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Case Report
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Chordoma of the Clivus with Endonasal Extension: A Case Report and Review of the Literature

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Abstract

Chordoma is a rare tumor of the axial skeleton, of notochordal origin, histologically benign with local malignancy. It develops mostly in young adults. The clinical evidence of this localization (clivus) often simulates that of a pituitary tumor or of the locally advanced cavum, it is only the histopathological examination on biopsy which guides the diagnosis and then the care which must be multidisciplinary (Neurosurgery, ENT and Radiotherapy).

Two management modalities are important in the treatment of skull-base chordomas: surgery and radiation therapy, but complete resection remains the only element of good, non-controversial prognosis. We report the case of a patient in charge of the ENT department and head and neck surgery at the Sheikh Khalifa International University Hospital and stress the diagnostic and therapeutic features of this type of tumor.

Keywords: Chordoma; Clivus; Immuno Histo Chemistry; Resection

Introduction

Chordomas are rare tumors of the axial skeleton, histologically benign, but their natural history and evolution under treatment are very similar to those of a tumor with local malignancy [1]. They constitute 1% of intracranial neoplasms, located at the level of the clivus in 25% to 39% of cases [2,3]. Developed from the remnants of the early morning fetal development [4]. This tumor initially poses a diagnostic problem because of its rarity and then therapeutic because of the difficulty of surgical access. Advances in this field have been made thanks to immunohistochemistry and new radiotherapy techniques [5].

Case Presentation

Mr. MM, a 25 years old, with no particular pathological history, was admitted to our department for bilateral nasal obstruction, hyposmia, otalgia and right hypoacusis, with transient headaches and discreet torticollis, all progressively evolving over the last 6 months.

On physical examination, the patient was conscious, apyretic without sensory-motor deficiency, with rhino-cavoscopy a bulky whitish mass lasts in places, filling the cavum and extending to the posterior third of the nasal fossae, not bleeding on contact. A high jugulo-carotid adenopathy was also found on the left side of the cervical examination, the rest of the ENT examination found no abnormalities (Figure 1).

Computed tomographic imaging revealed a lesion process 6 cm long, centered on the postero-superior wall of the cavum, weakly enhanced by the contrast medium, with central necrosis, lysis of the clivus and the body of the sphenoid, filling the cavum and extending to the 2 nasal cavities especially on the left side (Figure 2 and 3). An average jugulo-carotid adenopathy of 2 cm was also found on the right side.

The patient then benefited from multiple biopsies of the mass under general anesthesia twice, which's pathological and immunohistochemical examination was in favor of fibro-inflammatory material without any sign of malignancy. It was only at the 3rd surgical intervention that was programmed (biopsy resumption with panendoscopy and adenectomy) that the diagnosis of chordoma was retained after extensive immunohistochemical study using specific markers (brachyuria, expression of cytokeratin ae1-ae2, PS100 and membrane epithelial antigen) (Figure 4). The anatomo-pathological study of lymphadenopathy did not find any anomaly.

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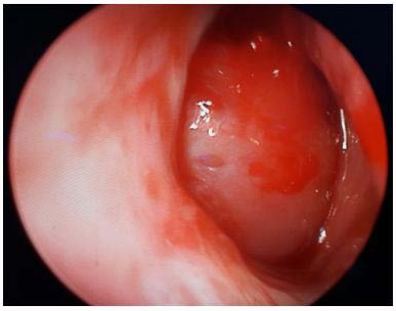

Figure 1: Endoscopic image (rhino-cavoscopy) evoking a mass filling the cavum and the posterior 1/3 of the left nasal fossa.

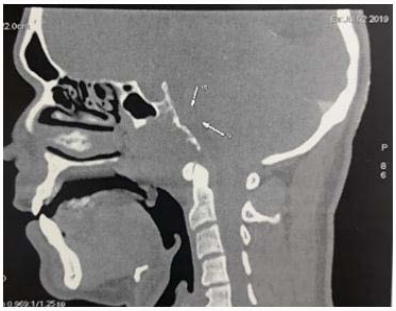

Figure 2: Cranio-facial CT scan in sagittal section, showing a tumor process filling the cavum, with lysis of the clivus and prepontic endocranial extension.

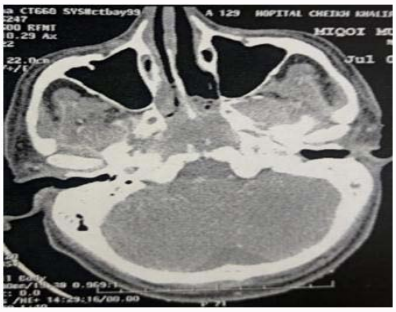

Figure 3: Cranio-facial CT in axial section, showing the endonasal extension of the tumor process.

The patient benefited from a complementary radiological assessment, magnetic resonance imaging, showing precisely the extension of the tumor process, and then transferred to the neuro-surgery department for surgical treatment and complementary proton therapy (Figure 5).

Discussion

Chordomas are rare, slow growing, infiltrative tumors which are most often diagnosed in the second or third decade of life [4]. The chordomas come from the notochordal embryonic vestiges, initially, they were regarded as of cartilaginous origin and that the confusion persisted a long time between chondrosarcomas and chordomas, today the difference is clearly established [1].

Although these remnants may persist anywhere along the axial skeleton, the sacrococcygeal region and the clivus are the most common sites. The clinical evidence of this localization (clivus) often simulates that of a pituitary tumor or cavum with invasion of the base of the skull, it is only the histo-pathological examination on biopsy

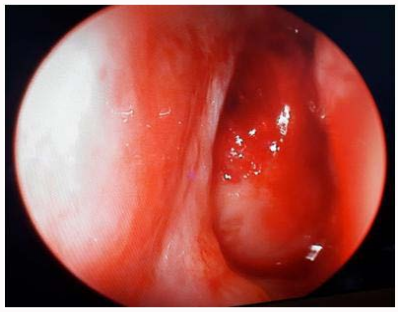

Figure 4: Intra operative image after deep biopsy of the tumor.

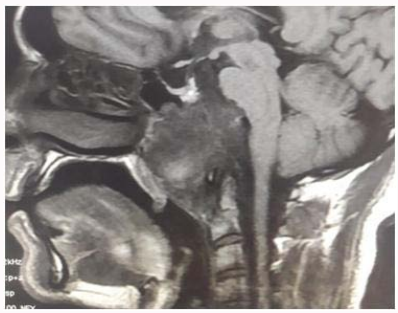

Figure 5: Sagittal section of the magnetic resonance imaging shows the tumor process.

(requiring a qualified team and the availability specific markers) that guide the diagnosis and then the care that must be multidisciplinary (neurosurgery, ENT and radiotherapy). Imaging has an essential place in diagnosis; the chondroma takes on the scanner an aspect of central lytic tumor sometimes lobulated with rare calcifications. MRI makes it possible to better specify the locoregional extension while the PET-CT scanner has a very small role especially in the detection of local recurrences [5]. Two management modalities are important in the treatment of skull-base chordomas: Surgery and radiation therapy [4].

The surgical strategy should be aggressive with good preoperative preparation (checkups and verification of cerebral perfusion quality) and taking into account the patient's age, general condition, desire, and volume and tumor extension. The addition of high-dose irradiation, using particular techniques preserving adjacent functional structures, protons or photons or both associated, is of great interest in the management of this type of tumor [1].

Magnetic resonance imaging should be done routinely one month after surgery. The treatment of recurrent forms after surgery and irradiation is not yet codified. The prognosis of the chordomas of the base of the skull, notably the clivus, seems less severe than that of the chordomas of the craniocervical hinge and the cervical spine [1].

Conclusion

Chordomas are rare, slow growing, infiltrative tumors. They may pose diagnostic problems because of their scarcity, and the unavailability of specific immunohistochemical markers in some centers, which explains the diagnostic delay observed in some cases. The reference treatment of chordomas of the skull base, in particular of the clivus, remains the surgery which must be maximalist, in several times if needed, associated with a complementary proton therapy. The complete resection is the only element of good prognosis

not controversial.

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