



## SACRAL CHORDOMA : CLINICAL CASE

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### **Abstract :**

Chordoma is the most common primitive malignant tumors of the sacrum, arises from embryonic notochord. The diversity of their clinical presentation, their management of diagnosis and therapy is illustrated through a clinical case collected in department of surgery II ,Mohammed V hospital in Rabat , Morocco .

Key words : chordoma , notochord, bone tumors

### **Introduction :**

We report our surgical experience a case of sacral chordoma at Mohammed V ,military hospital in Rabat , Morocco department of Surgery II, and we highlight current standard of management of in the light of recent literature data.

### **Observation :**

In 2016 a 53-year old man with no past medical or surgical history admitted in surgical department II for management of a pelvic mass . The history of his illness began 8 months back before his hospitalization, patient had history of loose weight, about 10 kg in last six months , pain in posterior pelvic region cruralgia with uralgia irradiating to lower limb, increase frequency of urination , urinary urgency ,and severe constipation.

On examination, abdomen soft, no tenderness, no palpable mass, no ascites, no peripheral lymph nodes noted.

A laboratory investigation revealed normocytic normochromic anaemia . Tumor markers : ACE and CA19-9 are normal, no sign of inflammatory process.

A rectal examination showed a rectal stenosis produced by mass compression. A CT scan abdomen and pelvis revealed a extramucosal mass , developed from the 1st and 2 ed sacral vertebrae (S1 and S2), the rectum shifted right - anteriorly, the urinary bladder collapsed and pushed superiorly ( Figure 1).



Figure 1 : Tomography examination showing the sacral tumor.

Magnetic resonance imaging of the pelvis showed a presence of a sacral polylobed tissue Mass , process developed at the expense of S1 - S2, this process infiltrates posteriorly to S3 and S4 with pelvic involvement ( Figure 2).

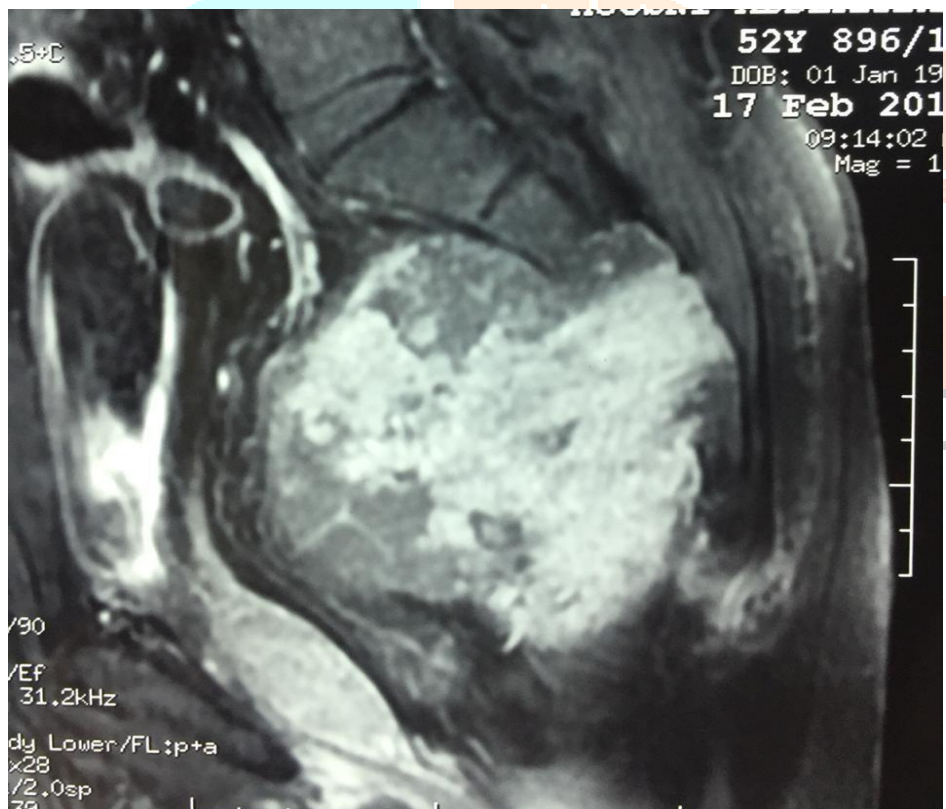


Figure 2 : Sagittal section of pelvic MRI, after gadolinium injection showing the tumor process.

The exact histological diagnosis was not established, and an indication of abdominal resection of tumor was made . The patient was informed about the risks of the intervention, including bleeding ,neurological risk and the possibility of digestive stoma.

Under general anesthesia, the patient was placed in decubitus dorsal position with hips pushed slightly upward , a median infra-umbilical incision carried out , peritoneum opened, no carcinosis or hepatic metastases were found.The retrorectal dissection in the avascular zone found an non -capsulated whitish mass with encephaloid aspect.

The mass is resequaled to the sacrum by a laborious dissection up to the pelvic floor with a loss of substance of about 4cm and extraction of intrarachidian tumor material. Hemostasis secured vacuum drainage placed, wound closed.

The final diagnosis is based on the morphological aspect and the immunocytochemistry profile compatible with a sacral chordoma.

The post-operative, marked by the rapid recovery of the patient and remarkable neurological improvement and the disappearance of constipation and pollakiuria.

One month after healing, our patient received adjuvant radiotherapy of 50 grays for 01 month at the tumor site to treat intra bones lesions in the sacrum.

A control pelvic MRI objectified a stable aspect of the process centered on S3, S4.

This process has an intracanalicular extension with infiltration of sacral nerves, rectum and urinary bladder: without anomaly (Figure 3).

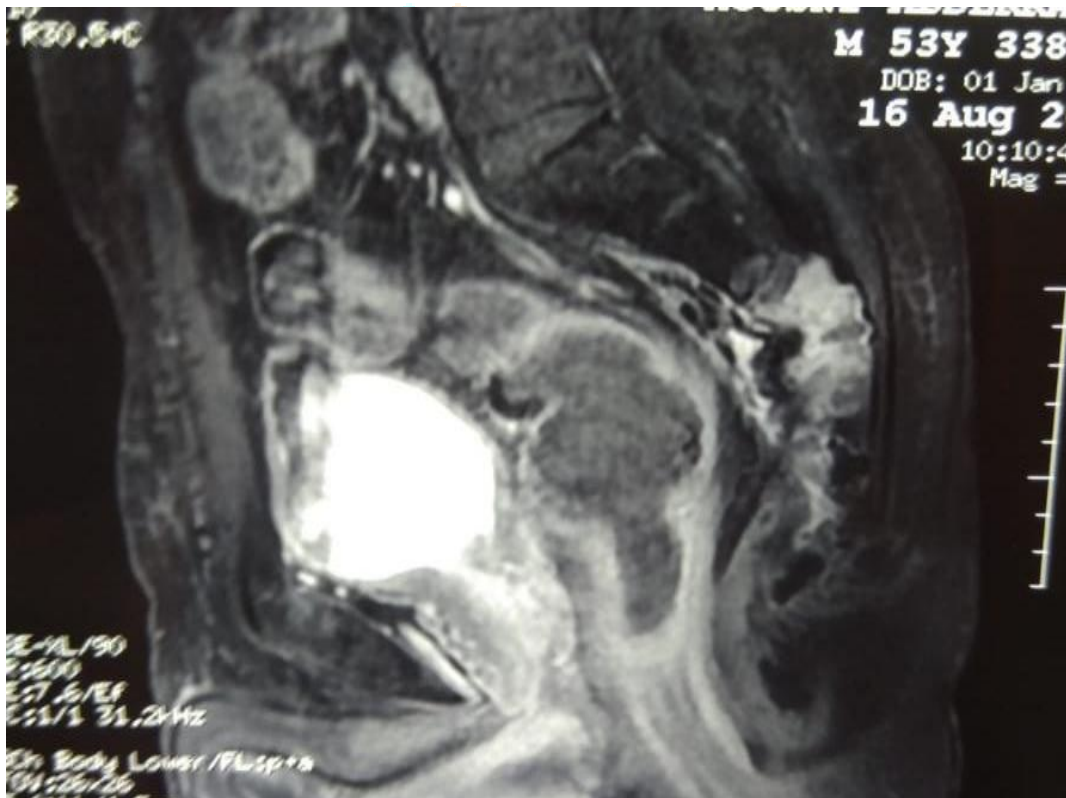


Figure 3 : control pelvic MRI

The evolution was favorable, the patient was reviewed regularly, within a period of one and a half years, and was sent to the Neurosurgery department for additional surgical treatment to reduce the neuralgia that appeared and were less intense.

**Discussion :**

Chordoma is a very rare tumor. Its incidence varies according to studies from 0.5 to 8 per million people per year. More common in the Caucasian population.

The chordoma represents from 1 to 4% of primary malignant bone tumors. It can be seen at all ages, the lesion is predominant in adults and the fourth in children under 15 years of age. Man is significantly more affected than woman (03) (sex ratio M / F = 3/1) [1]

The clinical approach to the sacral chordoma is always very delicate due to the clinical polymorphism.

The sacral location with extension to the retrorectal space is exceptional, characterized by the absence of a specific clinical picture; This phrase from MABREY illustrates it perfectly "There are no clinical signs characteristic of chordomas" [2].

The diagnosis is very late due to clinical latency. The diagnostic delay is often long : 27 months for the Mayo Clinic series with extremes of 1 to 18 months; 2 years on average for the literature review of SCIUBBA and AL [3] .

The positive diagnosis is based on digital rectal examination which shows a fixed, rough, and firm presacral mass.

Confirmation is done by CT and MRI. While the precise lesion diagnosis is only made on histopathological examination after complete surgical excision of the tumor.

The treatment is always surgical and which must consist of a total excision of the tumor as much as possible in order to avoid complications and exceptional cases of recurrence. Several approaches are proposed, the choice of which depends on the size and degree of invasion of the lesion [4] .

Radiotherapy intervenes as a complement to surgery, immediately or in the event of local recurrence, or even, when surgery is impossible. However, surgery of the sacral chordoma is difficult because of the anatomical reports of the retrorectal space, exposing the occurrence of per and postoperative complications dominated by hemorrhage, neurological lesions and lesion of the external sphincter.

The sacral chordoma is characterized by its malignant potential at high risk of local and locoregional recurrence, on the other hand the metastases are late and often occur several years after the initial presentation [5].

**Conclusion :**

The analysis of this observation and the data from the literature allows us to retain the rarity of this tumor, its potential for regional recurrence and its uncertain prognosis despite any therapeutic outcome.

**Conflicts of interest :**

The authors do not declare any conflict of interest.

**Authors' contributions :**

All authors contributed to the conduct of this work. All authors also state that they have read and approved the final manuscript

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