P315

THÈME RHINOLOGIE ET ALLERGIE

Chordoma of the nasal cavity: a case report

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Introduuction

- Chordomas are low grade and often malignant tumours derived of the notochordal tissue, they represent 4% of all bone malignancies.
- Most cases occur preferentially in the axial skeleton and the skull base.
- Extra-axial chordomas are exceptional subcategory of its axial counterpart with only 90 cases reported up to 2021.these localizations are explained by the occurrence of extraosseous notochordal rests that have penetrated these sites.
- Diagnosis is made after hisptopathological examination with immunochemical staining. Chordomas show a high expression of cytokeratin 7 and protein S100 and brachyurie, which is a highly sensitive and specific immunochemical marker.
- The main stay in chordomas treatment is ‘’in bloc’’ resection. Unresectable tumors are managed by external beam radiotherapy.
- The 5 years overall survival rate reaches 68.4%

Case report

- A 62-year-old woman with history of nasal myoepithelial tumor sought medical consultation for left-sided nasal obstruction and recurrent epistaxis.
- Endoscopy showed a smooth mass filling the left nasal cavity.
- Computed tomography (CT) : large filling the entire left nasal cavity and adherent to the nasal septum. No bone erosion was found.**(figure1)**
- Magnetic resonance imaging (MRI): well-defined mass in the left nasal cavity with a regular contour displaying heterogeneous hyperintensity on T2-weighted imaging and hypointensity on T1-weighted imaging, without diffusion restriction, and an intense enhancement after contrast injection **(figure2)**
- Treatment and follow-up: An endoscopic sinus surgery was performed. The tumor was resected in one piece The frozen-section examination concluded a malignant tumor. Histopathological examination concluded a chordoma and surgical margins were tumor free.
- Post operative course was uneventful and the patients is free from disease one year after surgery.

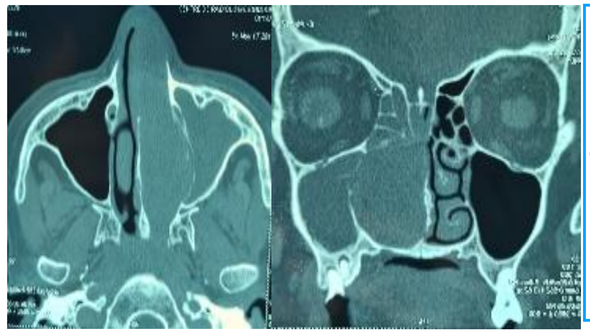

figure 1 : CT (axial and coronal section) of the head showing a mass in the the left nasal cavity

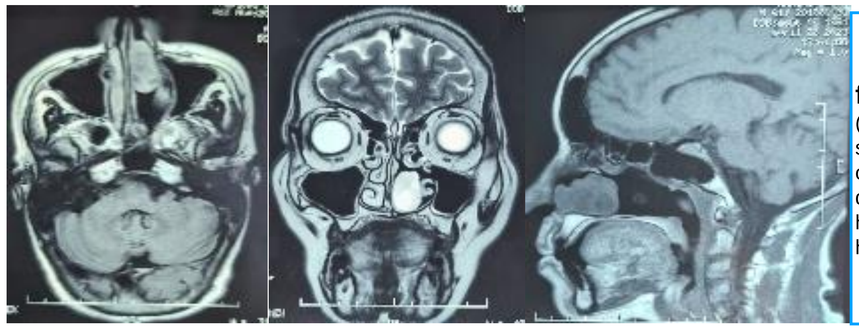

figure 2: facial MRI (axial coronal and sagittal sections) showing a mass of the nasal cavity displaying T2 hyperintensity and T1 hypointensity

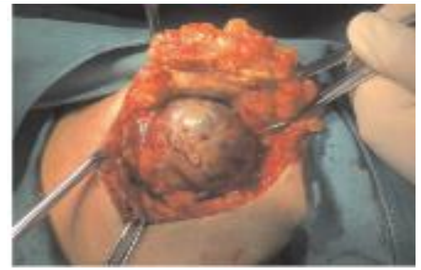

Figure 3 : Intraoperative view showing a firm encapsulated mass.

Aim

We aim to describe the clinical and histological features of the extra-axial chordomas and its management.

Conclusion

Nasal chordoma is an extremely rare malignant tumor wtih no specific clinical and radiological profile. Diagnosis requires histopathological confirmation with immunohistochemical staining.
Surgical resection is the gold standard in the management of chordomas.

Méthodes et Matériels

- We report a case of a patient referred to our ENT department for a nasal tumour.

Références

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2.Ulici V, Hart J. Chordoma. Arch Pathol Lab Med. 2022 Mar 1;146(3):386-395. doi: 10.5858/arpa.2020-0258-RA. PMID: 34319396..