Case Report

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An Extremely Rare Case of Granular Cell Tumour of Right Nasal Vestibule

Pathology Section

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ABSTRACT

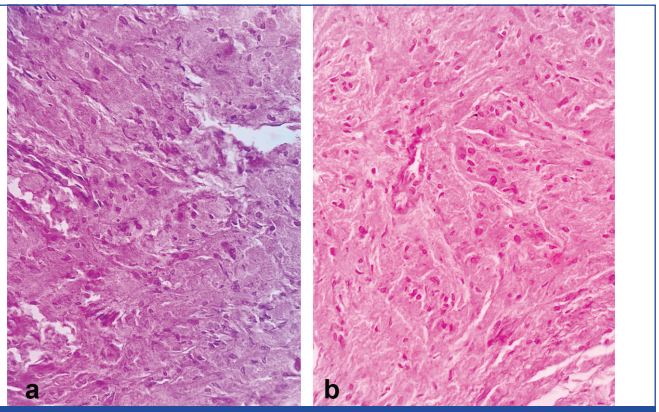
Granular Cell Tumour (GCT) of nasal and paranasal area is an extremely rare entity, while the most common site is the tongue in oral cavity. GCT was first time described by Abrikossoff in 1926, which occurs in 3rd to 5th decades of life and most frequently involving the head and neck region. Abrikossoff in 1926, first time described GCT, to be derived from smooth muscle and gave the term myoblastomas, also known as Abrikossoff's tumours. Schwann cell origin of GCT was demonstrated on immunohistochemistry through positive S100 protein identification. Herein, the authors report a case of GCT arising from right nasal vestibule in a 28-year-old female who presented with swelling since three months. On gross examination, the specimen revealed firm red and white pieces of tissue measuring 0.8x0.8x0.5 cm. Diagnosis was made by histopathological examination which was confirmed by PAS special stain and S100 immunohistochemistry. Author also abridged the clinical information, microscopic features, treatment and patient status after follow-up. This case is believed to be the 7th reported case of GCT as primary lesion at this location.

Keywords: Myoblastomas, Premalignant, S100, Sphingomyelin

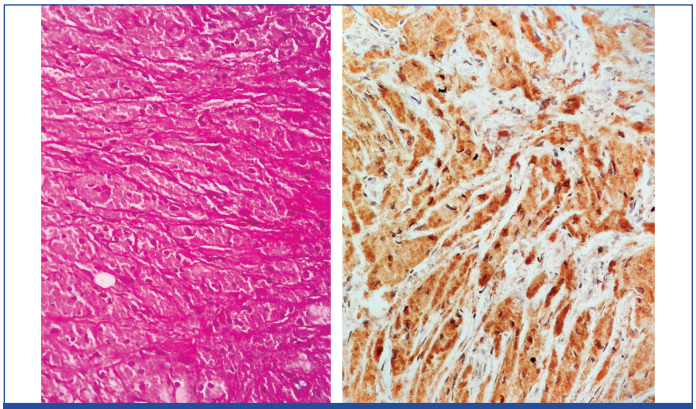
CASE REPORT

A 28-year-old female presented to Ear Nose Throat Outpatient Department with swelling over right nasal vestibule and right side nasal obstruction for three months. There was no history of nasal discharge, nose itching and epistaxis. On local examination, the swelling was small in size measuring 1.0×1.0 cm and was firm to hard in consistency with restricted mobility. The provisional diagnosis of alar cyst was given. There was a past history of the swelling at similar site; the swelling was excised in other hospital and had been sent for histopathological examination in a pathology lab and diagnosed as epidermoid cyst. Complete blood count, liver function test and kidney function test were found within normal limits. The swelling was excised and sent for histopathological examination in the Department of Pathology of this Institute.

On gross examination, the specimen revealed firm red and white pieces of tissue measuring 0.8×0.8×0.5 cm; the whole tissue was processed and 3 to 5 µm thick sections were cut and stained with Haematoxylin and Eosin (H&E) stain. On microscopic examination, the sections examined revealed sheets of round to polygonal cells with ill-defined cell borders, finely granular eosinophilic cytoplasm and centrally placed small, vesicular nucleus. Stromal component comprised of few benign spindle cells with small blood vessels [Table/Fig-1a,b]. On histological examination, the diagnosis of GCT was made, but since there was history of surgery at the same site, the possibility of granular cell reaction could not be entirely excluded. So, Periodic Acid-Schiff (PAS) stain and S100 immunostaining were done. The cells were found to be positive for both PAS and S100 [Table/Fig-2,3]. On the basis of PAS stain and immunohistochemistry for S100, the histopathological diagnosis of GCT was confirmed. The swelling was already excised completely and sent for histopathological examination. No further surgical intervention was done. On follow-up for one year there was no history of reoccurrence till date. GCT of nasal and paranasal area is extremely rare and only six such cases have been reported to the best of our knowledge.



[Table/Fig-1a,b]: Showing sheets of round to polygonal shaped large cells with ill-defined cell border, finely granular eosinophilic cytoplasm and centrally placed small nuclei with vesicular chromatin (H&E, 40x).



[Table/Fig-2]: Showing cytoplasmic granular PAS positivity (PAS Stain, 40x).
[Table/Fig-3]: Showing cytoplasmic granular S100 positivity (IHC, 40x). (Images from left to right)

DISCUSSION

Granular Cell Tumour (GCT), mostly benign, is a rare lesion of uncertain aetiology [1]. First time, Abrikossoff in 1926, described GCT to be derived from smooth muscle and gave the term myoblastomas, also known as Abrikossoff's tumours [2]. In 1935 Feyrter, proposed neural origin and termed them granular cell neuromas. Fust and Custer in 1948, confirmed the neuronal origin theory and gave a new term granular cell neurofibroma [3]. In 1962 Fisher and Wechsler, found that these tumours most likely originate from Schwann's cells on the basis of ultrastructural and immunohistochemistry finding, and proposed the term granular cell schwannomas. However, World Health Organisation (WHO) gave the nomenclature GCT [4]. Nasal mass accounts for a wide spectrum of differential diagnosis ranging from infective aetiology, inflammatory proliferation to neoplastic lesions

Study	Patient age (in years)	Sex	Location	Size	Presentation	Treatment
Salman RA et al., [14] (1989)	22	Male	Maxillary sinus	-	Facial swelling	Partial maxillectomy
Hwang JSG et al., [6] (2001)	6	Female	Nasal septum	0.6 cm	Nasal discharge	Excisional biopsy
Sasaki T et al., [15] (2007)	69	Female	Nasal septum	0.19 cm	Recurrent epistaxis	Excisional biopsy
Yang CF and Chin SY [1] (2012)	24	Male	Maxillary sinus	0.13 cm	Nasal discharge	Multiple sinusectomy
Bhadani PP and Jamal I [5] (2020)	15	Female	Nasal Cavity	5.0 cm	Nasal obstruction with epistaxis	Excision biopsy
Nkouo YCA et al., [16] (2021)	48	Male	Nasal Cavity	-	Nasal obstruction and hyposomia without epistaxis	Surgical excision by transfacial approach
Present case	28	Female	Right nasal vestibule	0.8 cm	Nasal swelling with obstruction without epistaxis	Excision biopsy

[Table/Fig-4]: Brief review of granular cell tumours of nasal cavity [1,5,6,14-16].

[5]. Historically, immunohistochemically and ultrastructurally identified concentric arrangement of granular cells around nerve end, lipoprotein and sphingomyelin in granular cells indicates that granules in granular cells are derived from myelin or from its degradation products [3,4]. GCTs can occur in any age group from 11-month-old infant to as old as 104 years, being more common in third to fifth decades of life [5,6]. GCTs can involve any organ or tissue of the body most frequently involving head and neck region such as tongue, larynx, parotid glands, labial mucosa, orbits and lacrimal sacs; however extremely rare in nasal cavity [5]. Females are affected more common than males [3]. It grows slowly, insidiously infiltrating the adjacent tissues [3,4]. The membrane contained cytoplasmic granulation is the hallmark of GCTs along with other features like, myelin formation, microtubules, increased density area and microvesicles [7-9]. On light microscopy, the cells appear large round to polygonal shaped cells arranged in sheets and nests with centrally placed small bland looking round to oval nucleus and abundant pale to eosinophilic granular cytoplasm. This can be mistaken as sheets or group of histiocytes, so it is often misdiagnosed as inflammatory condition or reactive process [1,3,5]. Abundant granular cytoplasm of GCTs can mimic other neoplastic condition such as granular cell ameloblastoms, rhabdomyoma granular cell variants of other tumours, like leiomyoma, dermatofibrosarcoma and angiosarcoma and congenital epulis, if present, in paediatric age group [10,11]. The pale granules are PAS positive and diastase-resistant. GCTs are positive for S100, CD56 and SOX-10 immunohistochemistry; however, consistent negativity for desmin, cytokeratin, smooth muscle antigen and epithelial membrane antigen helps in differentiating it from other neoplastic lesions [3,7-9,12]. GCTs are usually benign, but increased mitotic activity, nuclear pleomorphism, necrosis, cellularity and specific location over skin and subcutaneous tissues indicates malignant potential [5,13]. GCTs are neither premalignant nor progress into malignancy, but can co-exist with carcinoma. GCT was found to involve maxillary sinus in case report of Salman RA et al., and Yang CF and Chin SY, nasal septa in case report of Hwang JSG et al., and Sasaki T et al., nasal cavity by Bhadani PP and Jamal I, and Nkouo YCA et al., while in the present case report right nasal vestibule was found to be involved [Table/Fig 4] [1,5,6,14-16]. GCT is a benign lesion, so conservative excision to wide local excision with 2-3 cm margin is the treatment of choice. Local recurrence can occur in case of incomplete excision of lesion.

CONCLUSION(S)

Granular cell tumour is a rare benign tumour, while the malignant potential is very rare and distinguished on basis of histomorphological finding; however there is no specific immunohistochemical analysis to distinguish them. Histopathological examination and immunohistochemical analysis plays an important role in appropriate diagnosis and treatment is surgical excision.

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