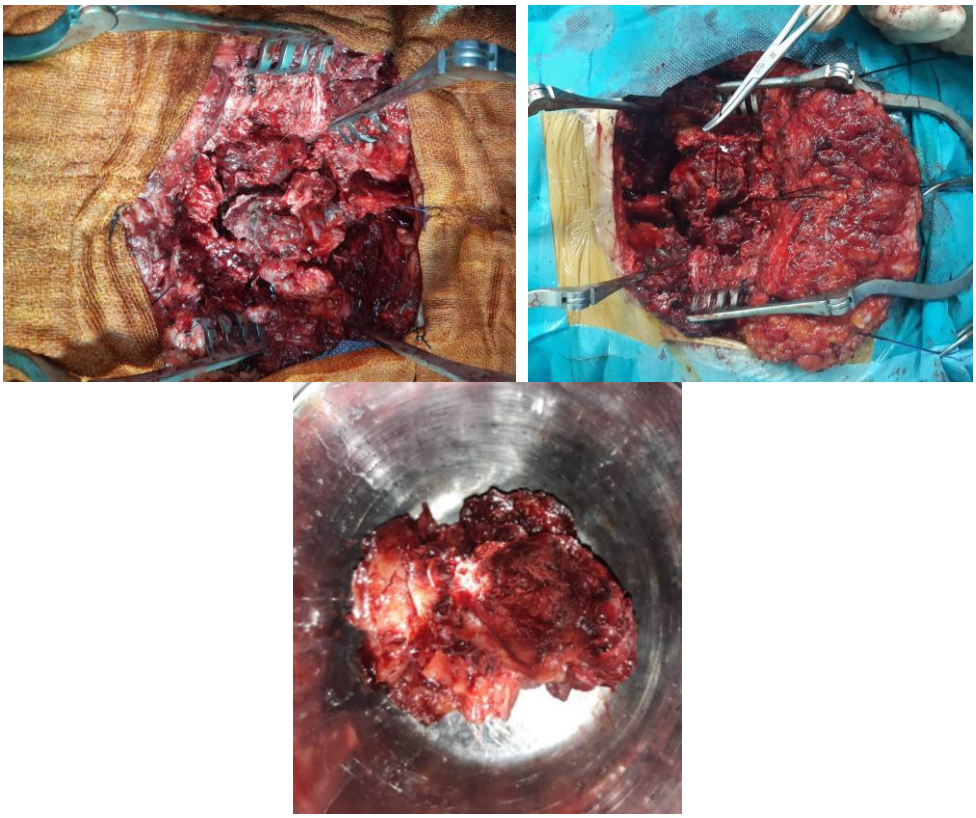

**Figure 4: Intraoperative aspect of the sacral chordoma (Case 1)**

**Case 2:**

A 62-year-old female patient with no particular pathological history consulted for progressive onset of a low back pain evolving for 2 years recently associated with sphincter disorders. On examination, hypoesthesia of the left lower limb and hypoesthesia in the saddle were found. On CT scan, an osteolytic solid mass with endopelvic extension and bilateral ureterohydronephrosis was found. MRI revealed a large tissue mass encompassing the lower third of S1 and all the other sacral pieces and the coccyx, with heterogeneous T2 hyper signal and T1 hypo signal, enhanced after injection of gadolinium, delineating areas of intra-lesional necrosis (Figure 5).

Ultrasound biopsy with anatomopathological examination revealed an aspect compatible with a chordoma.

Preoperative embolization (Figure 6) followed by lumpectomy was successfully performed.

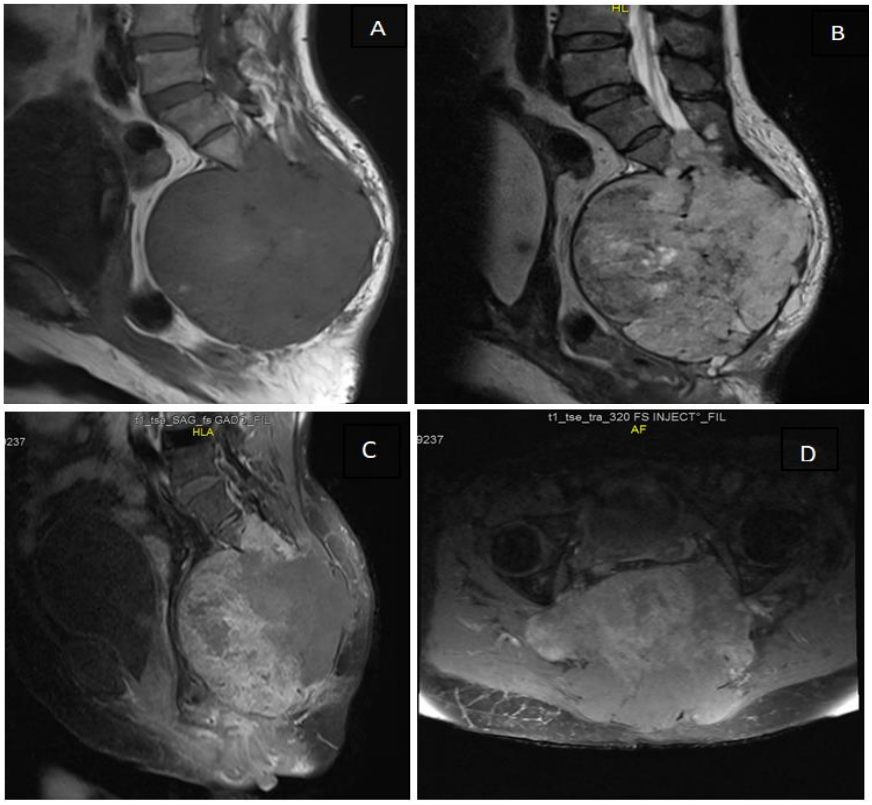


**Figure 5: MRI in sagittal (A, B, C) and axial (D) sections, T1 (A), T2 (B) and T1 weighted after injection of gadolinium showing a sacrococcygeal chordoma in T1 hypo signal, T2 hyper signal, intensely and heterogeneously enhanced after injection of gadolinium, delimiting areas of necrosis (C, D) (Case 2)**

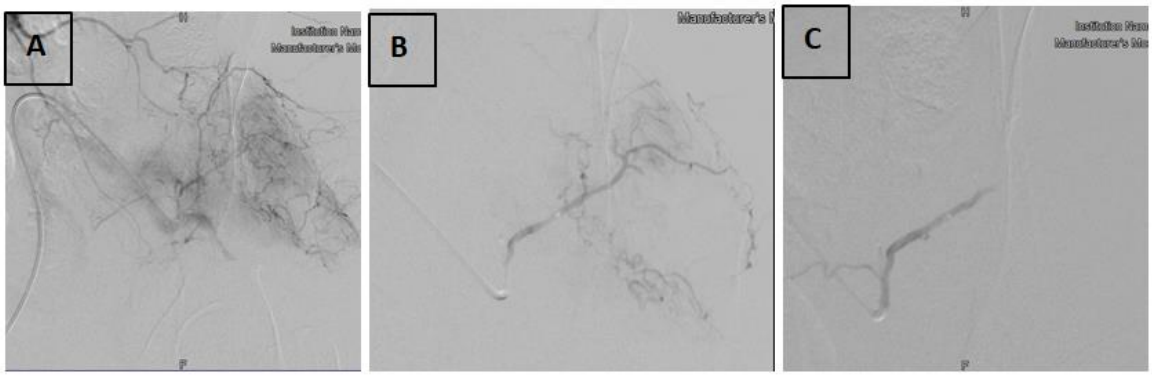


**Figure 6: Angiography. Pre, per and post embolization aspects of sacral chordoma (Case 2)**

## DISCUSSION

Chordomas are rare primary bone tumors. They represent 2 to 3% of primary malignant bone tumors. They affect the sacrococcygeal region in 50 to 60% of cases and the clivus or spheno-occipital region in 30 to 35% of cases [4]. According to the studies, the average age is between the fifth and sixth decade. In our observations, the age of our patients was included in

that range. Clinically, the symptoms often reported are related to the organs surronding the tumor. They may include a painful syndrome of the buttock or sacro-coccygeal region, numbness, constipation or incontinence [5].

On imaging, the signs are not specific. Standard X-rays and CT scans allow a good bone analysis. Four important signs are to be looked for on X-ray: bone expansion and rarefaction, intra-tumoral trabeculations, and calcifications [6]. On the CT scan, as in our case, a sacro-coccygeal osteolysis associated with a mass of similar density as the adjacent soft tissues is very often found. A strong argument in favor of the diagnosis would be the existence of areas of lower density within the tumor. On MRI, the tumor appears as a T1 hypo or iso signal, T2 hyper signal. The lobulated appearance of the tumor on T2 and T1 sequences after injection of Gadolinium is suggestive. Some authors defend the superiority of MRI compared to CT scan after injection of contrast, suggesting that a CT scan without injection of contrast should be performed first for optimal bone analysis, complemented by MRI in T1, T2, and T1 weighting after injection of Gadolinium [7]. In our observations, this protocol was followed. Another role of MRI lies in the accuracy of locoregional tumor extension. Indeed, it is important to know the state of the surrounding organs, in particular the rectum, the bladder, the limits of the lesion, the possible extension to the gluteal muscles, to the sciatic nerve but also the inclusion of the lumbosacral roots by the tumor. All this informatios are important for the surgical approach. According to Stephens and Schwartz, the rectal wall is respected in the majority of cases [7]. In our observations, the rectal wall was respected.

The differential diagnosis of sacrococcygeal chordomas includes chondrosarcomas, giant cell tumors, aneurysmal cysts, Ewing's sarcoma, plasmacytomas, and metastases.

Surgical resection, known as "block surgery", remains the gold standard [3]. However, the complex location of the tumor makes this procedure difficult as when it is located above the third sacral piece. In the literature, the chordomas that are most easily removed are those located below S3, hence the importance of providing the surgeon with this information [8].

However, there is a high risk of bleeding during surgery. Some authors describe the role of angiography and embolization for a good analysis of the vascular network and tumor neoangiogenesis and the reduction of the hemorrhagic risk. Wang et al describe satisfactory results in a series of spinal tumors [9]. In this case, embolization would aim to limit the risk of bleeding during surgery. This approach was adopted in our two observations with considerable therapeutic success.

## CONCLUSION

Chordomas are rare primary bone tumors. Imaging plays a considerable role not only in the diagnosis by assessing the locoregional extension of the tumor, but also in the therapeutic management by pre surgical angiography and embolization, limiting the risk of bleeding. However, surgery remains the gold standard in the management of these lesions.

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