Young man with nasal obstruction and anosmia

An 18-year-old man was admitted to the otolaryngology service with a 6-month history of headaches and 3-month history of nasal obstruction and anosmia. The patient subsequently underwent MRI of the head.

Imaging Findings:

MRI of the paranasal sinuses demonstrates a large mass, of which the epicentre appears to be the left aspect of the nasal cavity (Fig. 1, 2, 3, 4). The lesion contacts the cribriform plate (Fig. 2a), extends superiorly into the left frontal sinus (Fig. 2b) and obliterates the left choana, growing into the nasopharynx postero-inferiorly (Fig. 1b). There is no evidence of destruction of the cribriform plate and no extension into the cranium or orbital involvement. The outflow of the ethmoid and ipsilateral maxillary and frontal sinuses are apparently obstructed as these sinuses are filled with secretions. The mass is of soft tissue intensity, hypointense on T1 WI (Fig. 3), hyperintense on T2 WI (Fig. 1, 2) and demonstrates extensive and relatively homogeneous enhancement following contrast administration (Fig. 4).

Location and appearances are typical for an olfactory neuroblastoma which was subsequently proven histologically.

Discussion:

Esthesioneuroblastoma (ENB) is an uncommon malignant tumour that arises from bipolar sensory receptor cells in the olfactory mucosa. The incidence curve has a bimodal shape with the first peak in the 2nd decade and the second peak in the 6th decade. The main symptoms are nasal occlusion, proptosis, epistaxis, headache, excessive lacrimation, anosmia and visual disturbance [1, 2].

This tumour is almost always unilateral and may cause metastasis by lymphatic and haematogenous routes. Local recurrence has been reported in up to 57% of patients. A metastatic rate of 20% to 60% is reported with the most common site being the cervical lymph node [2].

On CT the tumour appears as a homogenous density mass, which usually enhances in a moderate and homogenous fashion. Intraleisional calcification and presence of cysts along the intracranial margins yield a definite diagnosis. Bone erosion is frequent and usually accompanied by modelling of bone [2].

On T1W MR Images after contrast administration, esthesioneuroblastoma presents as homogenously enhancing
tumour with intermediate signal intensity, whereas T2W images, the original intensity is increased [2]. MRI can delineate intraorbital and intracerebral extension. Gadolinium enhanced MR images help to differentiate tumour from obstructed secretions in paranasal sinuses, to determine meningeal and extradural spread and to detect perineural spread.

The use of scintigraphy with a radiolabelled somatostatin analog (Octreoscan) can provide information regarding the extent of tumour involvement and help guide clinical decision. The ability to detect cervical metastases and widespread systemic disease provides the rationale for use of Octreoscan for staging and surveillance imaging of patients with ENB [4].

The optimal treatment continues to be controversial. Endoscopic endonasal surgery can be applied with good results. Advantages of endoscopic endonasal surgery include improved visualization and decreased morbidity compared to traditional approaches. However, there are contraindications for doing an endonasal approach, particularly depending on the relationship to critical neurovascular structures. Cerebrospinal fluid leaks are the major complication in early endonasal procedures [5] and should be taken in consideration on imaging evaluation.

The benefit of adjuvant therapy, particularly radiotherapy, has been well described in the literature. The largest reported series evaluated neoadjuvant radiotherapy, with or without neoadjuvant chemotherapy, and found improved resectability with improved patient survival. Although there is no standard chemotherapy regimen used for ENB, the agents used are chiefly cisplatin and etoposide, and doxorubicin and vincristine with an alkylating agent [6].

The prognosis is doubtful, as local recurrence and distant metastases often occur [1, 3].

**Differential Diagnosis List:** Esthesioneuroblastoma, Non-Hodgkin Lymphoma, Nasal and paranasal squamous cell carcinoma, Sinonasal polyposis, Juvenile angiofibroma, Embryonal rhabdomyosarcoma, Neuroendocrine carcinoma, Ewing sarcoma

**Final Diagnosis:** Esthesioneuroblastoma

**References:**


Robert C. Rostomily, Maria Elias et al. (2006) Clinical utility of somatostatine receptor scintigraphic imaging (octreoscan) in esthesioneuroblastoma: a case study and survey of somatostatine receptor subtype expression. Wiley InterScience 28: 305–312 (PMID: 16470879)


Description: Axial T2 WI demonstrating a large hyperintense mass with its epicentre in the left nasal cavity. Origin: IPOFG, Department of Radiology
**Description**: Axial T2 WI showing that the mass obliterates the left choana and contacts the nasopharynx postero-inferiorly. **Origin**: IPOFG, Department of Radiology
Description: Coronal T2 WI revealing the contact of the lesion with the cribriform plate without clear invasion of it. Note the bright intensity of the ethmoid cells and the left maxilar sinus related to its obstruction. Origin: IPOFG, Radiology Department
Description: Coronal T2 WI demonstrating the extension of the mass into the left frontal sinus. Note the bright intensity of the left frontal sinus related to its obstruction as well. Origin: IPOFG, Radiology Department
**Description:** Coronal T2 WI showing the posterior extension of the lesion without evidence of destruction of the surrounding structures, namely the sphenoid sinus. **Origin:** IPOFG, Radiology Department
Description: Axial T1 WI demonstrating the hypointense soft tissue lesion extending into the nasopharynx. Origin: IPOFG, Department of Radiology
Description: Axial T1 WI after contrast administration showing the extensive enhancement of the lesion. Origin: IPOFG, Department of Radiology
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