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Ischio Rectal Chordoma: A Case Report and Literature Review

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Abstract

Ischio rectal chordoma is a rare bone tumor. The optimal therapeutic strategy for ischia rectal chordoma remains surgery. This case describes a 32-year-old adult with ischio rectal chordoma. Given the potential impact on quality of life and the technical platform available, the patient did not undergo tumor resection. The patient was treated with exclusive radiotherapy. The patient developed pulmonary metastases three months after the end of radiotherapy. This case highlights a minimally invasive strategy for the treatment of these rare tumors. The radiographic and histopathological features of chordomas are also reviewed, and the various treatment options reported in the literature are discussed to give another point of view on the management of this rare pathology in low-income countries where the technical facilities are unavailable.

Subject Areas

Oncology

Keywords

Chordoma, Ichio-Rectal, Radiotherapy

1. Introduction

Chordoma is a rare malignant tumor that accounts for 8.4% of all primary malignant bone tumors. It develops from the remnants of the notochord [1], an embryonic structure that forms the axis of fetal development and is involved in the development of the spinal column.

Around 40% of chordomas form in the base of the skull (the clivus) and the

craniocervical hinge 15% in the bones of the mobile spine and around 50% in the sacrum. These tumors are often pauci-symptomatic, and the main symptom of a sacral chordoma is pain. Optimal treatment of chordomas is based on wide surgical resection.

The primary purpose of this case report is to describe how we managed this rare disease exclusively with radiotherapy, and how it evolved in our socio-economic context.

2. Observation

We report the case of Mr. L.A., aged 32, with no notable pathological history, who developed a painful swelling of the right gluteal region 4 months prior to his consultation, aggravated by the onset of dysuria, evolving in a context of altered general condition, which prompted his consultation.

On physical examination, the patient had a swelling of the right gluteal region that was painful to palpation, with no inflammatory signs, and the inguinal-sus clavicular lymph nodes were free (**Figure 1**).

Soft tissue MRI revealed an ischio rectal tumor process infiltrating the gluteal muscles and subcutaneous soft tissue, measuring 15 × 18 × 13 cm (AP × T × H), suggesting a sarcomatous origin (**Figure 2**).

The patient underwent a biopsy of the tumor process described above, the results of which showed a poorly differentiated myxoid tumor process with round cells, initially suggesting a sarcomatous origin, immunohistochemical complement showed diffuse tumor cell expression of cytokeratin and EMA (epithelial-membrane antigen), no expression of CDX2, no expression of CK7, TTF1 with Ki67 at 10%, suggesting a chordoma.

The patient's case was discussed at a multidisciplinary consultation meeting, with the decision to be referred for radiotherapy, given the unresectable nature of the tumor process due to the lack of technical facilities in our context.

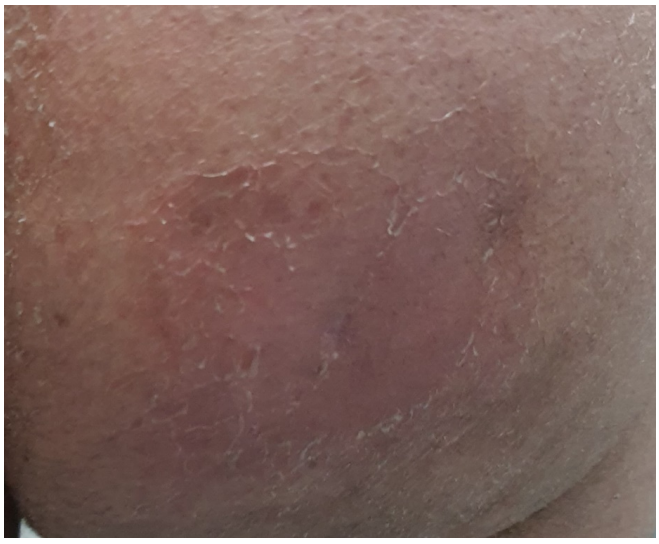

Figure 1. Appearance of the skin opposite the tumour on the right buttock.

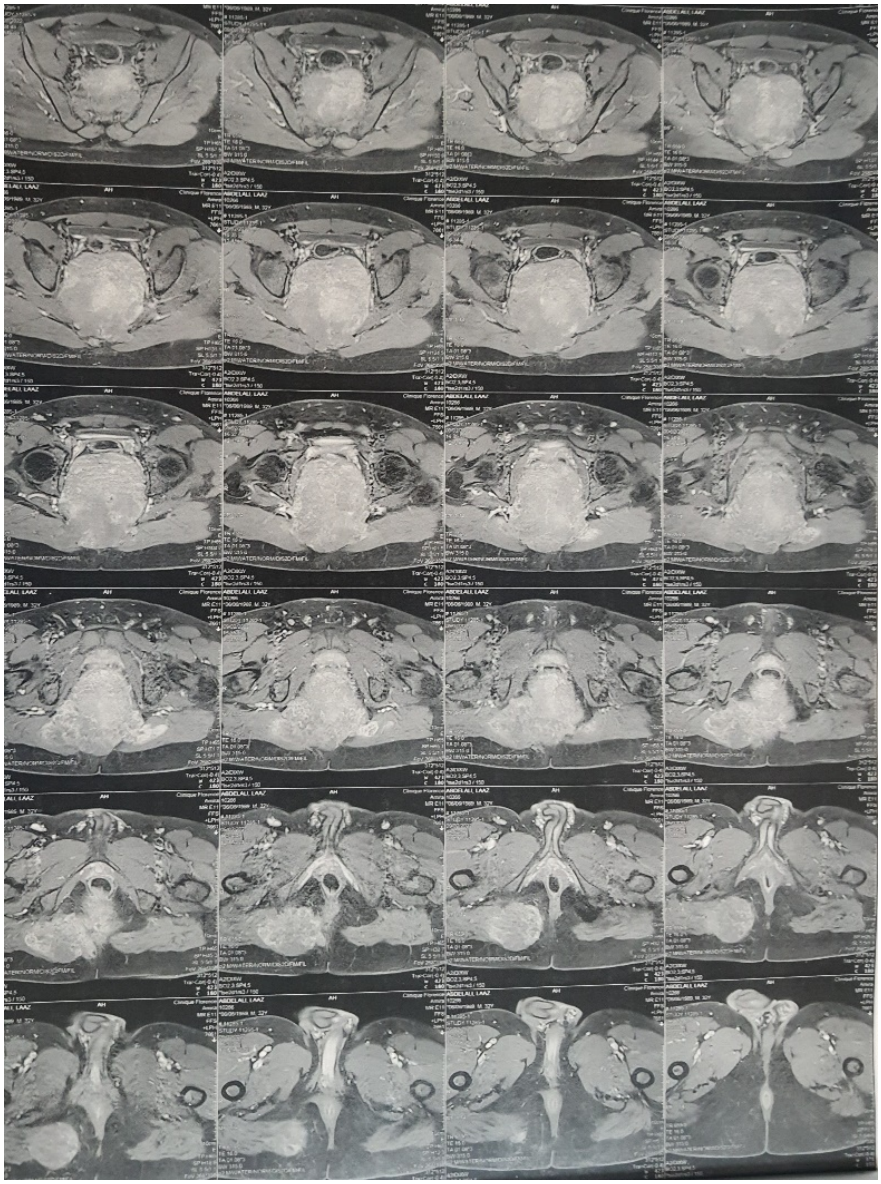

Figure 2. MRI images of our patient's tumour process.

The therapeutic strategy, the course of the radiotherapy sessions and any side effects of the treatment were explained to the patient. The patient underwent a centring CT scan, positioned in a comfortable, reproducible supine position, with hands on the chest and knee and callus supports, aligned with anatomical markers using 2 lateral and 2 transverse wall lasers, and 3 mm sections were acquired from the D11-D12 interline to the upper two-thirds of the femur in two C- and C+ steps, followed by transfer of the images to the TPS.

The patient underwent 3D external radiotherapy with photons of 10 to 18 Mev energy, using 4 beams with a total dose of 66 Gy in 33 fractions of 2 Gy per fraction over 37 days. During radiotherapy, the patient developed grade II radiodermatitis, which was treated symptomatically with good evolution.

Three months after the end of radiotherapy, the patient reported a marked

improvement in the initial symptomatology. He underwent an evaluation CT scan, which showed that the voluminous sacrococcygeal tumor mass measuring 13 × 17 × 12 cm (AP × T × H) was stable in size. The patient showed 3 secondary pulmonary nodules put by the medical oncologists on imatinib 800 mg daily with overall imaging stability.

3. Discussion

Chordoma is a very rare tumor. Its incidence varies from 0.5 to 8 per million inhabitants per year [2]. The sacrococcygeal region is the preferred site (50%). The spheno-occipital region accounts for 35% of cases. The cervical and dorso-lumbar spine is affected in only 15% of cases, while exceptional localizations have been described, notably in the orbit, sinuses or scapula [3].

Since MULLER in 1858, chordomas have been associated with their notochordal origin. Although they were initially considered to be of cartilaginous origin, confusion persisted for a long time between chondrosarcomas and chordomas. Today, the difference has been clearly established [4]. The notochord is a rod-shaped element that defines the primitive axis of the embryo, and is replaced by a bony skeleton that becomes the spinal column and part of the base of the skull. However, part of the notochord may persist in the form of vestigial islands, the degeneration of which gives rise to the chordoma.

The clinical approach to sacral chordoma is always very delicate due to its clinical polymorphism, and it is often unrecognized and belatedly diagnosed because of its location and the mass effect it exerts on neighboring organs [5]. Its sacral location with extension to the retro rectal space is exceptional, characterized by the absence of a specific clinical picture; MABREY's phrase illustrates this perfectly: "There are no clinical signs characteristic of chordomas" [6]. They can manifest themselves with a wide variety of clinical signs: pain, urinary signs, digestive signs or neurological signs [7]. Many of these patients had been treated for years for symptoms with NSAIDs, laxatives or antibiotics, as well as steroid injections by general practitioners [8].

A CT scan is the diagnostic confirmation test par excellence. It enables the diagnosis to be made in 100% of cases, even in the case of very small tumors that cannot be palpated by rectal examination. It provides precise localization of the tumorbed and can also determine the nature of the mass by measuring its density, assessing its character, topography and boundaries, and possibly its local and distant extension. [9]. CT is of great value both in the preoperative workup and in monitoring the progress of the tumor, as it perfectly locates the site of the tumor [7].

In addition to CT, pelvic MRI is highly accurate topographically, particularly for soft tissue, epidural and intradural space [10], and on T1 and T2 sequences in sagittal and axial sections, it can be used to assess whether the lesion is liquid (with intense hyper-signal in T2 sequences) or solid, its contiguity with the rectum and its limits, and to determine the extent of the tumor, particularly in the

upper region, and consequently the best surgical access, particularly on sagittal sections. MRI is also more sensitive than CT for studying relationships with nerve roots and the existence of any embracement, while angiograms and venograms provide a clearer picture of vascular relationships. MRI with gadolinium injection before and after neoadjuvant treatment enables tumor volume to be checked [11]. On MRI, sacrococcygeal chordomas are tumors with hypo- or iso-signal on T1-weighted sequences (compared with muscle masses), and heterogeneous hypersignal on T2-weighted sequences. Chordoma is lobulated in appearance, with extension into the sacral and epidural spaces [11].

MRI consistently shows tumoral lobulation, particularly in T2-weighted sequences and in T1-weighted sequences after gadolinium injection (enhancement of tumoral septa, much better visible than in post-injection CT), underscoring the value of this examination in the positive diagnosis of sacrococcygeal chordoma.

Diagnostic certainty is based on anatomopathological examination, which is a fundamental and indispensable element in the diagnosis of chordomas.

Macroscopically, the tumor is generally lobulated, of soft gelatinous consistency, with occasional firmer areas of cartilaginous consistency, of greyish or bluish-white color, but there may be recent or old hemorrhagic recurrences of ochre appearance. Tumor lobules may be well delineated when the tumor invades soft tissue, but the boundaries are often more indistinct in the bone itself. In the case of cortical rupture, the tumor often remains covered by the periosteum. This is particularly true of sacral tumors, which are removed as a whole; cervical and basal localizations are more often highly fragmented [10].

On immunohistochemistry, chordoma is the only tumor to show positivity for epithelial and connective markers, as well as for S100 protein. Pancytokeratins and cytokeratin 19, EMA (epithelial-membrane antigen) and vimentin are almost constantly expressed, but sometimes only focally, which can be problematic on a small biopsy. Brachyuriais a more recent marker, highly specific for chordoma, although it is also expressed by hemangioblastomas and some testicular tumors.

Its sensitivity is high, estimated at 90.2% in a study of 51 chordomas [10], but immunoreactivity may be lost after decalcification, as is common with nuclear staining. In dedifferentiated forms, these different markings are lost: diagnosis relies on the coexistence of recognizable chordoma sectors, or on the clinical history.

In our case:

- Diffuse tumor cell expression of cytokeratin and EMA
- Diffuse expression of PS100
- No CDX2 expression
- CDK4 shows moderate nuclear and cytoplasmic expression
- No expression of CK7
- No TTF1 expression
- Ki67 is expressed by less than 10% of cells

The treatment of chordomas is primarily surgical; however, complete excision can only be achieved in 60% - 70% of cases, due to the proximity of neurological structures whose involvement would be a source of post-surgical complications [12].

Radiotherapy can be used either as adjuvant therapy after surgery, or exclusively in cases of local recurrence, or even when surgery is not possible [13]. In view of the high relapse rates obtained after exclusive surgery, adjuvant photon irradiation was initially proposed, but progression-free survival was always less than 40%. The proximity of organs at risk to the tumor and the ballistic properties of photons often prevented the delivery of doses more than 60Gy to the tumor, which had a direct impact on local control [8].

Chordomas vary in radiosensitivity, but in most cases, they are radio-resistant tumors [14]. This raises the question of whether adjuvant radiotherapy is indicated, after R1 and R2 surgery, at a dose of 50 to 60 Gy over 5 to 6 weeks. Radiotherapy is sometimes indicated exclusively and palliatively, in the case of very large tumors that cannot be operated on. An initial series of external irradiation for decompressive and analgesic purposes is delivered up to a dose of 50 Gy over 5 weeks. In the event of a good tumor response, as assessed by a follow-up CT scan, a further 20 Gy of external irradiation can be administered over 2 weeks.

Exclusive external irradiation only exceptionally leads to complete tumor destruction, but often results in good comfort and, above all, a good analgesic effect, as in our patient's case. Given the possibility of metastasis, chemotherapeutic combinations have unfortunately been tried without success [15]. Thanks to the ballistic characteristics of protons, proton therapy increases the dose to the tumor and spares nearby critical organs as much as possible [8]. This physical feature is fundamental in explaining the dose gradient that can be obtained close to a critical organ. The dose varies from 10% to 15% per millimeter of tissue crossed [16]. It was then shown that local control was improved and the risk of toxicity acceptable in a large number of series. This is why, for several years now, proton irradiation has become the reference irradiation technique for the treatment of skull base chordomas after surgery [16]. The use of carbon ions represents an interesting modality. Indeed, they have the physical advantages of protons (Bragg peak) and relatively superior biological efficacy, which is of interest for radio-resistant tumors. Promising results have been obtained in Japan and Germany in terms of both efficacy and toxicity [14] [17].

The prospects of adjuvant chemotherapy or antiangiogenic therapy are still being evaluated [9]. In practice, the chemoresistance of these tumors is well recognized, although positive and isolated results have been reported with anthracyclines, alkylating agents, cisplatin and thalidomide [2] [18]. Imatinib alone or in combination with sirolimus has also shown some efficacy in terms of local control in PDGF-expressing chordomas [18] [19]. Phase II therapeutic trials using imatinib have shown an active clinical and radiological response in locally advanced forms [10]. The average survival time of untreated patients is estimated at 28 months from symptom onset. The time to recurrence after surgery,

whether or not followed by irradiation, varies from two to three years, but can be as long as ten years [8].

In Erikson's series of 48 patients (23 sacral chordomas, 20 skull bases, 5 spines), 35 non-operated, 11 operated, 2 metastatic, patients received external radiotherapy at a dose of 50Gy over 25 fractions. The mean survival in Erikson's series was 6.6 years for the radiosurgery group, compared with 5.7 years for surgery alone and 5.4 years for radiotherapy alone.

In our case, after 1 year from the end of radiotherapy, the patient is still alive with less pain and clinical and radiological stability of the tumor and secondary pulmonary localizations.

4. Conclusions

Adult sacral chordomas are a rare entity. They develop from embryonic residues and notochordal remnants. Diagnosis is very late, due to clinical latency. They often go unnoticed; they may be discovered by chance or as a result of a mechanical complication. Treatment is always surgical, involving total excision wherever possible to avoid complications.

Radiotherapy is used as a complement to surgery, either immediately or in the event of local recurrence, or even when surgery is impossible. Advances in radiotherapy have led to improved survival.

In our context exclusive radiotherapy remains as sub-treatment but may be the only treatment that can be considered to preserve our patient's quality of life.

Conflicts of Interest

The authors declare no conflicts of interest.

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