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Case Report

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Granular cell tumor that originated in a posterior ethmoid sinus

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ABSTRACT

A granular cell tumor (GCT) is a rare neoplasm. It grows slowly, presumably originates from a Schwann cell, and is typically benign. Histopathologically, GCTs are composed of loosely infiltrating sheets of large, pale, polyhedral cells with abundant granular eosinophilic cytoplasm and a pale, centrally situated nucleus. Immunohistochemically, GCTs express the S-100 protein and neuron-specific enolase. A GCT can occur anywhere in the body. Half of all GCTs occur in the head and neck regions, especially on the tongue, but they are rarely found in the nasal cavity. A GCT usually arises as a solitary tumor and can be confirmed only by a histologic examination. The appropriate treatment is excision of the lesion. Here, we present a rare case of a GCT originating in the right posterior ethmoid sinus in the nasal cavity. A GCT originating in a posterior ethmoid sinus has not been reported thus far. In our case, a simple nasal polyp was found in the left ethmoid sinus of the patient. Thus, we initially misjudged the GCT in the right nasal cavity as a simple nasal polyp.

Keywords: Paranasal sinus neoplasms, Granular cell tumor, Ethmoid sinus

INTRODUCTION

A granular cell tumor (GCT) is a rare neoplasm. It grows slowly, presumably originates from a Schwann cell, and is typically benign.[1,2] Although a GCT usually arises as a solitary benign tumor, it is rarely malignant.[3,4] This neoplasm is composed of loosely infiltrating sheets of large, pale, polyhedral cells with abundant granular eosinophilic cytoplasm and a pale, centrally situated nucleus. GCTs can occur anywhere in the body. Only a histologic examination can confirm the presence of a GCT. The appropriate treatment is excision of the lesion.

Half of all GCTs occur in the head and neck regions, especially on the tongue.[2] However, GCTs are rarely found in the sinonasal region. Only two cases of GCTs originating in the nasal septum and two cases of GCTs arising in the maxilla have been reported; a GCT originating in a posterior ethmoid sinus has not yet been reported.

Here, we present a rare case of a GCT arising in a posterior ethmoid sinus, a site of frequent mucopurulent rhinorrhea and nasal obstruction for the patient. To the best of our knowledge, this is the first report of a GCT originating in a posterior ethmoid sinus.

CASE REPORT

A 50-year-old woman visited our clinic with complaints of recurrent mucopurulent rhinorrhea from both nostrils and obstruction of the right nostril, both of which had been occurring for 3 years. An endoscopic examination revealed that the nasal septum was deviated to right, and a pedunculated polypoid mass that originated in the right posterior ethmoid sinus was seen at the ipsilateral superior meatus. The tumor was about 1 cm; its smooth

pink surface looked more opaque than the surface of an ordinary nasal polyp. An examination of the larynx, pharynx, and oral cavity did not reveal any tumors. The patient had no history of other diseases.

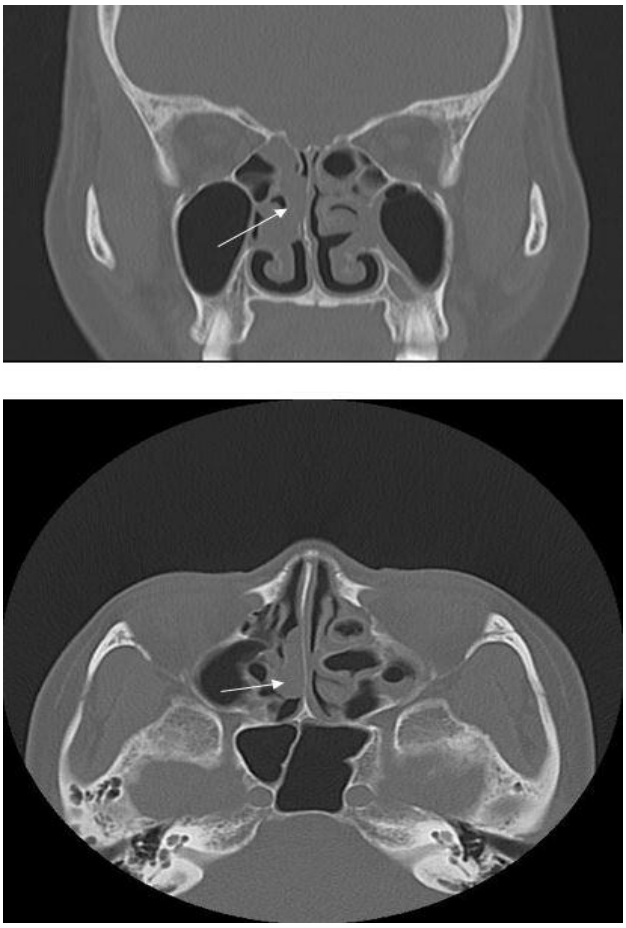

Figure 1: A non-contrast-enhanced computed tomography image showing an area in the paranasal sinus with the density of soft tissue in the right superior meatus and right posterior ethmoid sinus, but this area seemed no different than a normal nasal polyp.

On a paranasal sinus, noncontrast-enhanced computed tomogram, an area with a density similar to that of soft tissue was seen in the right superior meatus and right posterior ethmoid sinus, but this area seemed no different from a normal polyp (Figure 1). We performed endoscopic sinus surgery. The right nasal polypoid mass was subsequently excised.

Histologically, the polyp was composed of sheets of large oval cells containing abundant eosinophilic cytoplasm with fine granules. The tumor cells were small to medium in size and contained centrally located, round-to-oval nuclei. Mitotic figures were not noted (Figure 2A). Immunohistochemical stains indicated that the tumor cells were positive for the S-100 protein and CD68 (Figure 2B, C). The Ki-67 labeling index was less than 1%. The cells were negative for the epithelial membrane antigen and desmin. These findings were consistent with those for GCTs. After 16 months of follow up, there were no recurrences.

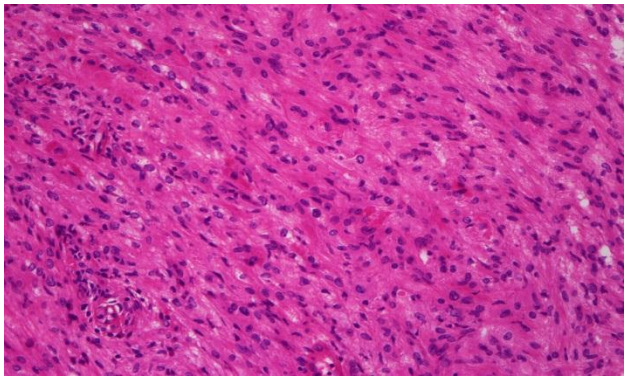

Figure 2A: In the tumor, large polygonal cells with abundant eosinophilic granular cytoplasm had proliferated (hematoxylin and eosin stain, ×200).

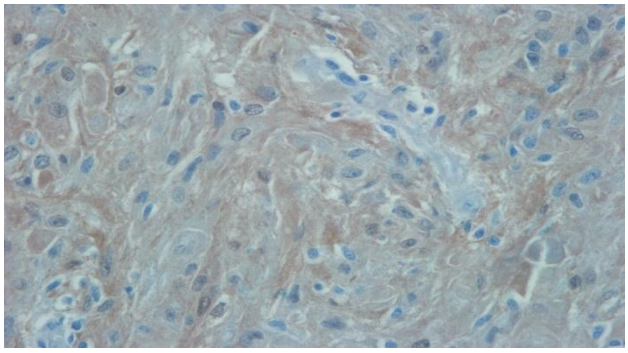

Figure 2B: Immunohistochemical staining indicates that the tumor cells were positive for s-100 (s-100, ×400).

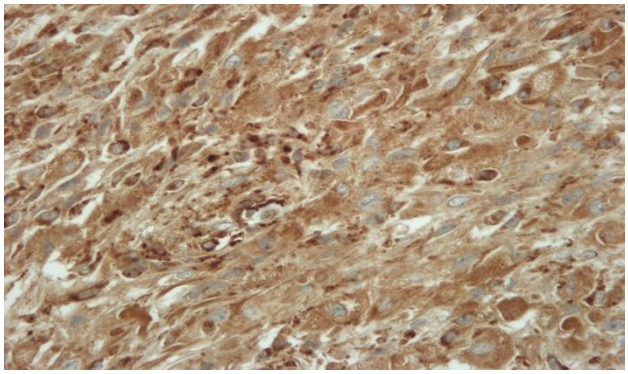

Figure 2C: Immunohistochemical staining indicates that the tumor cells were positive for cd68 (cd68, ×400).

DISCUSSION

Nasal masses can have a wide variety of possible diagnoses, including inflammatory proliferations, infective lesions, and neoplastic conditions that are either benign or malignant. A GCT is one of these possibilities.

GCTs have occurred in patients as young as 11 months and as old as 104 years, but these lesions occur most frequently in patients who are in between their fourth and sixth decades.[5] GCTs frequently arise in the head and neck regions; they are found most often on the tongue, but also occur on the skin and in the stomach, bronchi, and bile ducts.[6] Within the head and neck regions, GCTs have been found in the larynx, soft palate, labial mucosa, uvula, oral floor, gingivae, orbits, lacrimal sacs, nasolacrimal ducts, and parotid glands. GCTs are rarely found in a paranasal sinus: only four such cases have been reported. [6–9] In these cases, the tumors were found in the nasal septum or a maxillary sinus. Thus, this is the first report of a GCT occurring in a posterior ethmoid sinus (Table 1).

Table 1: Summary of granular cell tumors of the nasal cavity reported in the literature.

Case	Report	Age (years)	Sex	Location	Size (cm)	Presentation	Treatment	Outcome
1	Hwang (2001)	6	F	Septum	0.6	Nasal discharge	Excision Biopsy	NED
2	Sasaki (2007)	69	F	Septum	0.19	Recurrent epistaxis	Excision Biopsy	N/A
3	Salman (1989)	22	M	Maxilla sinus	N/A	Facial swelling	Partial maxillectomy	N/A
4	Yang (2012)	24	M	Maxilla sinus	0.13	Nasal discharge Hyposmia	Multiple sinusectomy	NED

Abbreviations: F, female; M, male; N/A, Not applicable; NED, No evidence of disease.

In general, surgical excision is the first choice of treatment for GCTs originating in common sites such as the skin. However, because they rarely arise in the nasal cavity, there are no accepted standards for the management of GCTs that originate there.

GCTs are composed of loosely infiltrating sheets of large, pale, polyhedral cells with abundant granular eosinophilic cytoplasm and a pale, centrally situated nucleus.[10] Immunohistochemically, GCTs express the S-100 protein and neuron-specific enolase. In addition to the S-100 protein, positive expression of CD68, a histiocytic marker, was reported in 77.7–100%, a finding that indicates the presence of intracytoplasmic lysosomes in GCTs.[11]

Although the majority of these tumors are benign, a rare malignant variant (less than 2% of all GCTs) exists. It is aggressive; its local recurrence rate has been reported to be as high as 70%, and its 3-year survival rate is less than 50%.[9] Differentiation between benign and malignant variants is based mainly on the presence of necrosis, increased mitotic activity, spindling of tumor cells, vesicular nuclei with large nucleoli, and a Ki-67 labeling index greater than 10%.[12] No adverse histologic features were seen in the present case.

GCTs usually occur in the head and neck regions; they originate most often on the tongue and in the oral cavity (30–50%), which are areas with high densities of peripheral nerves.[11] In one study, GCTs were positive for the S-100 protein and neuron-specific enolase, and the findings from an ultrastructural examination indicated that the GCTs differentiated from Schwann cells.[11] Therefore, GCTs are believed to originate from Schwann cells.

Histopathologically, GCTs can mimic granular cell variants of other tumors, such as leiomyoma, dermatofibrosarcoma, and angiosarcoma. Immunohistochemistry can be used to confirm a GCT diagnosis: GCTs are consistently positive for the S-100 protein and consistently negative for desmin, cytokeratins, the smooth muscle antigen, and the epithelial membrane antigen.

CONCLUSION

In summary, this is an unusual case of a GCT arising in a posterior ethmoid sinus and causing a nasal obstruction. Although GCTs rarely occur in this region, they should be considered in the differential diagnosis of a nasal mass.

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