



Case Report

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

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ABSTRACT

Granular cell tumors (GCTs) are uncommon benign lesions which usually occur in the head and neck region. However, paranasal sinus presentation is extremely rare. We report a case of a 24-year-old man with clinical symptoms of chronic sinusitis, which is believed to be the first reported case arising from the paranasal sinuses in the English literature. His symptoms included bilateral nasal obstruction, discharge, postnasal drip, and hyposmia. He received a multiple sinusectomy and there was no evidence of recurrence in the 6-month follow-up. We also summarize the clinical information, clinical presentation, microscopic picture, treatment, and patient status after treatment of three previously reported cases involving naso-paranasal areas along with our case. The diagnostic criteria for malignant transformation and the role of bone invasion are also reviewed.

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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 tumor 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.

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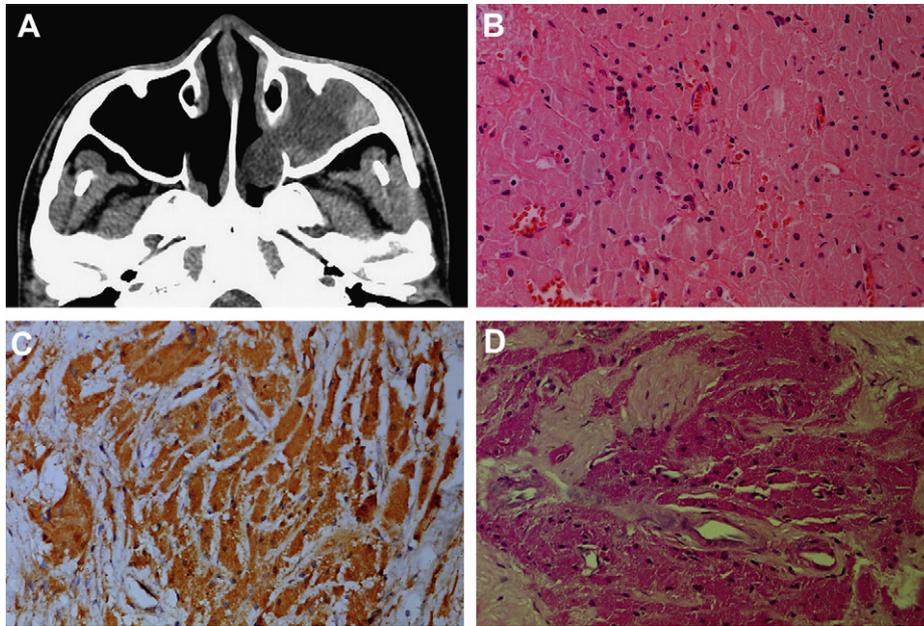


Fig. 1. (A) Computed tomography image revealing thickened mucoperiosteum in the bilateral paranasal sinuses, especially the left maxillary sinus, causing obstruction of the left ostiomeatal unit (asterisk); (B) granular cell tumor with cells with indistinct borders, abundant granular cytoplasm, small dark nuclei, and inconspicuous nucleoli (hematoxylin and eosin stain; $\times 400$); (C) positive cytoplasmic staining with S-100 protein (S-100 protein; $\times 400$); (D) intracytoplasmic granules are strongly positive for periodic acid Schiff stain and diastase resistant. (Periodic acid Schiff with diastase; $\times 400$).

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 GSTs. Congenital epulis is only seen in newborn infants and pseudoepitheliomatous hyperplasia is not present. Pseudoepitheliomatous hyperplasia is commonly noted in GSTs 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

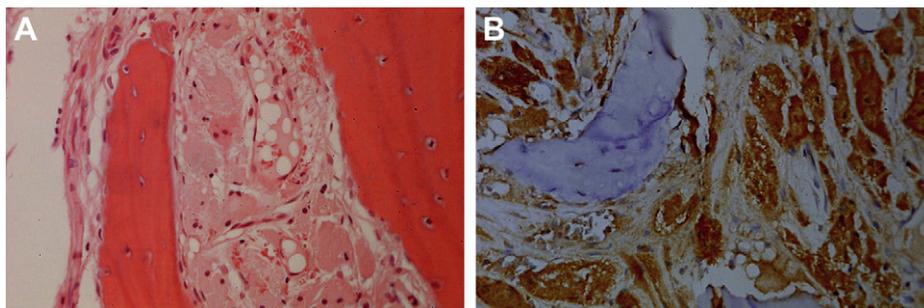


Fig. 2. (A) Tumor occupying marrow spaces, revealing bony invasion. (hematoxylin and eosin stain; $\times 400$); (B) tumor cells present in marrow spaces highlighted by S-100 protein (S-100 protein; $\times 400$).

Table 1
Granular cell tumor involving naso-paranasal region.

Reference	[6]	[1]	[5]	Present case
Age (y)/sex	22/male	6/female	69/female	24/male
Race	Black	Chinese	Japanese	Chinese
Symptoms	Right facial swelling and right intraoral mass	Nasal pain and discharge from the left nostril	Continuous serous discharge, stuffiness and occasional bleeding from the right nasal cavity	Bilateral nasal obstruction, discharge, postnasal drip, hyposmia
Symptom duration	2 wk	1 mo	Several years	Several years
Clinical impression	Right intraoral mass occupying alveolar ridge	Left nasal polyp	Right nasal polyp	Chronic sinusitis with polyposis
Tumor location	Right anterior maxilla, invading maxillary sinus and nasal cavity of the same side	Left anterior nasal septum	Kiesselbach's area of right nasal septum	Left maxillary sinus
Tumor size	Not mentioned	6 mm × 5 mm × 3 mm	19 mm × 14 mm	Pieces of tumor fragments measuring about 13 mm × 4 mm in total
Bone invasion	Yes	No	No	Yes
Treatment	Partial maxillectomy, split thickness skin graft and dental prosthesis	Excision	Excision with cauterization	Multiple sinusectomy
Patient status	Not mentioned	NED	NED	NED

NED = no evidence of disease.

GSTs arising from the nasal septum were reported by Hwang et al in 2001 [1] and Sasaki et al in 2007 [5]. One case reported by Salman et al in 1989 occurred in the maxilla and extended to a paranasal sinus and nasal cavity [6]. All four reported cases, including ours, were benign and occurred as a solitary lesion.

Bone invasion was noted in the present case and the one described by Salman et al [6]. A review of the literature showed that this feature does not seem to be potentially malignant. Malignant forms of GCT are even rarer, occurring in 1–2% of cases [1,2,25]. Fanburg-Smith et al clarified six histological criteria for malignant GCTs [24]: necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity, a high nuclear to cytoplasmic ratio, and pleomorphism. We did not observe any of these microscopic characteristics in our case. However, metastasis of histologically bland lesions has been reported [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 [1,8]. 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 GSTs 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.

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