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Chordoma of the Cervical Spine: A Case Report and Review of Literature

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Authors' contributions

This work was carried out in collaboration between all authors. Author MC was responsible for the coordination of the overall study, including: The study design and manuscript preparation. Authors MC, YD and MT managed the analyses of the study and the literature searches. Authors MC, HA and BB were responsible for manuscript review and study design. All authors read and approved the final manuscript

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Case Study

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ABSTRACT

Chordomas are rare low to intermediate grade malignant tumours derived from the notochordal remnants. Their location in the cervical spine is exceptional. The clinic radiologic features, anatomopathologic aspects, diagnostic difficulties, therapeutic methods and prognostic factors are discussed through a case report of a cervical spine chordoma with a literature review.

Keywords: Chordoma; notochordal tumor, spine tumor; cervical tumor; physaliphorous cells; complete resection; local recurrence.

1. INTRODUCTION

Chordomas are rare primary malignant tumors of the bones that recapitulate notochord. They can be found anywhere from the base of the skull to the coccyx. The most common locations are in the clivus and sacrum and only infrequently in the intervening vertebral column.

In the light of a case study of a cervical spine chordoma and a literature review, clinical aspects, imaging features, histological

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characteristics, therapeutic principles and prognosis of this tumor will be discussed.

2. PRESENTATION OF CASE

Mr. A. a 53-year-old man, without medical history, admitted for a chronic and isolated left neck swelling, gradually increasing in volume. Clinical examination revealed a hard swelling, painless, movable relative to the skin but attached to the deep planes, this mass was extended up to the submandibular region and down to supraclavicular region and measured 15 cm / 6 cm. The overlying skin was normal. The rest of the ENT examination especially endoscopic examination of the upper aerodigestive tract were normal. Neurological examination had found a limitation of elevation of the left upper limb. Imaging initially included a cervical ultrasound has objectified a multi-partitioned mass with hypoechoic content. The magnetic resonance imaging (MRI) revealed a huge process with hypersignal SET2 (Fig. 1), hyposignal T1 (Fig. 2), strongly enhanced after gadolinium injection (Fig. 3), and developed in the posterior left lateral cervical space causing displacement, forwards and inwards, of homolateral internal jugular vein and carotid artery. Back, the mass contracted an intimate relationship with the anterolateral parts of the vertebral bodies and the cervical spine pedicles at C4 and C5, with cortical bone erosions and intracanal extension, displacingthe cervical spinal cord to the right.

Trocar biopsy was performed but histological study was not conclusive, so a cervicotomy was decided with a latero-cervical incision. Intraoperatively, the mass was greyish, firm to friable, very adherent to the cervical vertebrae and inextirpablein totality. The surgical specimen was sent for frozen section and the answer was in favor of achordoma. The excision was therefore performed at the maximum of our possibilities with help of neuro-surgeons. The final pathological study confirmed the diagnosis, showing at the cyto-histological examination, physaliphorous cells with eosinophilic and vacuolated cytoplasm observed on a background of myxoid matrix and grouped into cords (Fig. 4), and at the immuno-histo-chemical study, the tumor positivity for anti-PS100, anti-EMA, anti-vimentin and anti-pan-keratin (Fig. 5).

An adjuvant postoperative radiotherapy has been decided and the patient was addressed to the radiotherapy department but since then, he is not seen in consultation for control.

3. DISCUSSION

Chordomas are rare low to intermediate grade malignant bone tumours derived from the remnants of the embryonic notochord. They account for 1-4 % of all primary malignant tumours and arise along the midline axial skeleton. Chordomas are most commonly found in the sacro-coccygeal region (60%), spheno-occipital region (25%) and in the mobile spine (15%), in descending order of frequency. Only 10% of all chordomas are developed in cervical spine. This neoplasm is most commonly presents after age 30 with a peak in the sixth decade (30 %), and is very rare under age 20 (1%). The sex-ratio male/female is 1.8:1 [1,2].

Clinically, pain and neurologic deficits specific to the site of origin are common symptoms, and some tumors, especially those arising in the sacrum or the mobile spine, may be present for years before they are diagnosed. In chordomas arising in the low or high cervical spine, the pain with or without brachialgia is caused by bone destruction, nerve compression or by an increased mobility of the vertebral segments. Posterior growth will lead to radicular or even medullary compression with paraparesis or tetraparesis. Anterior growth will take to loco-regional compression with cervical or parapharyngeal mass, dysphagia and / or dyspnea [2,3,4].

Radiologically and on CT-scan, chordoma is typically solitary, central, destructive lesion of the axial skeleton and is almost always associated with intra-tumoral foci of calcification and a soft tissue mass predominantly located anteriorly or laterally to the spinal column. MRI studies best visualise soft tissue extension and its relationship to anatomic structures. On MRI, T1 weighted sequences are hypo- or iso-intense, while T2 weighted sequences are of high signal intensity. Other caracteristics include contrast enhancement after gadolinium injection, and septated areas of low attenuation [2,5,6].

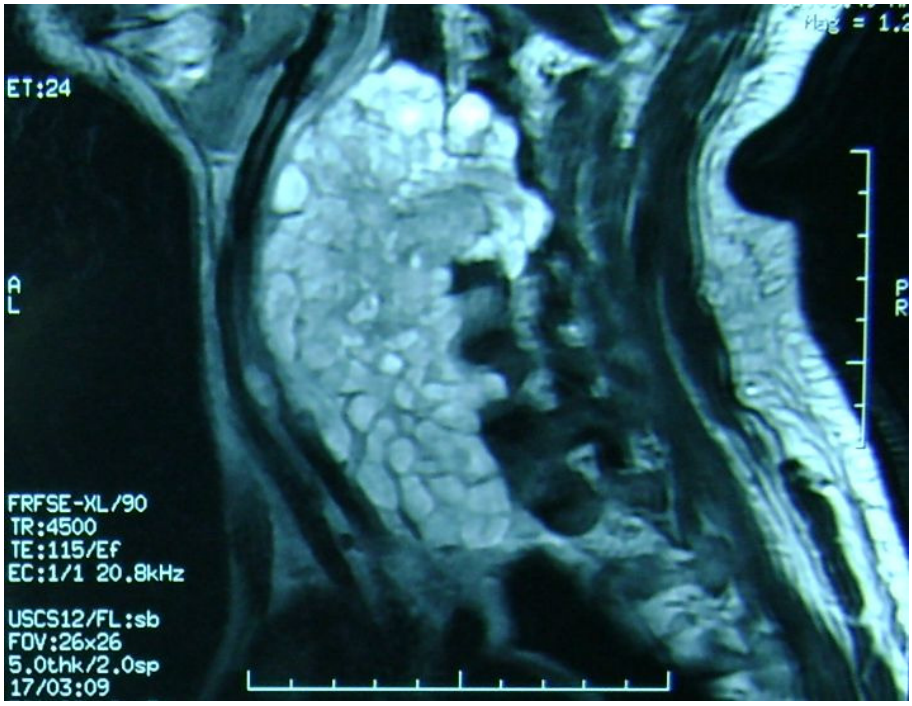

Fig. 1. Sagittal T2-weighted MRI showing hyperintense huge cervical mass with an intimate relationship with the vertebral bodies and an intracanal extension

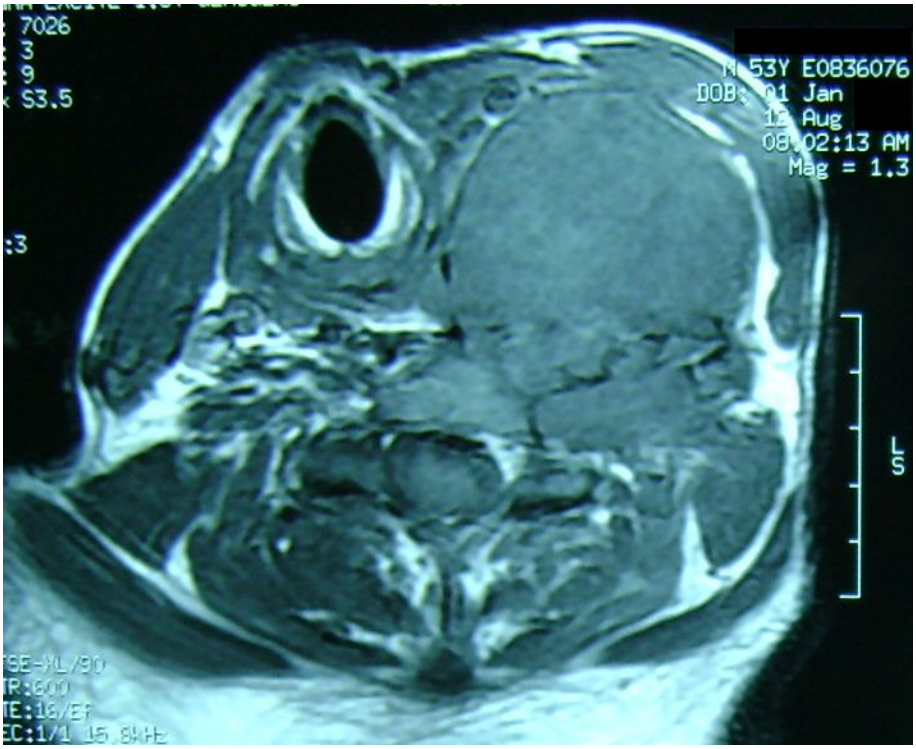

Fig. 2. Axial T1-weighted MRI showing hypointense mass in the posterior left lateral cervical space with an intracanal extension displacing the cervical spinal cord to the right

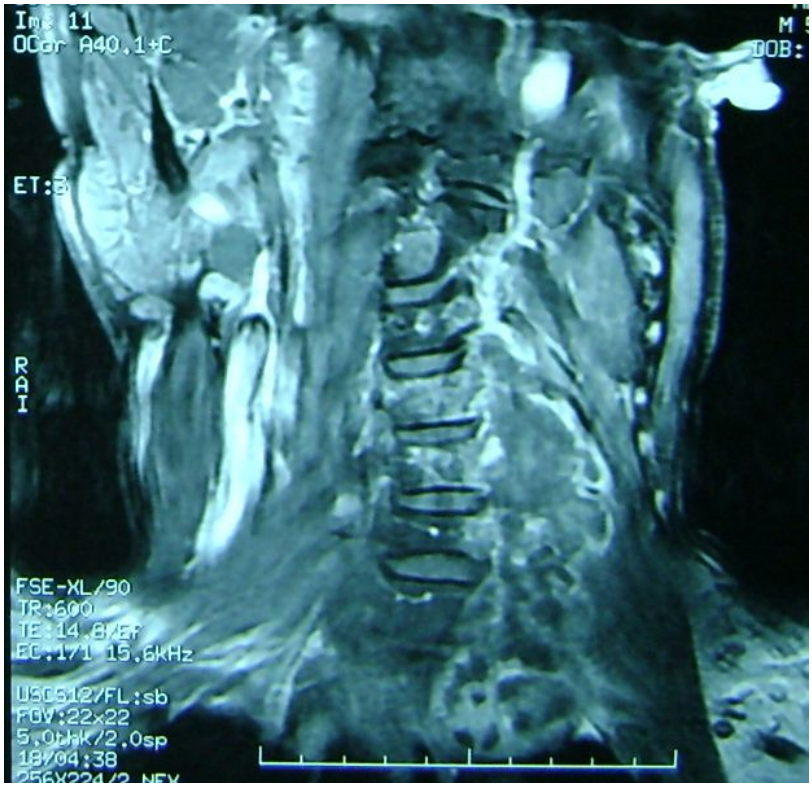

Fig. 3. Coronal T1-weighted MRI showing mass enhancement after gadolinium injection

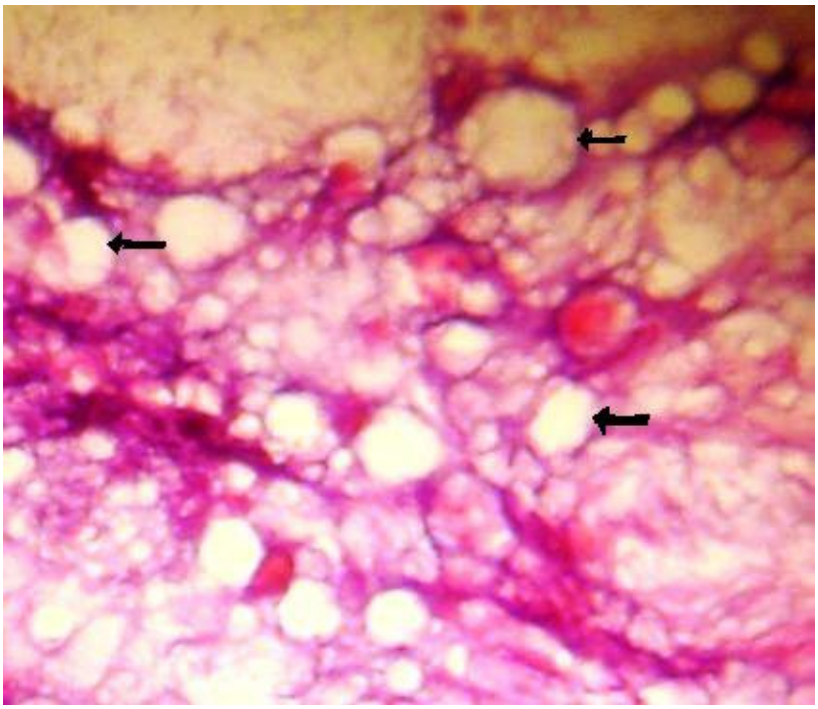

Fig. 4. Cyto-histology showing physaliphorous (arrows) cells with eosinophilic and vacuolated cytoplasm observed on a background of myxoid matrix and grouped into cords

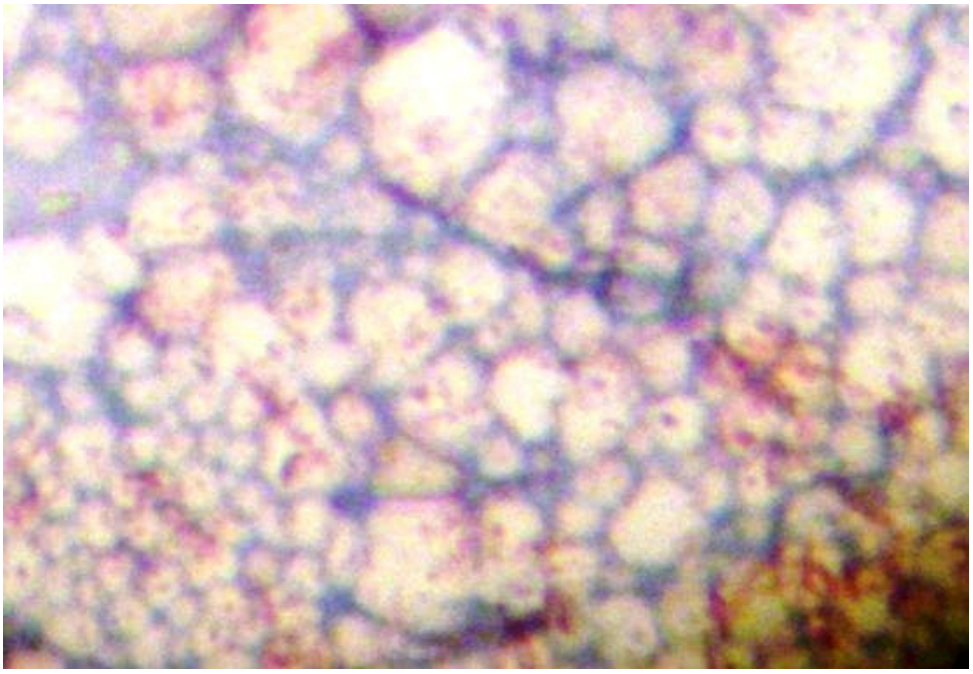

Fig. 5. Immunohistochemistry showing expression of cytokeratin

On gross inspection, chordoma is a greyish tan, glistening, gelatinous to friable and lobulated mass. In most cases, it is associated with extension beyond the contours of the bone into the surrounding soft tissues. The tumours vary in size, generally from 5 to 15 cm. Histopathologically, chordoma is classified into the conventional (the most frequent), chondroid (the most common in children) and dedifferentiated variant (the less frequent). A component of the conventional type is always present in the others variants. Chordoma is a lobulated neoplasm, with lobules separated by fibrous bands. The tumour cells, arranged in sheets or cords within an abundant myxoid stroma, have an abundant pale vacuolated eosinophilic cytoplasm. These cells are known as "physaliphorous cells" and approve the notochordal origin of the tumor. Mitoses and moderate nuclear atypia are infrequent and objectifiable with difficulty. In the chondroid chordome (approximately 14% of all chordomas), there are areas that may mimic hyaline or myxoid cartilage. In the dedifferentiated variant, called sarcomatoid chordoma (less than 5% of all chordomas), there is an association with a high grade sarcoma. Immunohistochemical studies show a positive reaction of chordomas with antibodies against PS100, pan-keratin, vimentin and EMA [1,7,8,9,10].

Some differential diagnoses should be considered in the diagnostic process. Firstly, chordomas are sometimes misdiagnosed as metastasis of carcinomas, essentially renal cells carcinoma. Glandular or acinar architecture, vascular wealth, and lack of physaliphorous cells, incite to carry those diagnoses. Secondly, chordoma may be confused with chondrosarcoma. For the latter, architecture is diffuse and immunostaining directed against cytokeratins are negative. Finally, benign notochordal lesions, including benign notochordal cells tumors and ecchordosis pysaliphora, are an important entity in the differential diagnosis of chordoma, since they have the same cells physaliphorous, signing their common notochordal origin, and the same immunohistochemical characteristics. The absence of clinical and radiological signs of aggression, lack of enhancement after injection of gadolinium in MRI and absence of mitosis and nuclear atypia at cytologic study straighten the diagnosis, knowing that, in some exceptional cases, both types of lesions may coexist in the same tumor, supporting the theory of malignant transformation of some benign notochordal lesions to chordoma [3,11,12,13].

A preoperative assessment, combining CT-scan and MRI, is essential to clarify the tumor extension and plan the surgical procedure. CT-

scan is better than MRI in the precision of bone involvement. MRI is unquestionably superior to CT-scan as regards the contrast between tumor mass and soft tissue. Sagittal T2 weighted MRI is good at showing tumor extension into the spinal canal. CT or MRI angiography can provide sufficient information on vascular reports. In case of large tumor or recurrence, it is useful to look ganglion extensions and distant metastases, which are mainly through the bloodstream, particularly in the lung and liver. Other secondary locations can be searched based on call signs [2,3,14].

Surgery is the mainstay of treatment for chordomas. The goal of surgery is to remove as much of the tumor as possible without causing unacceptable damage. Oncologic total resection ("en bloc" resection), when technically feasible, with margins of healthy tissue provides the best chances for local control and long-term survival. The endolesional curettage is the second method. It involves removing the entire tumor tissue as completely as possible. This curettage must be careful, even if it is not satisfactory carcino logic, hoping to reduce the speed of recurrence. This treatment by curettage should be reserved for very elderly patients or patients whose tumor volume does not allow adequate resection. Currently, according to some authors, multilevel oblique corpectomies seem to be an effective and safe surgical technique to treat a cervical chordoma when there is a precarotid and retrocarotid extension, essentially in young patients. For chordoma of the cervical spine, the surgical approach is anterolateral (sterno-cleido-mastoid) if tumor is limited to the vertebral body, and it is double, anterior and posterior, if there is extension to the posterior arch. Reconstruction and stabilization of the cervical spine after resection, requiring collaboration between ENT and neuro-surgeons, may be necessary and appeal to anterior cervical fusion with bone graft and internal fixation plate.

The post-operative radiotherapy appears to improve survival, especially in the case of proton beams therapy allowing delivery of a larger radiation dose to the tumor with a minimal dose to surrounding tissues. This is very important when radiating around critical structures such as the optic nerves, brain stem and spinal cord. However, the risk of recurrence appears to be related to only quality of resection. In case of recurrence, surgery is indicated if the tumor appears resectable. Embolization can be envisaged as palliative and analgesic treatment when the possibilities of surgery are exceeded. However, there is no need to renew an irradiation treatment in patients with active pursuit, because of the risk of life-threatening complications [2,15,16,17,18].

The prognosis of chordomas is affected by a variety of clinical, radiologic and anatomopathologic features. The essential characteristics include tumor location, size, and resecability, as well as the age of the patient and the histologic variant. The chondroid chordoma has been reported to be associated with a better prognosis, while the dedifferenciated variant has the worst prognosis of all chordomas and is usually rapidly fatal. In the mobile spine, the 5 year survival rate is approximately 58 %. However, local recurrence rates are very high since the complete excision of spinal chordomas is difficult to accomplish [1,8,15,19].

4. CONCLUSION

Chordoma is a raremalignant notochordal tumor. Its location on the cervical spine is exceptional. CT-scan and MRI are essentials to shed light on the tumor extension and plan the surgery. On the whole, chordomas are slow growing, but tend to recur after treatment. Because of their proximity to critical structures such as the spinal cord, chordomas are difficult to treat and need highly specialized care. Surgical resection represent the essential pillar of treatment. Radiotherapy, notably proton beam therapy, appears to improve survival, but only complete resection of the tumor can offer a low risk of local recurrence.

CONSENT

Not applicable.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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