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## Esthesioneuroblastoma: What Family Practitioners Should Know

***Esthesioneuroblastoma is an uncommon neoplasm of the nasal cavity which is curable if diagnosed early but fatal if neglected. We describe a case of Stage B esthesioneuroblastoma excised by using the lateral radical rhinotomy surgical technique followed by radiation therapy. The patient was free of the tumor and asymptomatic 13 years after treatment. Our diagnosis and treatment of this uncommon nasal tumor serves as an example of how family physicians and nurse practitioners must be alert to the possibility of such serious tumors and when to assertively seek consultation for their management.***

Esthesioneuroblastoma (EN) is an uncommon neoplasm which arises from the olfactory epithelium in the cribriform region of the nasal septum. Of neuroectodermal origin, EN accounts for 3% of all nasal neoplasms<sup>1,2</sup> and mainly affects white persons. Mean age at diagnosis is 48 years (range, 9 to 83 years).<sup>3</sup> EN occurs 1.6 times more often in women than in men.<sup>4</sup>

Differentiation of EN from other tumors is difficult because it resembles other neuroendocrine tumors of the head and neck. The cause of EN is unknown. Smokers are at increased risk for EN.<sup>4</sup> EN also is not as aggressive as undifferentiated carcinoma; however, if not treated early in its course, EN spreads rapidly and is fatal.

We describe our treatment of a case of EN and review its diagnosis and treatment.

### Case report

A 50-year-old white man was seen by an otolaryngologist for nasal obstruction, chronic sinusitis, a feeling of fullness in the head, and numbness of the left cheek and left maxillary teeth. He was given antihistamines but returned the next month with continued nasal ob-

struction as well as rhinorrhea and dizziness. Because he was noncompliant, the patient was not seen until three months later, when he was seen for epistaxis. Nasal biopsy was done. Computed tomography (CT) scans and x-ray films of the sinuses showed a soft-tissue mass in the nasopharynx which crossed the midline, left maxillary sinus, ethmoid, inferior orbit, and cribriform plate. The left paranasal sinus was eroded up to the orbit. The patient was sent to an urban medical center for surgical and adjunctive treatment. Initial biopsy results suggested small cell neoplasm or a metastatic lesion from occult lung cancer. Bronchoscopy, sputum cytology, and chest x-ray results were negative. Chemistry panel and liver function test results were normal. The patient had mild anemia. Expert review of pathology slides showed EN.

The patient, raised on a farm, had smoked for 35 years but had no history of alcohol abuse. One of the patient's two brothers had died of pancreatic cancer at age 65 years; and the other, a heavy smoker, died of a nasopharyngeal neoplasm.

Because the tumor was located in the postnasal cavity and extended

superiorly and posteriorly to the ethmoid labyrinth and into the maxillary sinus, the patient had subtotal maxillectomy and complete ethmoid and sphenoid exenteration through the lateral rhinotomy approach on the left side. The sphenoid bone was partly eroded anteriorly. The left nasal bone was removed with the maxillary mucosa. The orbital floor was eroded. The tumor was peeled off the petriorbital fascia, which was intact. The eroded sphenoid sinus was excised with the sphenoid mucosa. Three days after having maxillectomy, radiation treatment was begun.

The tentative diagnosis of undifferentiated, small cell, undifferentiated cancer was changed to EN at further review. The tumor weighed 25 g; consisted of soft, tan-to-yellow, friable tissue; and measured 2 × 2 × 3.2 cm. As is consistent with EN, no true rosette was seen.

Careful evaluation by his physician and oncologist every six months showed that the patient remained disease-free 13 years after surgery.

### Discussion

#### *Clinical Diagnosing EN*

Knowing the clinical manifestations of EN enhances the role of the primary care physician, the nurse practitioner, the ophthalmologist, the surgeon, and the oncologist in the vital early detection and prompt treatment of the disease. The most common initial signs and symptoms of EN are nasal obstruction, epistaxis, exophthalmos with visual field defects, blurred vision, eye pain, conjunc-

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tival erythema, eyelid edema, headache, blindness, excessive tearing, anosmia, diplopia, facial numbness and sweating, a polypoid intranasal mass with a granular red appearance, shortness of breath, fatigue, weight loss, and chest pain.

Our patient's smoking history and family history of neoplasms made us suspect that the etiology of EN has a hereditary component.

#### **Treating EN**

Timely surgery with radiation therapy yields a good prognosis in Stage A disease, which is defined<sup>5</sup> as a tumor confined to the nasal cavity. Stage B disease is defined<sup>5</sup> as a tumor confined to the nasal cavity and one or more paranasal sinuses. A craniofacial approach followed by radiation therapy is required. In Stage C disease (defined as a tumor extended beyond the nasal cavity or paranasal sinuses into the orbit, base of skull or intracranial cavity, cervical lymph nodes, or distant sites<sup>5</sup>), chemotherapy with such agents as cyclophosphamide is required in addition to surgery and radiation therapy.

The importance of salvage therapy to prolonging survival in patients with advanced EN has been recognized: a retrospective review of EN treatment<sup>4</sup> reported survival rate of 88% in patients with Stage C disease after treatment with surgery, radiation therapy, and chemotherapy, in contrast to a survival rate of 50% after treatment with only combined craniofacial resection and radiation.

#### **Conclusion**

EN is an uncommon, complex olfactory tumor which is curable if diagnosed early but which may be fatal if misdiagnosed and neglected. To identify

possible EN, the treating family physician, oncologist, surgeon, nurse practitioner, and pathologist should be alert for any initial complaints of nasal obstruction lasting longer than one month or of persistent rhinorrhea or epistaxis with neurologic deficits (eg, anosmia, focal numbness of the face or cheek, or visual defects) especially in patients who have a history of smoking and a family history of neoplasm. X-ray films of the sinuses and CT scans should immediately be obtained when the family physician or nurse practitioner is in doubt about the differential diagnosis, and the patient must be promptly referred for head and neck or neurosurgery consultation for possible tumor resection. As in other conditions, patients should be instructed that noncompliance could affect prognosis.

Although esthesioneuroblastoma is still seen uncommonly, it is being observed with increasing frequency. Therefore, as for other such increasingly observed conditions, family practitioners must increase their awareness of this tumor. ♦

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## **Inside Out**

The greatest revolution of our generation is the discovery that human beings, by changing the inner attitudes of their minds, can change the outer aspects of their lives.

*William James, Institute of Noetic Sciences*