Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

ENT and Neck and Head Surgery

Granular Cell Tumor of the Hypopharynx: A Rare Case Report

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DOI: 10.36347/sjmcr.2023.v11i04.024 | **Received:** 25.02.2023 | **Accepted:** 03.04.2023 | **Published:** 14.04.2023

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Abstract **Case Report**

***Aims*:** Granular cell tumor is an extremely rare tumor, accounting for 0.5% of all soft tissue tumors. This rare neoplasia have variable age, sex and ethnicity predilection. Positive staining for S-100 protein confirms the diagnosis and no harmonized treatment guideline is established. We report true this manuscript a rare case of benign of granular cell tumor of the hypopharynx. ***Materials and Methods*:** Case report. ***Results*:** We add to the literature arsenal of this entity by reporting this rare case of benign of granular cell tumor of the hypopharynx in A 74 years-old female patient with hypertension, suffered from dysphonia for about 6 months. Examination identified a budding tumor in the left piriform sinus. Ct scan showcased a 1 cm irregular, spontaneously hypodense, slightly enhanced after injection mass, infiltrating the left piriform sinus. Excisional biopsy confirmed the diagnosis and follow-up was uneventful. ***Conclusion*:** Originating from the Schwann cells; Granular cell tumor has nonspecific symptoms and usually displays as small, smooth, submucosal lesion. Variant of this tumor can be classified as either as synchronous or metachronous, or either malignant, begin or atypical.

Keywords: Granular cell tumor, hypopharynx**.**

INTRODUCTION

Granular cell tumor (GCT), also known as Abrikossoff's tumor is an extremely rare tumor, originating from the Schwann cells and accounting for less than 0.5% of all soft tissue tumors [1, 2]. despite the fact that this neoplasia is found in the head and neck localization in up to 50% of patient, the hypopharynx localization is extraordinary rare with a meager cases report in the literature [2].

No pathognomonic symptoms are associated with GCT, though the majority tends to present as solitary lesion; but in 5 to 6% multiple tumors is documented [2]. There are two distinct entities; one malignant and the other benign; determined by the pathologic examination [1, 2].

We report a scarce case of granular cell tumor of the hypopharynx, adding to the rare count of this entity in this localization, in a 74 years-old female patient.

CASE REPORT

A 74 years-old female patient with hypertension, suffered from dysphonia for about 6 months without any dyspnea, or odynophagia nor dysphagia. Physical exam was within the norms; with a normal oral and cervical examination and no palpable cervical lymph nodes. Nasofibroscopy identified a budding tumor in the left piriform sinus with thickening of the aryepiglottic fold, the other floors of the larynx; including the glottis one; were normal figure (1). Ct scan showcased a 1 cm irregular, spontaneously hypodense, slightly enhanced after injection mass, infiltrating the left piriform sinus and aryepiglottic folds and the corresponding para-laryngeal space figure (2). Direct rigid laryngoscopy was performed, revealing a pedunculated smooth lesion at the level of the anterior angle of the left piriform sinus; the bottom was free. No other synchronous or metachronous tumors were found with a normal larynx and esophagus figure (3). Body CT and ultra sound in search of metastasis were negative. Excisional biopsy was performed, revealing a morphological aspect of round cell tumor proliferation, with histiocytic appearance, with a positive immunostaining of anti-PS 100 confirming a benign GCT.

The immediate follow-up was uneventful and regular follow-ups up to 1 year mark were also without any incident.

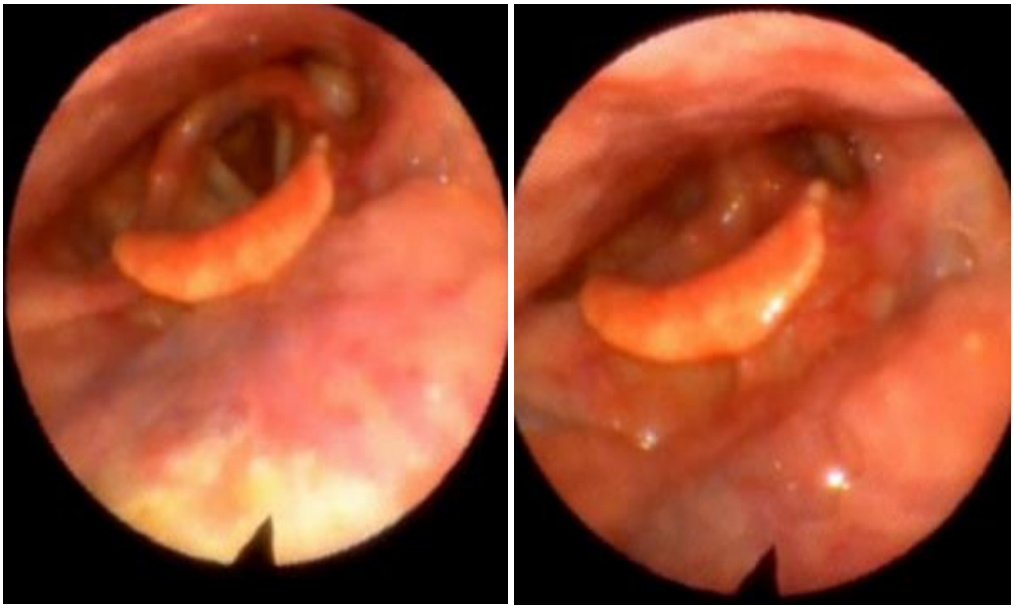

Figure 1: Endoscopic image of budding tumor in the left piriform sinus with thickening of the aryepiglottic fold with closed and open vocal chord

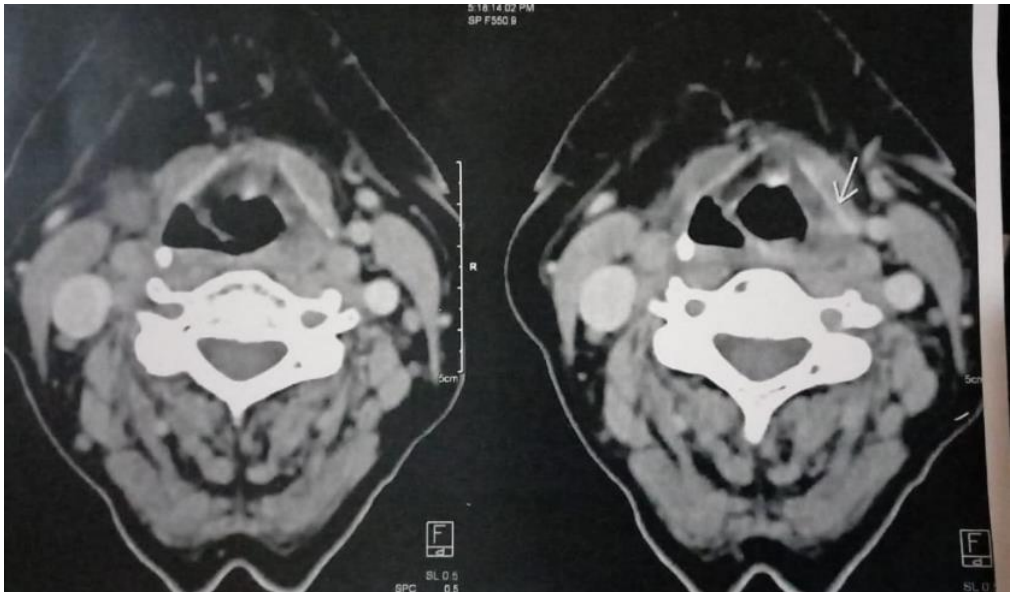

Figure 2: Axial CT scan images showcasing a irregular, spontaneously hypodense, mass, infiltrating the left piriform sinus and aryepiglottic folds and the corresponding para-laryngeal space

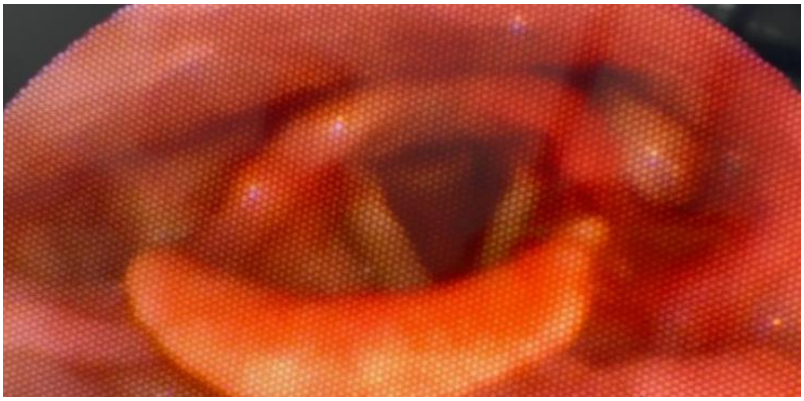

Figure 3: Direct rigid laryngoscopy images revealing a pedunculated smooth lesion at the level of the anterior angle of the left piriform sinus

DISCUSSION

First described by Abrikossoff as myoblastenmyome tumor [3]; GCT said to be arising from Schwann cells, though this neoplasia can be found at any anatomic site, the most common ones are the head and neck region; specifically in the 2/3 of the oral tongue [2, 3], followed by skin and subcutaneous tissues, breast, respiratory and gastrointestinal tracts [1, 4].

What account for 0.5% of all soft tissue tumors, have variable age, sex and ethnicity predilection; depending on the localization; the larynx locating have a predilection for male and the same distribution between caucasians and blacks [5, 6]; the few hypopharynx cases reported however, have a preponderance for females and black ethnicity and wide age range from 29 to 61 years old [1, 2, 7, 8].

Nonspecific symptoms have been reported in relation with hypopharynx sites of GCT, such as globus pharyngeus, dysphagia, associated or not with lenient dyspnea and hoarseness or even dysphoniabut asymptomatic tumor have also been documented [1, 2, 9, 10].

GCT usually displays as small, smooth, submucosal lesion, unencapsulated, but also polypoid or sessile and pedunculated [1]. The submucosal appearebce explains the differential diagnosis such as angiomyoma, synovial sarcoma and salivary gland tumors [1, 2].

A complete ENT and panendoscopy examination is mandatory; as the tumor may present as numourous or solitary lesion, the first variant can be either as synchronous or metachronous lesions [1, 2, 7, 8]. Speaking of variant; GCT can be either malignant, begin or atypical determined by Fanburg-Smith criteria that includes: tumor cell spindling, necrosis, increased rate of mitosis with more than 2 mitosis per 10 hpf, vesicular nuclei with large nucleoli, pleomorphism and a high nuclear to cystoplasmic ratio. Two criteria define atypical GCT (aGCT), three or more indicate mGCT and none of the criteria or focal pleomorphism describe the 98% of GCTs; the benign ones [1, 2, 11]. To note that; the first group have metastatic tendencies in 50% of the cases, in regional lymph nodes, bone or lung [11].

Microscopically GCTs are polymorphic with a large acidophilic cytoplasm. Positive staining for S-100 protein and ubiquitin carboxyl-terminal hydrolase confirms neuroectodermal origin [1, 2, 8].

GCT is shown in CT imaging as a solid mass with homogenous enhancement on contrast and portrayed as hypo intense homogenous tumor on contrast enhanced T1 sequence MRI [1, 2].

The standard and recommended treatment is wide surgical resection with preferential for the endoscopic approach as Piazza *et al.*, [1] report a successful excision of hypopharynx GCT in two cases by endoscopic co2 laser [1, 2]. Lack *et al.*, [3] also delineated 8% recurrence rate following the treatment of choice as the surgical margin were invaded, though numeours authors reported incomplete resection of the tumor without any reccurence in the long follow- up [1, 4].

CONCLUSION

The hypopharynx localization of GCTs is extremely rare with a predilection in the female variant. Asymptomatic or nonspecific symptoms are both reported regarding this entity; however the diagnosis is well established by anatomopathological and immunohistochemical examinations. The treatment of choice remains surgical with a large preference to the endoscopic route and follow-up surveillance visits are a must especially facing the malignant variant of GCT.

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