Case Report

Granular cell tumor of the left maxillary paranasal sinus in a 24-year-old man

Chia-Fen Yang, Szu-Ying Chin

1. Introduction

Granular cell tumors (GCTs) are uncommon lesions of uncertain etiology and histogenesis and the large majority are benign [1–3]. They are characterized microscopically by sheets and nests of large, polygonal, and pale to eosinophilic tumor cells with a granular cytoplasm and small, centrally placed nuclei. GCTs more commonly occur during the third to the fifth decades of life [3]. They can occur in any part of the body but appear most frequently in the head and neck region, with the tongue being the most common site [3,4]. Lesions involving the naso-paranasal areas are extremely rare. Only two cases of a GCT originating from the nasal septum and one case arising in the maxilla, extending to a paranasal sinus and nasal cavity, have been reported [1,5,6]. We describe a GCT arising from a paranasal sinus (left maxillary sinus) in a 24-year-old man, which is believed to be the first reported case of GCT as a primary lesion in this location in the English literature.

2. Case report

A 24-year-old man complained of a bilateral nasal obstruction with nose itching, nasal mucoid discharge and postnasal drip for approximately 7–8 years. He had received a bilateral multiple sinusectomy for this condition twice previously. A computed tomography scan with contrast medium showed thickened mucoperiosteum in the bilateral paranasal sinuses, especially the left maxillary sinus, causing obstruction of the left ostiomeatal unit (Fig. 1A). Clinical symptoms, examination and image studies all indicated chronic sinusitis. He received another bilateral multiple sinusectomy.

Gross examination of the specimen showed multiple pieces of soft to elastic mucosal tissue and bony fragments measuring up to 0.6 × 0.4 × 0.3 cm. No identifiable tissue was seen. Microscopically, the sections showed pieces of bilateral paranasal sinus tissue and bone with moderate lymphoplasmacytic and eosinophilic infiltrates, as seen in chronic sinusitis. In the materials from the left side, there were four pieces of fibrous tissue infiltrated by ill-defined sheets and groups of plump, eosinophilic cells. The largest infiltrative focus measured about 1.2 × 0.2 cm. These cells were polygonal or elongated with indistinct cell membranes, abundant granular cytoplasm, central small dark nuclei and inconspicuous nucleoli (Fig. 1B). The cellular groups were present in the bone marrow at one focus (Fig. 2), indicating bony invasion. No tumor necrosis, mitotic figures, or nuclear pleomorphism was evident. These cells were strongly and diffusely positive for S-100 protein (Fig. 1C). The cytoplasmic granules were strongly highlighted by periodic acid Schiff (PAS) staining, which was diastase resistant (Fig. 1D). It was confirmed to be a granular cell tumor. The tumor cells did not show connection to the mucosal epithelium and pseudoepitheliomatous hyperplasia was not observed.

The postoperative course was smooth and there was no evidence of recurrence at the 6-month follow-up.
3. Discussion

A report of five cases of GCTs by Abrikossoff [7] in 1926 is often referred to as the first description of a GCT [5]. However, Ordonez, in his recent review, referred to even earlier descriptions of similar lesions by Virchow and Weber in 1854 [3]. Several different cellular origins of this lesion have been proposed, including myocytes [3,8,9], fibroblasts [3,10], histiocytes [3,11], undifferentiated mesenchymal cells [3,12], and Schwann cells [3,13,14]. At present, most investigators believe that GCTs have a neural origin, documented ultrastructurally and immunohistochemically [3].

GCTs can affect people of any age but are more common during the third through fifth decades of life [3]. GCT may occur in all organs and tissues [5,15,16]. About one-half of reported cases arose in the head and neck region [3,17–20] and the tongue was the most common site of involvement, comprising 23–28% of all GCTs and 65–85% of those in the oral cavity [3,4,17,18,21,22]. The larynx and lip are the most commonly affected sites within the upper aerodigestive tract [21,23].

GCTs are characterized microscopically by sheets and nests of large, polygonal, and pale to eosinophilic tumor cells with a granular cytoplasm and small, centrally placed nuclei. The correct diagnosis is usually not difficult because of the typical microscopic appearance of hematoxylin and eosin-stained sections. On occasion, a GCT can be confused with a reactive process or with a neoplastic condition. In these instances, positive staining for S-100 protein and PAS-positive cytoplasmic granules are very valuable in the differential diagnosis. When the lesion occurs in the alveolar ridge of the mouth, the GST must be differentiated from congenital epulis, which has very similar cells to GCTs. Congenital epulis is only seen in newborn infants and pseudoepitheliomatous hyperplasia is not present. Pseudoepitheliomatous hyperplasia is commonly noted in GCTs closely attached to the epithelium. Other neoplastic tumors with cells which may display an abundant granular-like appearance include granular cell ameloblastoma and rhabdomyoma. These diseases usually reveal a more typical microscopic appearance, at least in some areas. Immunohistochemical and PAS stains also help to demonstrate GCTs. Groups of histiocytes in a reactive process can simulate the appearance of a GCT to some degree, but they do not cluster to the same extent and their nuclei are larger and more variable in shape [3].

A GST occurring in and involving the naso-paranasal region is extremely rare, and only three cases have been reported previously [1,5,6]. Our case is believed to be the first report in the English literature of a GST originating from the paranasal sinuses. Table 1 [1,5,6] lists these three cases along with the present case. Two
Gusts arising from the nasal septum were reported by Hwang et al. Table 1 Fanburg-Smith et al. described by Salman et al. [6]. A review of the literature showed that were benign and occurred as a solitary lesion. Sinus and nasal cavity [6]. All four reported cases, including ours, in 1989 occurred in the maxilla and extended to a paranasal forms of GCT are even rarer, occurring in 1% of cases [1, 2, 25]. Clinically, malignancy is suggested by local recurrence, rapid growth, tumors larger than 5 cm, and metastases [1, 2]. Bone invasion is not encountered.

The treatment of benign GCT varies from conservative excision [1, 26] to wide excision [1, 18]. A wide local excision with a 2–3 cm margin on all sides is advised by some investigators [3]. Local recurrence of benign lesions is seen in less than 7% of cases [1, 26], and most of those are directly related to inadequate excision. At present, there is little information on the response of malignant GCTs to therapeutic modalities other than surgery [3]. Salman et al. did not report the patient status after wide excision of a biologically aggressive and destructive case [6]. Our 24-year-old patient had no evidence of tumor recurrence 6 months postoperatively. Because bone invasion and suspicion of incomplete excision were noted in our case, close follow-up is highly recommended to detect any local recurrence.

In summary, we report the first case of GCT arising from the paranasal sinuses with a clinical presentation of chronic sinusitis. GCT, although uncommon, should be considered in the differential diagnosis of a patient with chronic sinusitis and polyposis of the paranasal sinuses. Because suspicion of incomplete excision of the tumor and bone invasion were noted microscopically, intensive clinical follow-up is recommended for our patient.

References

[25] D’Amore ESG, Ninno V. Tumors of the soft tissue composed of large eosino