The patient, a 13-year-old male, presented with the chief complaint of a mass in the lower aspect of his right orbit which had been present since his birth. On inspection, a large mass could be seen on the inferior part of the right eye that caused superior displacement of the right globe. He denied any history of acute or chronic sinusitis, paresthesia, swelling, or facial pain. Visual acuities of the right and left eye were 4/10 and 10/10, respectively. Refractive error on the right eye was -0.25+-9.0×15°; on the left eye, it was plano. A firm mass could be palpated in the upper part of maxillary area just inferior to the right globe. In the medial aspect, the inferior orbital rim could be palpated only with firm compression over the mass. In the lateral aspect, however, the inferior orbital rim could be palpated easily over the mass. The mass was not moveable and had no tenderness. On ocular examination, superior displacement of the right globe could be seen easily (Figure 1). Some limitations in ocular motility of the right eye could be seen in all gazes, especially toward the medial and inferior aspects. Other ocular examinations were normal. His dental and nasal examinations were normal. Computed tomography (CT) showed a well-circumscribed, expansile, unilocular, hypodense lesion in the superior aspect of the right maxillary sinus with extension to the right orbital cavity (Figure 2).

What is Your Diagnosis?

See next pages for the diagnosis
The patient underwent surgery. After a subciliary incision and opening of the skin, subcutaneous tissue, and orbicularis muscle, a large mass was exposed. The mass had eroded the orbital floor and entered the orbit. A large cystic mass was opened. The main portion of the cyst and its content, which was a white sebaceous material with some hair in it, were removed. After removing the cyst, we had an opening to the maxillary sinus through which we removed the residual material of the cyst within the maxillary sinus as much as possible. At the end of the procedure, there was only some residual atrophic mucous membrane left in the maxillary sinus wall. After completion of the cyst removal and its materials, the orbital floor was repaired with insertion of a silicone sheet (Figure 3).

Macroscopically, the sac-like resected tissue measured 2×1.8×0.7 cm. In cut sections, the cyst wall was irregular and 0.1 cm thick. Microscopically, the tissue consisted of a cyst lined by squamous epithelium supported by variably dense fibrovascular connective tissue (Figure 4). Numerous foci of lacrimal glands, cartilage, smooth muscle fibers, and sebaceous tissue were identified within the tumor that was compatible with complex choristoma. Abundant keratinous material was noted within the cystic lumen.

After six months of follow-up, the patient had no problems in the nasal and maxillary area. On ocular examination, some limitations could be seen on ocular motility but superior displacement of the globe was corrected to a large extent.

Dermoid cysts are developmental choristomas
that arise from embryologic stem cell rests that have the potential to give rise to tissues of all three germ layers. They comprise 3% to 9% of all orbital masses with an average in pooled series of 4.7%. Ten percent of head and neck dermoid cysts are orbital, and in most series the upper outer quadrant dominates. The first and second cases of dermoid cyst within the maxillary sinus were reported by Torske et al and Bodner et al, respectively, and to the best of our knowledge, our case is the third report of such cases in the literature.

The natural history of dermoid cysts is slow expansion and, depending on their sites, displacement of adjacent structures. The location, relationship to bone, and cystic nature help to identify dermoid cysts. The differential diagnosis depends on the location of the mass. Any of the solid tumors should be included in the differential diagnosis, especially if there is a focal bony defect.

The pathogenesis of this lesion within the maxillary sinus remains enigmatic. The development of maxillary sinus during embryogenesis is as an outgrowth of the walls of the nasal cavity (Figure 5). As dermoid cysts are well known within the nasal cavity and nasopharynx, it is plausible that embryonic tissue entrapped within the nasal cavity may have migrated into the developing maxillary sinus, eventually giving rise to a dermoid cyst.

As noted above, the development of paranasal sinuses that are derived from ectoderm begins as evaginations of the nasal mucous membrane during the second and fourth months of pregnancy. Further development takes place after birth. The maxillary sinus is the first to begin significant pneumatization within the first year of life. It begins to enlarge laterally along the floor of the orbit at the age of three years. The floor of the maxillary sinus reaches the level of the nose by 12 years of age (Figure 5). Adult size is achieved in mid-adolescence.

Recognizing the association of dermoid cysts with sinus tracts, the phrase “nasal dermal sinus cyst” was introduced by Sessions. While the exact embryopathology related to their development is unknown, a basic premise is that these cysts are formed from incomplete sequestration of neuroectoderm arising from epithelial ectoderm. This separation normally occurs between the third and fifth weeks of intrauterine life.

The association of choristomas with various craniofacial and systemic defects is well-documented in the literature. Nasal choristoma may be associated with bony cranial defects, intracranial abnormalities, CSF leakage, and the potential for fatal meningitis, if not handled properly.

The finding of a choristoma may signal a simultaneous occurrence of other lesions of an analogous origin in other localities than that of the bulbar or the orbit, which should lead the ophthalmologist to a complex examination of the patient and an interdisciplinary cooperation with a neurologist, otolaryngologist, and dermatologist, among other specialties. We believe that the orbital floor defect in our case was secondary to erosion caused by the dermoid mass; hence, it was an acquired lesion rather than a congenital defect.

The treatment of these lesions can be complicated owing to their size, location, and involvement of orbital structures and should not be undertaken by the occasional orbital surgeons. The operative approach should be based on thorough preoperative assessment of the size, location, extent, and relationship to adjacent structures. The base of the lesion is thought to be the active growth center. Thus, total removal of the lesion is mandatory to prevent recurrence or fistulization.

References