

## Olfactory Neuroblastoma—An Unusual Presentation

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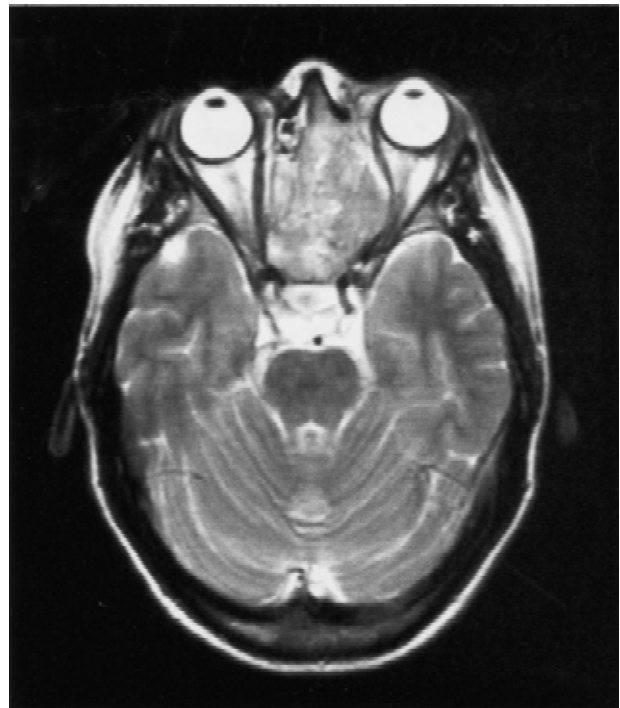
Olfactory neuroblastoma, also referred to as esthesioneuroblastoma, is a rare neuroectodermal tumor that originates from the olfactory sensory epithelium in the roof of the nasal fossa (1). The majority of patients present with chronic and progressive symptoms related to the nose or sinuses, such as nasal bleeding and persistent nasal discharge, which may have been present for years before seeking medical advice (2). The most common presenting ocular complaints include chronic peri-orbital pain, increased lacrimation, and visual disturbance, with diplopia being relatively infrequent at this stage (2). We report a patient with olfactory neuroblastoma who presented atypically with a brief history of diplopia, followed by rapidly progressive proptosis associated with profound unioocular visual loss.

### CASE REPORT

A 23-year-old woman presented to the ophthalmology department with a 3-day history of diplopia. She also reported a 1-month history of nasal stuffiness. She had no past medical or ocular history. Clinical examination revealed visual acuities of 20/20 bilaterally, with no evidence of proptosis. Her pupil reactions were normal, and optic discs were healthy bilaterally. Ocular motility assessment showed a restriction of abduction OS. She was also found to have a nodal mass in the left upper cervical area. A computerized tomography (CT) scan was performed, which showed an extensive lesion affecting the left nasal cavity, both ethmoid sinuses, and sphenoid, left maxillary, and frontal sinuses. There was extension through the skull base into the anterior cranial fossa and laterally into the orbit where there was also evidence of retrobulbar disease.

The patient was referred to otolaryngologists for nasal biopsy. Histology showed a malignant round cell tumor infiltrating the respiratory mucosa, with the tumor cells immunoreacting to the neuronal markers neuron-specific enolase (NSE), synaptophysin, and neurofilament. These appearances were believed to be consistent with the di-

agnosis of olfactory neuroblastoma. During the 2-week period of these investigations, the patient's vision OS deteriorated rapidly to perception of light only. This deterioration was associated with the development of marked proptosis and further limitation of ocular movements. In addition, she had a left relative afferent pupil defect and bilateral optic disc swelling. In view of this finding, an urgent neurosurgical and neuro-ophthalmologic opinion was sought. Magnetic resonance imaging (MRI) confirmed a large enhancing soft tissue mass probably originating in the ethmoid and sphenoid sinus regions, extending laterally into the left orbit, and displacing the muscle cone and optic nerve laterally and inferiorly (Fig. 1). The tumor extended into the brain bilaterally, occupying much of the floor of the anterior cranial fossa. There was extensive fluid within the left maxillary antrum thought to be caused by obstruction of the sinus outlet. Chest radiograph, bone marrow aspirate, cerebrospinal fluid (CSF) for cytology, and



**FIG. 1.** Axial magnetic resonance image showing the olfactory neuroblastoma extending into the left orbit.

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bone scan results were all reported as normal. CT chest, abdomen, and pelvis results were also normal.

The tumor was thought to be inoperable, and because of the risk of permanent visual loss, the patient was urgently referred to the oncology service for chemotherapy. By this stage, she had no perception of light OS. Chemotherapy consisting of intravenous ifosfamide, vincristine, and actinomycin, combined with oral dexamethasone, was started. Within 3 days, there was an improvement in her proptosis and return of diplopia, the latter suggesting some recovery of optic nerve function. The size of the left neck node was reduced by approximately half. During the next 4 months, she received a full series of six courses of chemotherapy. By the end of this treatment period, her ophthalmic status had improved considerably. Her visual acuity was 20/20 bilaterally, with diplopia only noted on extreme left gaze associated with a slight restriction of abduction. The proptosis and optic disc swelling had resolved completely, and her pupil reactions were normal.

### COMMENT

Malignant lesions of the nose and paranasal sinuses may occasionally present with ophthalmic symptoms; however, olfactory neuroblastoma, an uncommon neurogenic tumor of the olfactory region, is rarely included in the differential diagnosis. This case illustrates the importance of considering such a diagnosis even when nasal symptoms are associated with unusual presenting features such as diplopia and rapidly progressive proptosis with visual loss. It also demonstrates that in advanced cases, appropriate and prompt management with chemotherapy can result in significant recovery of visual function.

### REFERENCES

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