Olfactory Neuroblastoma: MR Evaluation

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PURPOSE: To evaluate MR in the diagnosis and staging of olfactory neuroblastoma. METHODS: Five patients with histologically proven olfactory neuroblastomas were studied by MR. Four also had CT scans. RESULTS: The extent of disease delineated by MR agreed with surgical findings and surgical staging of the tumor in all five patients. All cases of olfactory neuroblastomas originated in the nasal cavity and involved the ethmoid sinuses. In three of the five cases, the tumors extended to the anterior skull base. In one case MR correctly suggested skull base involvement missed on CT. Lesions were isointense to hyperintense with muscle on T1-weighted scans. Compared with fat on T2-weighted scans, olfactory neuroblastomas were variable in signal intensity. Enhancement was minimal to moderate in all cases. CONCLUSION: In the evaluation of olfactory neuroblastoma, MR is most useful in delineating the extent of the tumor and may be more accurate than CT. However, the signal intensity characteristics of olfactory neuroblastomas may overlap other tumors.

Index terms: Neuroblastoma; Esthesioneuroblastoma; Nose, neoplasms; Nose, magnetic resonance

Olfactory neuroblastoma (ON), also called esthesioneuroblastoma, is an uncommon intranasal tumor originating from the olfactory neuroepithelium lining the roof of the nasal vault and in close proximity to the cribriform plate. ONs occur in all age groups with a peak incidence in the age groups of 11 to 20 years and 51 to 60 years (1). There is a slightly greater incidence of the tumor in women. The common clinical symptoms are nasal obstruction, recurrent epistaxis, hyposmia, rhinorrhea, headaches, and visual disturbances (1-4). Computed tomography (CT) has been shown to be extremely useful in staging and treatment planning of this tumor (2, 5). However, magnetic resonance (MR) imaging has shown tremendous promise in surpassing CT for the mapping of neoplasms of the nasal cavity and paranasal sinuses (6-9). The authors have encountered five patients with histologically proved ON during the past 5 years who had MR studies. This entity has not been well described in the MR literature, and it remains to be seen whether ONs have stereotypical MR imaging features that can distinguish ON from other lesions.

Materials and Methods

Five patients with histologically verified ONs were examined by MR (n = 5) and CT (n = 4). There were three women and two men with ages ranging from 17 to 76 years (mean 49.4 years) (see Table 1). MR imaging was performed on a General Electric (Milwaukee, WI) 1.5-T Signa scanner using the quadrature head coil and a Medical Advances (Milwaukee, WI) anterior-posterior volume neck coil. T1-Weighted images were performed in the axial, coronal, and/or sagittal plane, 600-800/11-30/1-2 (repetition time/echo time/excitations). Double-echo long-TR scans (2500-3500/18-25) were performed in the axial plane. In the three of the five cases, gadopentetate dimeglumine (Berlex Industries, Wayne, NJ) was administered at a dose of 0.1 mmol/kg. T1-weighted scans (with the identical parameters above) were performed immediately after administration of the gadolinium in the axial and coronal planes. Fat-suppression techniques for the postcontrast scans were variably employed. All MR scans used a 256 X 192 matrix, 5-mm-thick sections, and inferior saturation pulses. Intersection gaps of 2 to 2.5 mm were standard for long-TR scans; the short-TR scans had no intersection gaps.
TABLE 1: Clinical information in five patients with olfactory neuroblastoma

<table>
<thead>
<tr>
<th>Case</th>
<th>Age, Sex</th>
<th>Presenting Symptoms</th>
<th>Primary Tumor Location</th>
<th>Tumor Staging</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>76, F</td>
<td>Nasal obstruction, decreased smell, headache</td>
<td>Right upper nasal cavity</td>
<td>C</td>
</tr>
<tr>
<td>2</td>
<td>65, F</td>
<td>Epistaxis, nasal obstruction</td>
<td>Left upper nasal cavity</td>
<td>B</td>
</tr>
<tr>
<td>3</td>
<td>44, M</td>
<td>Decreased smell and vision on left</td>
<td>Left upper nasopharynx</td>
<td>C</td>
</tr>
<tr>
<td>4</td>
<td>17, F</td>
<td>Nasal congestion, right &gt; left</td>
<td>Right sinonasal cavity</td>
<td>B</td>
</tr>
<tr>
<td>5</td>
<td>45, M</td>
<td>Epistaxis</td>
<td>Left upper nasal cavity</td>
<td>C</td>
</tr>
</tbody>
</table>

CT was performed on a GE 9800 scanner. Three- to 10-mm-thick sections were obtained in both the axial and coronal planes, after administration of intravenous contrast material. Supplementary 5-mm scans were made through regions of special interest.

Both the MR and CT scans were carefully evaluated for the tumor's original location, extension, bony structure changes, anterior skull base invasion, and some associated changes, including postobstructed secretions, hemorrhage, and edema. ONs were staged from A to C according to the classification system developed by Kadish et al (2). In stage A, the tumor is limited to the nasal cavity. In stage B, the tumor is confined to the nasal cavity and paranasal sinuses, and in stage C, the tumor extends beyond the nasal cavity and paranasal sinuses.

The scans were retrospectively reviewed by two radiologists in concert without knowledge of surgical findings. The CT and MR scans were viewed separately but one after the other in a random order. The CT and MR scans were performed within 1 to 13 days of each other with no intervening therapy. The imaging findings were compared with surgical findings.

Fig. 1. Case 1. Seventy-six-year-old woman with reported hyposmia and pathologically confirmed olfactory neuroblastoma.

A, Sagittal (600/20) MR scan through the midline demonstrates a soft-tissue mass (arrow) which breaches the cribriform plate and demonstrates extraaxial intracranial tumor (arrowheads).

B, Axial (3000/90) MR scan through the same level demonstrates persistent low signal intensity of the mass. Note the high signal intensity (arrowhead) of obstructed secretions in the posterior ethmoid air cell behind the lesion.

C, Axial (3000/35) MR scan through the level of the superior orbits demonstrates a low-intensity mass (arrow) within the right nasal cavity and ethmoid air cells with extension across midline to the left ethmoid air cells.

D, Coronal (600/20) MR scan through the anterior nasal cavity demonstrates the intracranial extension of the mass (arrowhead) through the cribriform plate invading the region of the olfactory lobe on the right side. Postobstructive secretions are present within the maxillary antrum (A).
Results

Clinical data on the patients with ON are summarized in Table 1.

On T1-weighted (short-TR, short-TE) MR examinations, all ONs were hypointense to fat, but the intensity compared with muscle and inflammatory mucosal disease was variable. On T2-weighted (long-TR, long-TE) sequences, the tumors were always hyperintense to muscle and hypointense to inflamed mucosa, but had variable intensity compared with fat. On proton density-weighted (long-TR, short-TE) scans the five ONs were hypointense to fat, hyperintense to muscle, and hypointense to mucosal disease.

Gadopentetate dimeglumine was administered to three patients. Tumor enhancement was heterogeneous in the three cases. In these three cases, the solid tumor enhancement was readily discernible from the peripheral (rim) enhancement of obstructed sinonasal secretions. Meningeal invasion, or the lack thereof, was well demonstrated after gadolinium.

Two tumors demonstrated speckled calcifications on CT scans; the calcified portion of the mass was not detected, even in retrospect, on MR scans.

The extent of disease delineated by MR agreed with surgical findings in all five patients. Four cases of ON originated in the high nasal cavity and involved the ethmoid sinus (Figs 1 and 2). In three of the five cases, tumors extended to the anterior skull base. One tumor invaded the brain and produced white matter edema (Fig 1). Another case was unusual in that it involved the maxillary antrum more than the nasal cavity (Fig 3).

Although the staging by CT (in four cases) was equivalent to MR, MR demonstrated intracranial extension not apparent on CT in one case. Why the intracranial extension by this ON was not
Fig. 3. Case 4. Seventeen-year-old girl with nasal stuffiness and pathologically proved olfactory neuroblastoma.

A, Sagittal (600/20') MR scan to the right of midline demonstrates a soft-tissue mass that completely opacifies the right nasal cavity. Note the heterogeneity of the signal intensity within the mass with the anterior portion more hyperintense than the intermediate posterior portion. The bony margin of the cribriform plate (arrowheads) is intact, as surgically confirmed.

B, Axial (600/20') MR scan through the maxillary antrum demonstrates complete opacification of the maxillary antrum by the neoplastic process (T). Whenever one confronts an isolated maxillary sinus opacification, one should consider neoplasm rather than an inflammatory condition. The bony margin of the maxillary antrum is breached medially, as invasion into the nasal cavity is evident.

C, Coronal (2500/80) scan further anterior to C demonstrates the low intensity of the mass, again isointense with gray matter. Although there is high signal intensity caused by postobstructive secretions in the frontal-ethmoidal recess on the right side, no tumor is seen to invade the cribriform plate or extend intracranially. MR is exquisitely useful for differentiating neoplasm from postobstructed secretions because of the difference in the signal intensity, as noted in this example.

seen on CT was unclear but probably related to the very minimal destruction of the cribriform plate. The tumor may have traveled through the sieve-like openings through the cribriform plate rather than aggressively destroying bone. Nonetheless, because of concomitant orbital involvement, this ON was preoperatively labeled as stage C by both CT and MR. Therefore, although the staging was identical, the MR more accurately mapped the tumor, as was confirmed at surgery.

Discussion

ONs are rare, slow-growing malignant tumors of the sinonasal cavity. The incidence of ON has been estimated to range between 2% to 3% of all malignant intranasal neoplasms (4). Most of the cases of ON arise from the olfactory neuro-epithelium lining the cribriform plate, upper one-third of the nasal septum, superior turbinate, and anterior ethmoid air cells. Symptoms associated with early ON include: 1) gradually decreasing sense of smell; 2) some degree of unilateral nasal obstruction; and 3) episodic epistaxis. These early clinical symptoms are nonspecific but, in combination, should lead one to consider the diagnosis of ON.

The tumors tend to grow slowly, spread submucosally, and eventually extend into adjacent structures, especially the sinuses, palate, orbit, and brain. Remote metastases may occur in advanced cases.

The preoperative assessment of a patient with ON requires medical imaging, and herein lies some controversy. Although CT is superior to MR in the evaluation of bony structures, soft-tissue boundaries between tumor, retained secretions, and muscle are more accurately delineated with MR (9). Which study should be ordered?

In this short report, we found that MR was more accurate than CT in delineating ON extent. Because the staging system is so general, the improved accuracy of MR in delineating the tumor is not demonstrated just by stage classification. Extension to the orbit on one CT study led to a stage C classification. The concomitant intracranial extension noted by MR was not seen on CT. Although this is not reflected by differences in the staging classification (stage C includes both orbital and intracranial extension), the finding on MR made a difference in the surgical approach. A craniofacial (neurosurgical-otorhinolaryngologic) combined-team approach was necessary. We have noted this improved
ability to detect intracranial spread by MR in a study of inverted papillomas (10). It would seem that intracranial spread need not require cribiform plate destruction detectable by CT. Perhaps a tumor can cross the fenestrations of the plate without destroying it to a degree detectable by CT.

The ability of MR to differentiate tumors from obstructed secretions in the paranasal sinuses was evident in this study also and has been previously described with other sinonasal tumors (7). Obstructed secretions are generally (but not exclusively) high intensity on T2-weighted scans and show peripheral enhancement; tumors are usually (but not exclusively) intermediate in intensity on T2-weighted scans and enhance in a solid fashion. Gadolinium enhancement is therefore very helpful in the mapping of sinonasal neoplasms. Its utility in determining meningeal and extradural extrameningeal spread is widely known; it also can be helpful, in other settings, to detect perineural spread of sinonasal neoplasms.

It should be noted that the signal-intensity characteristics of ONs in this limited study population overlap those of other neoplasms in the sinonasal cavity (11). One of the distinguishing features of this tumor is its propensity for calcification, although inverted papillomas and chondroid tumors can calcify as well. Spin-echo MR is less sensitive for the detection of calcification than CT is. In this regard, CT findings may suggest the specific histology of ON when there is a calcified superior nasal mass. However, by virtue of the lesion's location high in the nasal vault, the diagnosis of ON usually will be also suggested by MR. In any event, a biopsy is required for histopathologic confirmation. At that point, the superior mapping ability of MR is of paramount importance.

References
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