MRI features of esthesioneuroblastoma. A descriptive analysis of 7 cases

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Learning objectives

The goals of this paper were to analyse the MRI characteristics of esthesioneuroblastomas including extent of tumor, differentiation from obstructive sinus disease, MR signal and pattern of contrast enhancement.

Background

Estheioneuroblastomas are uncommon tumors originating in the olfactory epithelium of the superior nasal cavity. Accurate staging appropriately guides therapy and predicts survival. The MR appearance and pattern of contrast enhancement in these tumors have not been well described in the literature. The goals of this paper were to analyse the MRI characteristics of esthesioneuroblastomas including extent of tumor, differentiation from obstructive sinus disease, MR signal and pattern of contrast enhancement.

Findings and procedure details

Methods and materials:

The MR examinations of seven patients with proven advanced esthesioneuroblastoma were reviewed. Exams were performed using a 1.5 Tesla GE (General electric) MR machine for 4 patients and using Siemens 1.5 Tesla MR machine for 3 patients. Standard SE T1- and T2-weighted axial images were obtained [550-600/15-25 and 2,000-2,800/80-90 (TR/TE), respectively], followed by postcontrast axial, sagittal and coronal T1-weighted sequences in all patients.

Results:

Seven patients (3 females and 4 males) with olfactory neuroblastoma were treated at our institution (Monastir and Sousse medical university hospitals during the past 12 years. The age was ranging from 18 years to 68 years with a mean of 56 years. Unilateral nasal obstruction was noted in 5 cases, recurrent epistaxis in 3 patients, anosmia in 6 cases, proptosis in 2, and frontal headache in 2 cases.
The MR signal, pattern of contrast enhancement and extent of tumor were examined. MR images taken in 7 patients with histologically confirmed olfactory neuroblastoma were retrospectively reviewed.

Compared with brain gray matter, tumors were hypointense (4 cases) and isointense (3 cases) on T1-weighted images, 3 homogeneously and 4 heterogenously (Fig 1, 2).

4 tumors were isointense on T2-weighted images, 2 heterogenously hyperintense although their appearance was less intense than that of sinusitis, and in one case the signal was iso and hypointense (Fig 3, 4).

Gadolinium enhancement was moderate in one case and marked in 6 of the 7 cases, 4 homogeneously and 2 heterogeneously. 5 of the 7 tumors showed smooth regular shaped margins; 2 of these tumors exhibited irregular infiltrating margins with dural enhancement on gadolinium-enhanced images, compared to the pre-contrast T1-weighted images (Fig 5, 6). The best MRI sequence for detection of intracranial extension was the Gadolinium enhanced t1 weighted image in the coronal plane.

2 of the 7 cases exhibited intracranial cysts on T2 and T1 gadolinium-enhanced images(Fig 7). In 5 cases the intracranial extension with dumbbell shape was clearly identified. Orbital extension was noted in 4 cases.

Postobstructive sinus disease was encountered in 6 cases. T2-weighted or gadolinium-enhanced images successfully distinguished sinusitis from tumors in 4 cases (Fig 8).

In most cases, olfactory neuroblastomas were hypo to isointense on T1-weighted images, isointense on T2-weighted images, and show marked homogeneous enhancement with well-demarcated regular margins upon gadolinium enhancement.

**Discussion:**

Olfactory neuroblastoma (also known as esthesioneuroblastoma (ENB)), first described by Berger and Luc in 1924 (1, 2, 3, 4), is rare, accounting for 3% of all intranasal tumors1. It is usually found in young men, with a secondary peak at the age of 50-60 years (4, 5, 6). Only one patient was 18 years old in our series whereas all the other 6 patients were more than 50 years older.

Esthesioneuroblastoma is an uncommon malignant tumour that arises from bipolar sensory receptor cells in the olfactory mucosa. These cells originate in the neural crest and differentiate into the olfactory sensory elements.

The main symptoms are nasal occlusion, proptosis, epistaxis, headache, excessive lacrimation, anosmia and visual disturbances. Because sensory nerves of smell
originates in olfactory bulb and pass through cribriform plate to olfactory area of nasal mucosa which is located in the most superior part of both nasal fosse, usual primary sites of occurrence include superior nasal cavity or nasal septum, the turbinates, the ethmoid, or the cribriform plate, although an extra nasal site of origin has been suggested (1, 2).

As they grow, they tend to destroy surrounding bone, and can extend in any direction. This invasion may be superiorly into the anterior cranial fossa, laterally into the orbits and across the midline into the contralateral nasal cavity. They can also obstruct the ostia of paranasal sinuses resulting in opacification of the sinus with secretions (2, 5, 7, 8). Hyperostosis may be an inflammatory reaction related to obstructed neighboring air cells. This neoplasm is locally aggressive and cause metastasis by lymphatic and hematogenous routes.

The first clinical staging system was proposed by Kadish et al. in 1976 (7, 9, 10), and still is an important prognosis predictor (11). Survival for stage A and B tumors is usually excellent. Tumor extension to orbits or through the cribriform plate has significant prognostic implications (5). Morita (12) proposed a revised Kadish staging, with stage C consisting of local disease extending beyond the paranasal sinuses and stage D consisting of cervical or distant metastases. Both Biller (13) and Dulguerov (14) have proposed systems based on TNM staging, but neither has been widely accepted, as there is no evidence that they provide better prognostic differentiation than the Kadish system (9). Our cases fit into the Kadish A stage (2 cases) B stage (5 cases), with extension to ethmoidal air cells, therefore carrying a good prognosis.

CT and MRI are complementary examinations used for initial diagnosis, staging and follow-up.

The MRI appearance is often that of a large, soft tissue, dumbbell-shaped mass centered within the superior nasal cavity and extending intracranially. The tumor is generally hypointense on T1-weighted images and iso- to hyperintense on proton density and T2-weighted images. Contrast enhancement is usually mild to marked and can be uniform or mildly heterogenous. This appearance was noted in the majority of our cases. These MR findings are however relatively non-specific (1, 2, 3). Som et al. showed that cystic components at the intracranial margins of the tumor may be highly suggestive of ENB (3, 6) which was noted in 2 of our cases. One case of our series was atypical, and presented iso and hypointense signal in T2 weighted images.

MRI is also important for staging, as it exhibits the extent of soft tissue invasion with involvement of local structures, such as orbits and sinuses.

The use of MRI is crucial for discriminating between postobstructive secretion and tumor tissue (8). Postobstructive secretions showed a markedly hyperintense signal on T2 weighted images, thus differentiating them from the isointense tumor in the same images. In addition lack of contrast uptake of sinusal retention cysts on Gadolinium enhanced MR images can help to differentiate tumour from obstructed secretions in paranasal
sinuses, and is useful in determining meningeal and extradural spread and to detect perineural spread. Enhanced images, particularly in the coronal plane, were very helpful in identifying intracranial extension in our series.

Thus, olfactory neuroblastomas can be detected, delineated and its characteristics suspected by MRI, but definite diagnosis however is still based on histopathology (2).

Images for this section:

![Axial T1 weighted image: Left ethmoido orbital mass displaying isointense signal compared to grey matter.](image)

**Fig. 1:** Axial T1 weighted image: Left ethmoido orbital mass displaying isointense signal compared to grey matter.
**Fig. 2:** Coronal T1 weighted image showing large isointense mass involving the left nasal fossa and extending superiorly to ethmoidal cells as well as left orbital cavity. Intra cranial extension noted through the cribriform plate.
**Fig. 3:** Coronal T2 weighted image: Large ethmoido nasal mass with intra cranial extension through anterior cranial base displaying heterogeneous hyper intense signal. Note the hyper intense cysts at the superior margins of the tumour.
Fig. 4: Coronal T2 weighted image through orbits showing isointense intra cranial iso and hypointense lesion based on the olfactory grooves associated with extensive bilateral frontal white matter oedema.
**Fig. 5:** Sagittal T1 weighted image after IV contrast injection: Extra axial anterior cranial fossa mass with extension into ethmoids, displaying moderate heterogeneous contrast uptake.
**Fig. 6:** Coronal T1 with contrast injection: The large ethmoido nasal and frontal mass exhibits intense heterogeneous contrast enhancement.
Fig. 7: Axial T2 weighted image showing the hyper intense cysts at the periphery of the mass.
**Fig. 8:** Axial T2 weighted image: Left ethmoido nasal mass obstructing the left ostio meatal unit with cyst retension within left maxillary sinus.
Conclusion

Although MR features are nonspecific, they may suggest an imaging diagnosis of olfactory neuroblastoma when seen in the superior nasal cavity. Magnetic resonance signal characteristics helped to distinguish obstructive sinus disease from tumor. Enhanced images, particularly in the coronal plane, were very helpful in identifying intracranial extension.

Personal information

References