Sinonasal lymphoma: A case report

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Abstract
Sinonasal lymphomas are uncommon malignancies. They are difficult to differentiate from carcinomas, and immunohistochemistry is needed to make the diagnosis. We describe an unusual case of a T cell lymphoma that involved only the paranasal sinuses in a middle-aged man. The patient presented with a complete loss of vision in one eye and lateral rectus muscle palsy, but no nasal symptoms.

Introduction
Malignant lymphomas arising in the nasal cavity and paranasal sinuses are relatively uncommon, although they are the most common nonepithelial malignant tumors of the nose. An obstructive mass in the sinonasal area is more likely to be a carcinoma. When sinonasal lymphomas do occur, most are non-Hodgkin's lymphomas. A high index of suspicion and ultimately a tissue biopsy are necessary to differentiate sinonasal lymphomas from other possibilities. The incidence of sinonasal lymphomas is higher in Asian countries than in the West; these malignancies account for 2.6 to 6.7% of all lymphomas in Asia, and they are the second most common extranodal lymphoma, behind only gastrointestinal lymphoma. Non-Hodgkin's lymphomas of the sinonasal tract are an important cause of destructive lesions of the nose and midface; their course progresses slowly but relentlessly.

We report an unusual presentation of sinonasal lymphoma in which the patient presented with no nasal symptoms—only a loss of vision in one eye and lateral rectus muscle palsy. At the time of presentation, the disease had involved the orbit and extended into the anterior cranial fossa and skull base.

Case report
A 44-year-old man presented with a complaint of a sudden loss of vision in his right eye. His sight had begun to fail 20 days earlier, and the loss had become complete within 2 days. The patient reported no history of headache, double vision, loss of consciousness, neurologic deficits, nasal discharge, nasal obstruction, or epistaxis. He was not hypertensive or diabetic.

Examination determined that the patient's visual acuity in the right eye had been reduced to mild light perception. Examination also revealed an afferent pupillary defect of the right eye and right lateral rectus muscle palsy (figure 1). A fundus examination identified a pale optic disk. The remainder of the examination revealed that cranial nerve function was normal.

Anterior rhinoscopy detected an anterior deviated nasal septum with a spur to the right; no mass was seen. There was no paranasal sinus tenderness. Findings on the postnasal examination were also normal. A diagnostic nasal endoscopy did not reveal any additional findings. However, computed tomography (CT) identified a homogenously enhancing soft-tissue density that involved the right middle and posterior ethmoid cells and the entire sphenoid sinus. The mass had destroyed the orbital plate of the ethmoid sinus and the body of the sphenoid sinus (figure 2). The mass had eroded the lamina papyracea and infiltrated the medial rectus muscle. Superiorly, the lesion extended into the sellar area and eroded the right anterior clinoid process. The mass also extended inferiorly into the skull base. No extension into the maxillary sinus or nasal cavity was seen.

With a provisional diagnosis of carcinoma, we took the patient to the operating room for sinoscopy and biopsy. We performed a right uncinectomy, removed the anterior ethmoid cells, and perforated the ground lamella in order to approach the posterior ethmoids, which were filled with the soft, smooth mass. We removed the mass and sent it for histologic analysis, where it was identified as a highly cellular tumor with dense sheets of compactly arranged cells with scanty cytoplasm and hyperchromatic nuclei suggestive of a poorly differentiated carcinoma or lymphoma. To confirm the diagnosis, immunohistochemistry was performed. It revealed that the specimen was strongly positive for the leukocyte common antigen CD45, which is diagnostic of non-Hodgkin's lymphoma (figure 3).

The patient was referred for chemoradiation. He was given cycles of therapy with cyclophosphamide, hy-
droxyurea, vincristine, and prednisolone along with 60 Gy of irradiation. The patient was followed for 5 months, during which time he exhibited no evidence of any lesion on nasal endoscopy.

Discussion

Because malignant lymphomas are associated with surface crusting, widespread necrosis, and inflammation, they were once considered to be inflammatory lesions, known as lethal midline granulomas or nonhealing granulomas. It was not until the introduction of immunohistochemistry that most of these lesions were found to be malignant lymphomas.

On immunohistochemistry, the three phenotypes of malignant lymphomas are T cell, natural killer (NK) cell, and B cell. In Asian populations, more than 90% of sinonasal lymphomas are of T cell origin. In addition, many proliferating T cells have been shown to express an additional marker (CD56), which suggests an NK cell origin; these tumors are classified as T/NK cell lymphomas. Malignant lymphomas have a predilection for males, and they tend to occur in younger adults.

Nasal T cell lymphomas usually spread from their site of origin in the nasal cavity and invade adjacent structures (e.g., the paranasal sinuses, nasopharynx, and oral cavity) via palatine necrosis. In our patient, however, the tumor appeared to have originated in the ethmoid sinus and then spread to the intracranial cavity. T cell lymphomas are characterized by progressive ulceration and necrosis, which are not typical of B cell lymphomas. They are also characterized by their angiotropism or angiocentricity, as tumor cells infiltrate and destroy blood vessel walls and cause variable degrees of geographic necrosis. They express T cell markers such as CD2, CD45RO, and CD43. Also, they often express NK cell marker CD56, but they lack other NK cell markers, such as CD16 and CD57.

Numerous studies have shown that patients with T and NK cell lymphomas of the sinonasal area have a high incidence of Epstein-Barr virus (EBV) infection. Although T and NK cell lymphomas are less common in the West than in Asia, their association with EBV in the West parallels that seen in Asia and in South America. It is unclear what role EBV plays in the origin of sinonasal lymphomas, but one possible explanation involves clonal proliferation in response to viral stimulation.

Most T cell lymphomas are of intermediate grade (diffuse mixed small cell lymphomas, diffuse mixed large cell lymphomas, and diffuse large cell lymphomas) and high grade (large cell immunoblastic lymphomas). The diagnosis of T cell lymphoma can be extremely difficult to make. Histologically, these tumors are characterized by a polymorphic infiltrate that contains variable amounts of malignant cells and widespread areas of tumor necrosis. Therefore, diagnosis requires a high index of suspicion, an adequate amount of biopsy material, and immunohistochemistry.

Sinonasal B cell lymphomas, which predominate in Western populations, usually arise from the paranasal sinuses rather than the nasal cavity. They primarily involve the maxillary and ethmoid sinuses, and they extend locally to involve the orbit, cheek, and anterior cranial fossa. They generally contain a monomorphic population of large atypical lymphoid cells without a prominent mixture of reactive cells. They do not usually manifest angiocentrism or angioinvasion, and so vascular necrosis does not occur.

The usual presenting symptoms of sinonasal lymphomas are nasal obstruction and discharge, epistaxis, unilateral facial or cheek swelling, and headache. Patients may also show signs of infiltration, such as proptosis, blurred vision, and cranial nerve palsies secondary to orbital or skull base extension. T cell lymphomas are more aggressive than...
the other phenotypes, and they can cause soft-tissue and bony destruction. Dissemination is infrequent, but when it does occur, it typically involves other extranodal sites. Radiologic examination shows bony erosion, which is a predominant feature.

Patients with sinonasal lymphomas have a better prognosis than those with nodal lymphomas of similar grades. Favorable prognostic factors include young age, diagnosis at an early stage of the disease, and an absence of fever, weight loss, and night sweats. Favorable outcomes are also associated with a combination of anthracycline-based chemotherapy and locoregional radiotherapy. Local failures are more common in patients with T cell phenotypes.

References