



Posterior Approach for Total Resection of a Sacral-Coccygeal Chordoma: A Case Report

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Abstract

A young patient refers progressive worst of lower back pain that became localized in the sacral area getting worst while sitting or standing. On clinical examination, a small area over the coccyx gave painful sensations on pressure, VAS scale was 7. Images from RMN showed a well defined coccygeal mass that demonstrated a low signal on T2, bright signal on T1 and a disomogeneous structure with enhancement. Destruction of the coccyx without invasion of the gluteal musculature and rectum were present and confirmed by Ct scan. The incisional biopsy was performed to make a certain diagnosis of chordoma. Patient was undergone to radical excision from a posterior approach. The lesion was excised en bloc with the coccyx and fourth to fifth sacral vertebra with at least 2 cm healthy surgical margins. Combined use of post-operative high let radiation to increase disease-free interval was used. Follow-up was performed by MRI at 40 and 70 days after surgery showing no chordoma recurrence. Effective management of coccygeal chordoma requires early diagnosis, accurate preoperative staging, definitive and adequate surgical resection with proved tumor-free cut margins and closed follow-up.

Keywords: Chordoma; Coccygeal; Sacral; Posterior; Resection

Introduction

Chordoma is a rare, low-grade, slow growing primary malignant tumor arising from primitive notochord remnants of the axial skeleton.

It accounts for 1-4 % of all primary skeletal tumors and its incidence rate is inferior to 0.1 per 100.000 inhabitants per year.

The most common location is the sacrococcygeal region (40-50%) and the base of the skull (35-40%), followed by the vertebral bodies (15-20%).

Chordoma biological behavior is characterized by a generally slow aggressive local growth with a low tendency in metastasizing to distant sites including the lung, bone, soft tissue, lymph nodes, liver, and skin. Although it is considered to be of low metastatic potential, up to 40-60% of patient are, however, reported to develop distant metastases over the course of their disease.

Adequate wide surgery still remains the cornerstone of chordoma treatment even though safe margins are often hard to obtain because of its anatomical sites of origin.

The achievement of negative surgical margins favourably correlates with the rate of local relapse and survival.

Sacrococcygeal chordomas are insidious tumors that are difficult to diagnose; many patients are treated for an assortment of unrelated diagnoses before the correct diagnosis is made.

Case Report

The patient was a 39 years old Caucasian man who presented himself to his family physician complaining in the last 6-7 years pain when sitting and especially when rising from the sitting position [1-2].

Lower back pain was referred to multilevel lumbar herniated disc already known to the patient.

From 2009 patient refers progressive worst of lower back pain that became localized in the lower gluteal area and sacral area while sitting or after prolonged periods of standing.

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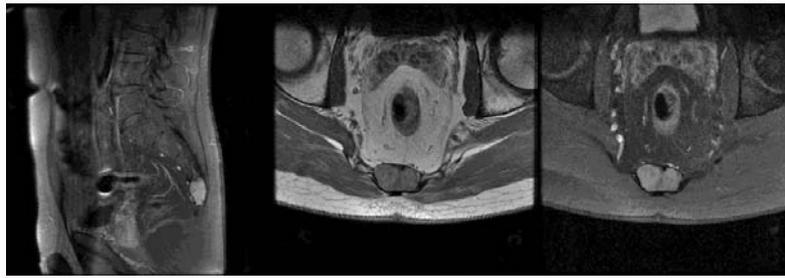


Figure 1: Destruction of the coccyx without insinuation into gluteal musculature and anterior displacement of the rectum without invasion were present.

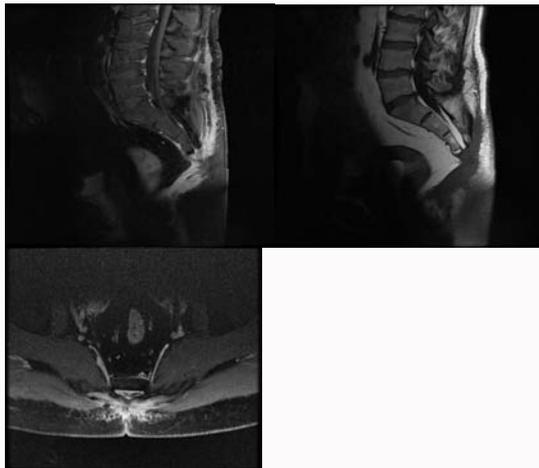


Figure 2: These findings are not correlated to a chordoma recurrence.

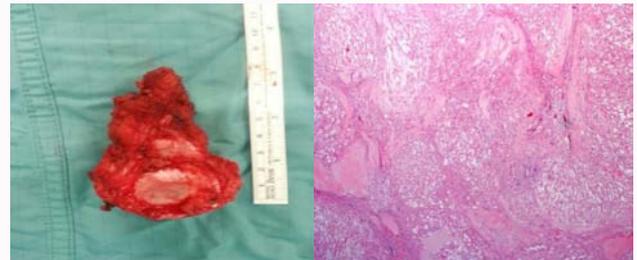


Figure 3: Morphological and intraoperative aspects suggest a diagnosis of chordoma confirmed by istopathologic exams.

Therefore he performs X-rays of the lumbosacral spine and coccyx, and then the patient decides to contact us and arrived to our observation [3].

On clinical examination, a small area over the coccyx gave painful sensations on pressure, not detectable lump in this area could be shown; on visual analogue pain scale (maximum=) -he defined his pain between 7 and 8 before surgical intervention.

He was subsequently evaluated with magnetic resonance imaging (Rmn) and computerized axial tomography (TC) , the images were obtained in axial and sagittal planes using various T1 and T2 - weighted sequences.

Images from RMN showed a large, well defined, lobulated mass (measuring about 3x6x2 cm), identified at the coccyx. Destruction of the coccyx without insinuation into gluteal musculature and anterior displacement of the rectum without invasion were present (Figure 1).

On MRI without enhancement the lesion demonstrated a low signal on T2 -weighted images and bright signal on T1 waiting. On RMI with enhancement the lesion demonstrated disomogeneous structure.

Unfortunately TC scan was performed only in the axial plane [4]. The exam confirmed partial sacral (S5) and total coccygeal invasion; in particular the images showed anterior and posterior coccygeal cortical interruptions.

Hematological and biochemical profiles were within normal limits.

The results of images studies and clinical diagnostic procedures

were discussed in an interdisciplinary groundround composed by orthopaedic and neurosurgeon.

The incisional biopsy was the first surgical procedure made to gain a certain diagnosis [5].

Under antibiotic intravenous coverage of 2 gr cefazolina, using a dorsal approach was performed incisional biopsy at level of the terminal part of coccyx in the interglutealsolcus; a partial skeletrozation let us to expose distal portion of lesion.

Macroscopically lesion has cerebroid aspect, soft texture, and purple color (Figure 3).

Morphological and intraoperative aspects suggest a diagnosis of chordoma confirmed by istopathologic exams (Figure 3).

After three weeks the patient was undergone to radical excision.

The patient lies prone in Kraske’s position; skin median vertical incision was about 20 cm long, from S2 to coccyx; the coccygeal chordoma was excised en bloc with the coccyx and fourth to fifth sacral vertebra [6]. The whole capsule of the tumor was kept intact and at least 2 cm healthy surgical margins were achieved macroscopically. The histopathological report confirmed free surgical margins [7]. The patient recovered uneventfully and was discharged on the fifteenth postoperative day. He was sent to the oncologist for the subsequent treatment and follow-up.

Although chordoma is resistant to the effects of conventional radiotherapy, our institution combined the use of post-operative high let radiation with surgery to attempt to increase disease- free intervals [8].

Follow -up was performed by MRI 40 days after surgery. It showed fluid collection in the surgical site with irregular margins, soft tissues enhancement. Probably these reports were compatible with fibrous scar tissue development in the surgical area.

Next MRI made at 70 days after surgery displayed fluid collection

resolution with persistent soft tissue enhancement due to scar tissue remodelling [9]. In the sub-coccygeal area, on the pelvic plane, death tissue assumed nodular aspects (7, 3 x 4, and 5) with irregular margins. These findings are not correlated to a chordoma recurrence (Figure 2).

Conclusion

Chordoma account for less than 5 % of all bone tumours and it's observed especially among 30 and 60 years with a predilection for male sex (12). Due to its ectodermal origin, chordoma is not properly a sarcoma even if it has clinically retained and classified as such being a primary tumor of bone.

Its histological assessment is often delayed due to non typical signs and symptoms of disease with a frequent clinical diagnosis of pelvic or vertebral and irradiated pain due to discogenic or a specific pathology.

Histology shows multiple lobules of abundant myxoid matrix with cords, strands, and solid nests of phylaphorous cells, which show enlarged atypical nuclei and eosinophilic cytoplasm with variable sized vacuoles. Dedifferentiation and development of high grade sarcoma like areas are possible.

Some of the sacrococcygeal region located pathologies are chordoma, giant cell tumor, neurofibroma, teratoma, metastasis, mixopapillaryepandimoma, myeloma, osteoblastoma, aneurysmal bone cyst, lipoma, osteosarcoma and condrosarcoma, anal fistulas sacral dermatoids and microtraumas, post-partum lesions.

The slow modality of biologic growth, associated to a relatively low incidence of metastatic spread makes surgery the primary treatment of this rare bone tumor [10-11].

The extension of margins is, in fact, a very important prognostic factor being correlated with the incidence of local relapses and overall survival. Local recurrence was significantly associated with an increased risk of metastases and tumor-related deaths.

Conventional radiotherapy with high energy photons is poorly active and need, moreover, to be delivered in doses as high as 60-65 Gy. It may offer some temporary benefit in disease control in patients with inadequate surgery (ie close or positive margins) or, as exclusive treatment, in patient with unresectable /inoperable disease.

Sensitivity to chemotherapy is very low and generally reported in the small subgroup of patients with high grade dedifferentiated chordomas with agents active in high grade sarcomas.

In the experience of Maggiore Hospital of Bologna (Italy), on 52 patients treated over a period of fifty years until 2002, only 20% of patients operated on with safe margins had a local recurrence with a relapse free survival of 56-94 months. This percentage has grown up to 100% of patients who underwent radiotherapy alone or inadequate surgery with intralesion margin with the detection of local recurrence

within 17-20 months from primary surgery.

Although potentially curative, a margin free "en bloc" resection is often very hard to obtain due to the anatomical sites of origin of chordoma ie skull base, spine, and sacro-coccygeal area.

Considering the above situation, we conclude that effective management of coccygeal chordoma require

early diagnosis, accurate preoperative staging, definitive and adequate surgical resection with proved tumor-free cut margins and closed follow-up.

To conclude, despite the progress of current surgical techniques and some encouraging results with the use of targeted therapy, disease control and long term patients prognosis are still poor and chordoma results generally, in a long-lasting life affecting disease.

Nevertheless, specific experience of the multidisciplinary team (surgeons, medical oncologists, radiotherapists, pathologists, radiologists) is a very important pre-requisite in succeeding to improve patients' quality of life and, hopefully, outcome.

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