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CASE REPORT

Transnasal endoscopic marsupialization for a large midline maxillary odontogenic keratocyst in a 6-year-old child

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Summary An odontogenic keratocyst is a histologically distinct lesion that is thought to arise from basal cells of the oral epithelium, usually occurring in the posterior portion of the mandible or the mandibular ramus. We report on a large odontogenic keratocyst that presented as nasal airway obstruction in a 6-year-old boy. To our knowledge, this is the youngest patient for whom a midline odontogenic keratocyst in the anterior maxillary region has been documented in the English literature. We designed a less-invasive surgical approach for transnasal endoscopic marsupialization instead of enucleation of this large pediatric odontogenic keratocyst.

1. Introduction

The odontogenic keratocyst (OKC) was first described by Philipsen in 1956 [1]. Approximately 65% of OKCs occur in the mandible, with a predilection for the third molar-ramus region [2,3]. However, OKCs can occur anywhere within the jaws and can mimic other lesions. Reports in the pediatric literature are sparse.

The following case report documents a large OKC that caused nasal airway obstruction and mimicked a nasopalatine duct cyst in a 6-year-old boy. We discuss the clinical features, diagnosis, and treatment of OKCs, along with a transnasal endoscopic marsupialization surgical approach that we used for this case. Based on the postoperative observations,

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we also propose that this less-invasive surgical approach is appropriate for managing developmental cystic lesions around the nasal cavity.

2. Case report

A 6-year-old boy presented with a 3-month history of worsening snoring and bilateral nasal obstruction. Examination revealed a large, nontender, nonpulsatile mass filling the lower aspect of the nasal cavity bilaterally. The overlying mucosa was intact. No definite abnormality was seen on the oral surface of the palate. The rest of the head and neck and systemic examination was unremarkable. A preoperative computed tomography (CT) scan revealed a cystic lesion at the midline of nasal floor (Fig. 1). The extensive bone remodeling with intact cortices enclosing a homogeneous fluid-filled cavity indicated a benign origin. The preoperative diagnosis of a nasopalatine duct cyst was made.

A transnasal endoscopic marsupialization method via the right nostril was designed to treat this patient. With the patient under general anesthesia, the right nasal cavity was topically anesthetized and decongested with cotton gauze soaked in a solution of 1% lidocaine and 1:100 000 epinephrine. Later, the protruded cyst roof was locally infiltrated with 1% lidocaine containing 1:100 000 epinephrine. The roof of the cyst, which was firmly attached to the mucous membrane of the nasal floor, was removed transnasally with a sickle knife and scissors. There was a large amount of whitish turbid fluid aspirated from the cyst. Under the guidance of a nasoendoscope, the opening of the cyst was thoroughly widened with bite forceps. The edges of the nasal mucosa and the cyst lining were adequately cut to closely match one another. Multiple biopsies of the residual cyst wall were done at the same time. Loose nasal packing was applied at the end of the procedure and removed after 48 h.

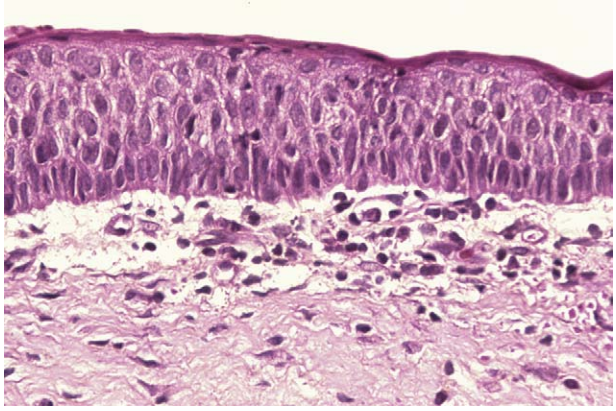

Fig. 2 The histological section shows a uniform epithelial thickness of 5—7 cells, palisaded basal cell layers, and a corrugated parakeratinized surface.

Histopathologic examination of the surgical specimen revealed a uniform epithelial thickness of 5—7 cells, palisaded basal cell layers, and a corrugated parakeratinized surface (Fig. 2). The features were consistent with an OKC.

The boy was seen on a weekly basis for 4 weeks for debridement of the nasal floor area. He was also instructed to flush the cavity by gentle nasal douching to remove the mucus accumulation in the pocket. He underwent CT scanning of the sinuses in the postoperative follow-up period. Six months after surgery, neither dentition displacement nor midface deformity was noted. The cyst had decreased in size and had a patent opening in the right nasal floor (Fig. 3). The patient's difficult nasal breathing and snoring had completely resolved.

The postoperative period was uneventful until a traffic accident supervened 3 years after surgery. The resultant facial swelling and submental laceration brought this patient to our hospital again. Radiographic investigation by panoramic X-ray revealed multiple cystic lesions in the mandibles and maxillae. A CT scan revealed multiple expansile

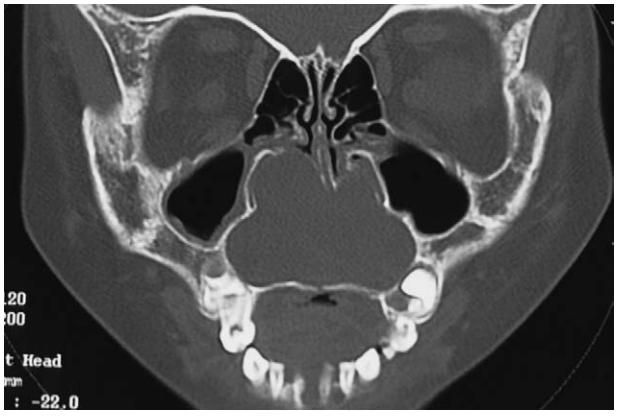

Fig. 1 A preoperative coronal computed tomography scan revealed a midline cystic lesion of the maxilla.

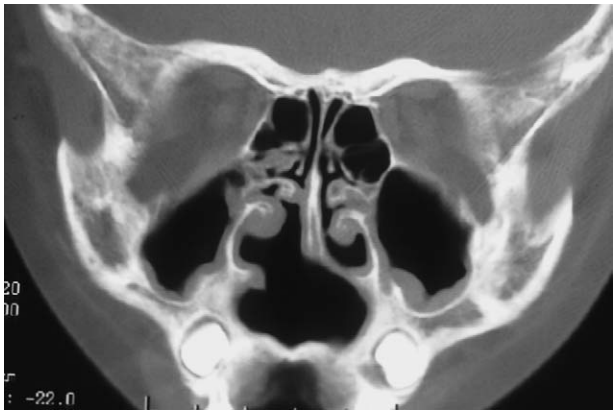

Fig. 3 Six months after surgery, the cyst had decreased in size, and it had a patent orifice in the right nasal floor.

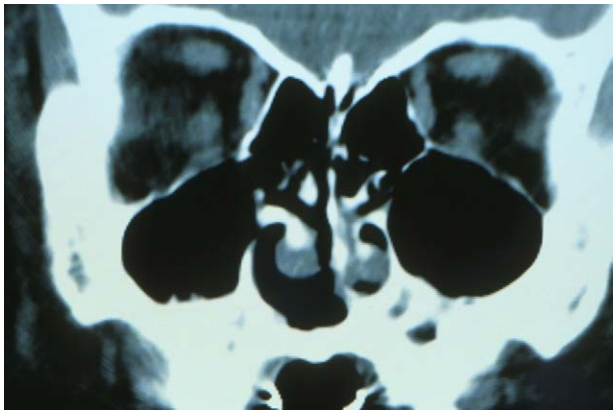

Fig. 5 Five years after surgery, the previously marsupialized midline odontogenic keratocyst was replaced by an air-containing sinus with a wide opening in the right nasal floor.

cystic changes on both sides of the body and angle of the mandible (Fig. 4). These multiple OKCs indicated the possibility of nevoid basal cell carcinoma (Gorlin's) syndrome, although there is no documented presence of this syndrome in his family.

The patient has undergone several maxillofacial surgeries for the subsequent OKCs. However, 5 years after the original surgery, the large midline OCK that we had previously marsupialized was replaced by an air-containing sinus with a wide opening in the right nasal floor (Fig. 5).

3. Discussion

The odontogenic keratocyst (OKC) is a histologically distinct lesion that is thought to arise from basal cells of the oral epithelium [4]. However, the pathogenesis of OKCs in the anterior midline maxillary region is not consistent. Woo et al. considered OKCs in this location to be developed from the primordium of a mesiodens, a common supernumerary tooth [5]. On the other hand, Neville et al. suggested that these cysts arise from remnants of the dental lamina in the anterior maxilla. Odontogenic keratocysts in the anterior midline maxillary region also exhibit different demographic features [6]. Odontogenic keratocysts in this particular location tend to occur in older individuals, with a mean age of 70 years; while the peak incidence for OKCs in general is in the second and third decades of life [3,5,6].

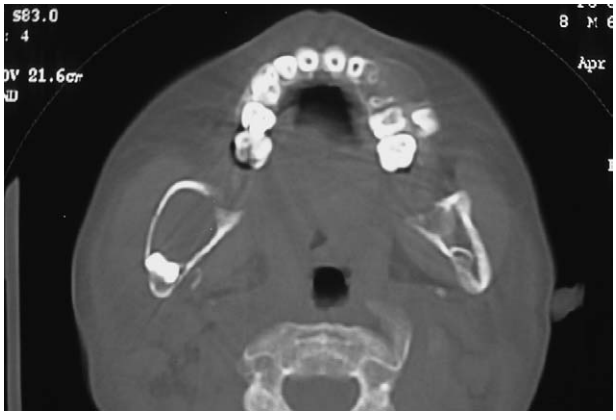

Fig. 4 A CT scan 3 years after surgery revealed multiple expansile cystic changes on both sides of the body and angle of the mandible.

The OKC is the specific type of odontogenic cyst that may be associated with the nevoid basal cell carcinoma syndrome (Gorlin's syndrome) [3,7,8]. Gorlin's syndrome comprises multiple OKCs, early basal cell carcinoma, skeletal developmental anomalies, dyskeratotic pitting of the hands and feet, and dural calcification. The OKCs associated with Gorlin's syndrome are usually multiple with a greater potential for proliferation and a higher tendency for recurrence. Pediatric OKCs should alert the clinician to the possible underlying diagnosis of nevoid basal cell carcinoma syndrome [9].

The characteristic clinical presentation includes localized swelling; spontaneous drainage of cystic fluid is common. Other manifestations include pain, trismus, cellulitis, and occasionally paresthesia or displacement of teeth [9]. Nasal obstruction is unusual except for the OKCs that are located in the anterior midline maxillary region [10]. Most studies report a slight male and white predominance [2,3,6].

The diagnosis of OKC was made after direct histopathologic examination of the specimen. Radiographically, it is not possible to differentiate OKCs from dentigerous cysts. The histopathologic characteristics include a uniform epithelial thickness of 5—7 cells, a basal layer of hyperchromatic cuboidal cells, and a thin, corrugated layer of parakeratin on the epithelial surface.

The appropriate initial therapy for an OKC is enucleation with or without extraction of the associated dentition. Some adjunctive treatments, including Carnoy's solution (tissue fixative containing absolute alcohol, 6 mL; chloroform, 3 mL; glacial acetic acid, 1 mL; ferric acid, 1 g) fixation of the cyst wall before enucleation; marsupialization in combination with secondary enucleation; and combination of enucleation and liquid nitrogen cryotherapy, may decrease recurrence rate [11,12]. Recurrent OKCs indicate the need for more aggressive surgical management that includes maxillectomy or mandibulectomy [13].

The clinical presentation of this 6-year-old boy was not typical for an OKC. It is extremely rare for a 6-year-old boy to develop a large OKC in the anterior

midline maxillary region; in fact, this is the youngest patient for which such a lesion has been documented in the English literature [3,9]. The differential diagnosis of a solitary cystic lesion in the anterior maxillary region may involve consideration of various forms of fissural cysts, odontogenic cyst, dermoid cyst, and benign primary bone tumor or cyst. However, the diagnosis of an OKC was not suspected at the time of surgery.

The large midline cyst with extensive involvement of deep structures in a pediatric patient presented a special treatment dilemma: complete extirpation of this extensive midline nasal cyst in this 6-year-old boy might have been difficult or have led to significant morbidity, but surgery was clearly indicated. Therefore, we used a less-invasive surgical approach that involved transnasal endoscopic marsupialization of this cyst. This approach not only relieved the difficult nasal breathing, but it also minimized morbidity, and the patient's dental and craniofacial development was not disturbed. Although 3 years later this patient developed multiple OKCs in other areas of the jaws, we considered the transnasal endoscopic marsupialization to have been an effective method for the first OKC. The subsequent OKCs that developed in this patient were additional primary cysts rather than recurrences of the original one. Based on the observation of postoperative sinus CT scans, we propose that this transnasal endoscopic marsupialization method may also be the treatment of choice for nasopalatine duct cysts that protrude toward the nasal cavity. Su et al. proposed a similar approach in surgical management of nasolabial cysts [14]. The same approach may also be feasible in treating various forms of developmental cysts that surround the nasal cavity.

In conclusion, maxillary OKCs should be considered in the differential diagnosis of a midline maxillary lesion, even in the pediatric population. Long-term follow-up is necessary following initial OKC treatment. Pediatric OKCs, particularly those that are multiple or recurrent, should alert the clinician to the possible underlying diagnosis of nevoid basal cell carcinoma syndrome. Treatment of OKCs ideally involves early diagnosis and complete removal of the cyst lining. A tailored procedure that takes into account the lesion site and extent and patient age is important in preventing potential complications. In our experience, transnasal endoscopic marsupialization was a less-invasive surgical approach and produced a satisfactory result. In addition, we believe this treatment modality is suitable in managing various forms of developmental cysts surrounding the nasal cavity, especially for pediatric patients with large cysts.

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