Noncontiguous Bilateral Esthesioneuroblastoma: A Case Report

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ABSTRACT

Esthesioneuroblastoma is a rare malignant neoplasm arising from the olfactory epithelium. This tumor has a tendency to originate from one side of the nasal cavity or paranasal sinuses with frequent extension into the contralateral nasal cavity through direct invasion. A review of the literature reveals numerous case reports describing esthesioneuroblastoma with unilateral or bilateral involvement; however, there have been no known reports of bilateral noncontiguous (multifocal) esthesioneuroblastoma. We present a unique case of a patient diagnosed with two separate primary esthesioneuroblastomas who was managed with preoperative radiation followed by surgical resection of the tumor. This case illustrates that esthesioneuroblastoma must remain as part of the differential diagnosis for a multifocal, noncontiguous intranasal and paranasal sinus mass.

KEYWORDS: Esthesioneuroblastoma, noncontiguous sinus mass, olfactory neuroblastoma

CASE REPORT

A 49-year-old male presented with a 6-month history of left nasal obstruction and anosmia. He had no history of associated epistaxis, headaches, diplopia, or sensory changes. Endoscopic examination revealed a polypoid mass in the left nasal cavity

arising from the posterior ethmoid sinuses. Examination of the right nasal cavity was normal, as was the remainder of the head and neck examination.

Computed tomography (CT) with contrast (Fig. 1) showed a heterogeneously enhancing mass within the left nasal cavity and maxillary sinus with opacification of the sphenoid and ethmoid sinuses.

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Received: April 7, 2007. Accepted: July 10, 2007. Published online: September 4, 2007.

DOI 10.1055/s-2007-986459. ISSN 1531-5010.

Figure 1 Computed tomography (CT) coronal image demonstrating bilateral maxillary sinus involvement of soft tissue mass as well as left ethmoid mass abutting the orbit and skull base/cribriform plate.

Figure 2 Coronal magnetic resonance imaging (MRI) shows findings similar to the computed tomography (CT) image. Some of the opacification seen on CT in the maxillary sinuses, which appear to be more prominent on the left, appear to be inspissated mucus.

The cribriform plate, lamina papyracea, infraorbital floor, and nasal septum were all intact with no evidence of disease extension across the midline. Partial opacification of the right maxillary sinus was noted. A diagnosis of a grade II esthesioneuroblastoma was made on biopsy of the nasal mass. Immunohistochemistry was positive for neuron-specific enolase, synaptophysin, and chromogranin; however, it was negative for keratin (AE1:AE3) and S-100.

Magnetic resonance imaging (MRI) (Fig. 2) revealed a soft tissue mass measuring 2.2×1.6 (anteroposterior $\times$ transverse) cm in the left posterior nasal cavity, which did not extend to the cribriform plate. An enhancing soft tissue nodule was seen within the inferior aspect of the left maxillary sinus. There was no evidence of direct extension of the tumor from the left nasal cavity. In addition, a heterogeneous focus of tissue, found to be predominantly of low signal on T2 sequence, was located within the right maxillary sinus antrum. The signal characteristics were reported to be more in keeping with neoplastic tissue.

A polypoid lesion completely confined to the right maxillary sinus was found on transnasal endoscopic examination. Biopsy of this lesion was reported as an esthesioneuroblastoma. Based on endoscopic examination, imaging, and pathologic results, this patient was diagnosed with two separate primary esthesioneuroblastomas. The patient was managed with preoperative radiation followed by a craniofacial resection including a right and left partial maxillectomy, ethmoidectomy, and sphenoidectomy. Intraoperatively, there was no evidence of direct extension of tumor between the left and right nasal cavities. Furthermore, the septum was completely intact. Final pathology confirmed bilateral esthesioneuroblastomas.

DISCUSSION

Esthesioneuroblastoma is a rare tumor that is believed to arise from the olfactory epithelium

and has a predilection for invading surrounding regions, such as the paranasal sinuses, orbits, and skull base.¹ It is a rare disease that was first described in 1924 in the French medical literature² and since that time, this tumor continues to be uncommon. The true incidence is difficult to ascertain; however, it may account for ~3% of all intranasal tumors.³

The most common symptoms of patients with this tumor may include nasal obstruction, epistaxis, anosmia, and headache.^{1,3,4} Most of these symptoms are nonspecific, and patients often present with locally advanced disease. A unilateral mass within the nasal cavity is frequently noted at presentation.⁵ This was the main clinical finding on examination of our patient. It was only after further investigation that it was discovered our patient had bilateral disease. Initially, due to the rarity of a noncontiguous bilateral esthesioneuroblastoma, we theorized the patient had a left-sided tumor with inflammatory changes on the right maxillary sinus, as indicated by the opacification on CT imaging. Previously, there have been no known reports of a patient with two separate primary sites of an esthesioneuroblastoma.

Diagnosis of this tumor may initially be difficult, and it has been referred to as "the great imposter."⁶ Many tumors may mimic an esthesioneuroblastoma, and one must be cognizant of the differential diagnosis, particularly in a situation such as ours where the specific tumor is not only rare, but the noncontiguous bilateral focus of the disease is highly unlikely. Specific tumors to be aware of include inverting papilloma, squamous cell carcinoma, adenocarcinoma, sinonasal undifferentiated carcinoma, hemangioma, and metastatic carcinoma.⁷⁻⁹ Certainly, in addition to imaging, the diagnosis must be made definitively through histopathology. Homogeneous small cells in a rosette or pseudorosette formation characterize a well-differentiated esthesioneuroblastoma,

whereas an undifferentiated esthesioneuroblastoma consists of anaplastic hyperchromatic small cells with little cytoplasm. The latter may be difficult to diagnose; therefore, further studies in the form of electron microscopy and immunohistochemical staining should be performed.^{5,10}

ACKNOWLEDGMENTS

The authors would like to acknowledge the support of the Rallying Against Cancer Hardcore (RACH) Fund administered through the Princess Margaret Hospital Foundation in support of research in esthesioneuroblastoma and skull base malignancy. We would also like to thank Dr. Doron Sommer for his assistance with the figures.

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