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Grand Rounds

A 60-year-old woman with an asymptomatic left lacrimal gland mass found incidentally

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History

A 60-year-old woman with a history of recurrent, poorly differentiated adenocarcinoma of the left maxillary sinus was referred to the Orbital Oncology Service of the University of Texas MD Anderson Cancer Center for evaluation and management of a slow growing, asymptomatic left lacrimal gland mass that was incidentally identified on routine surveillance magnetic resonance imaging (MRI). The patient denied significant vision changes, diplopia, and pain associated with the lesion.

Other past pertinent oncologic and ophthalmologic history included treatments for the maxillary sinus adenocarcinoma with multiple surgical resections, including a maxillectomy and high-dose radiation of 60 Gy to the nasal cavity and 50 Gy to the right and left neck lymph nodes, and repeated proton beam therapy to the sinonasal region for a local recurrence. The patient's ocular history was significant for a bilateral dacryocystorhinostomy and left conjunctivodacryocystorhinostomy for nasolacrimal duct blockage and canalicular stenosis secondary to treatments for her sinonasal cancer. Additionally, she was diagnosed with iritis of the right eye 5 months prior to presentation by her local ophthalmologist that had resolved with topical steroids and was felt to be idiopathic in nature.

Examination

On examination, the patient's visual acuity was 20/25 bilaterally, unchanged from previous visits. Her intraocular pressure was 19 mm Hg in the right eye and 20 mm Hg in the left eye. Her pupils were normal. Her visual fields were intact, and she had full extraocular motility. The eyes were not proptotic, eyelids did not have an S-shaped or other deformity, and the lacrimal gland mass was not readily palpable. Her intraocular examination was unremarkable.

Ancillary Testing

MRI demonstrated a hyperintense and well-circumscribed, homongeneously enhancing mass measuring 1.8 × 1.1 × 1.1 cm in the superolateral aspect of the left orbit, suggestive of a lacrimal gland mass (Figure 1). Three years prior her lacrimal gland measured 0.7 × 0.5 × 0.5 cm on MR imaging, indicating slow growth over the previous 3 years.

Treatment

Given the concerns for growth of the lacrimal gland mass and the possibility of a metastasis from the patient's known previously treated recurrent sinonasal carcinoma, the patient underwent a left anterior orbitotomy with total resection of the lacrimal gland mass 2 weeks after this presentation. Gross evaluation of the surgical specimen revealed an encapsulated mass with a homogeneous yellow, tan cut surface (Figure 2). Histopathologic analysis of the tumor revealed an encapsulated tumor with a surrounding fibrous capsule (Figure 3A), consisting of a variable density of benign spindle cells with densely cellular areas (Antoni A areas, Figure 3B), including focal Verocay body formation (Figure 3C) juxtaposed with less cellular regions, where the

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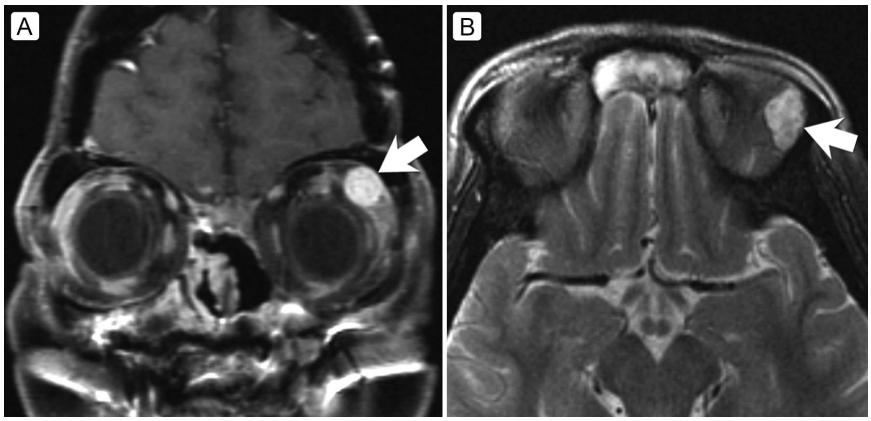

Figure 1. A, T1-weighted post-contrast coronal magnetic resonance imaging (MRI) demonstrating a well, circumscribed homogeneous mass in the left superolateral orbit. B, T2-weighted precontrast axial MRI demonstrating heterogeneous signal consistent for schwannoma.

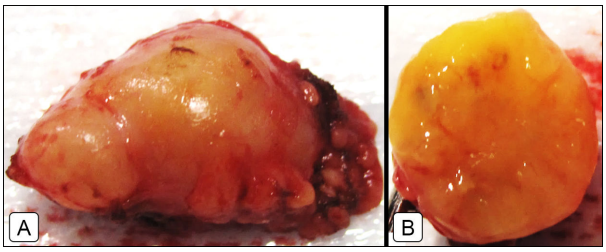

Figure 2. A, Gross surgical specimen showing the appearance of the lacrimal gland mass. B, Gross specimen in cross section.

spindle cells were loosely arranged in a myxoid background with prominent hyalinization of the tumor-associated vessels (Antoni B areas, Figure 3D).

Differential Diagnosis

The clinical history and imaging in this patient suggested a benign lacrimal gland epithelial tumor, such as a pleomorphic adenoma (PA), metastatic disease from the patient's primary adenocarcinoma, adenoid cystic carcinoma, or lacrimal gland lymphoproliferative disease. Among these, the well-circumscribed appearance and reported history of slow growth indicated a benign process. Taken collectively with the histopathologic findings, however, the diagnosis of lacrimal gland schwannoma was supported.

Diagnosis and Discussion

We describe a rare case of an asymptomatic lacrimal gland schwannoma detected incidentally on routine imaging. The annual incidence of lacrimal gland tumors is approximately 1 in 1 million, with pleomorphic adenoma (benign mixed tumor) being the most common epithelial tumor of the lacrimal gland.[1,2] Patients with lacrimal gland tumors can present with findings of diplopia, restricted ocular motility, and globe dystopia with an inferomedial shift.[3] A Schwannoma of the lacrimal gland is an extremely rare tumor; only 8 previous cases have been described in the literature.[4–9]

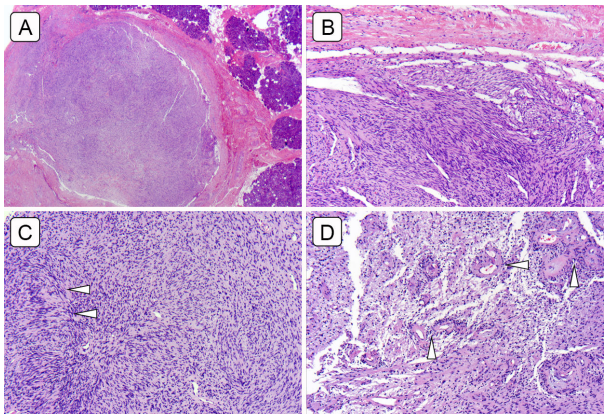

Figure 3. Lacrimal gland schwannoma. A, Scanning magnification reveals an encapsulated spindle cell tumor with areas of hyper- and hypo-cellularity adjacent to glandular tissue (hematoxylin-eosin [H&E] original magnification ×40). B, Spindle cell tumor comprised of areas of relative hypercellular Antoni A areas (H&E ×200). C, with Verocay body formation (arrowheads; H&E ×200). D, Antoni B area consisting of loose arrangement of spindle cells in a myxoid background with hyalinized thick-walled vessels (arrowheads; H&E ×200).

Schwannomas are benign, peripheral nerve sheath tumors, composed of neural crest–derived Schwann cells; they comprise 0.7%–2.3% of orbital tumors.[10,11] It commonly manifests along the supraorbital branch of

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the frontal nerve.[12] We postulate that our patient had an extension of her schwannoma from the lacrimal branch of the ophthalmic division of the trigeminal nerve.

Schwannomas classically consist of spindle cells arranged as hypercellular and alternating hypocellular regions known as Antoni A and Antoni B, respectively.[13] This biphasic phenomenon underlies the classic signal heterogeneity on T2-weighted imaging, as demonstrated in Figure 1B.[14,15] There can also be palisading arrangements of nuclei known as Verocay bodies.

The clinical and radiologic diagnosis of the lacrimal gland tumor in our patient was further hindered by her complex history of an aggressive primary adenocarcinoma of the maxillary sinus and local radiation. Our presumptive diagnosis was a benign epithelial tumor or metastatic disease. The gross specimen after surgical resection had a yellow appearance with a potatolike consistency characteristic of a pleomorphic adenoma. However, it was not until histopathologic assessment that a diagnosis of Schwannoma of the lacrimal gland was made. In addition to nonspecific S100 protein and glial fibrillary acid protein expression in schwannomas, markers such as calretinin, podoplanin, and SOX10 may also be useful in elucidating the diagnosis.[16–18] In a recent review of the literature, Yamanaka and Hayano reported 28 cases of radiation-induced schwannomas.[19] Because the primary tumor was on the left maxillary sinus and nasal cavity, the radiation field may have included the left orbit. Since the schwannoma was not present on imaging prior to radiation, it is possible that adjacent surrounding structures may have been affected thereby promoting tumorogenesis of this rare, neoplastic variety.

Our case demonstrates that, although rare, schwannoma of the lacrimal gland should be considered on the differential diagnosis of a well-circumscribed lacrimal gland mass. Gross total surgical resection is the appropriate treatment particularly if there are signs of growth or concerns for other more malignant diseases, as was the case in our patient with a history of significant malignant carcinoma of the maxillary sinus.

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