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Granular Cell Tumor of the Hypopharynx Treated by Cervical Surgery: A Case Report

Ait Taleb Oum'Hand Hajar[1*], Boumendil Ikram[1], Allouch Ihsane[1], Nitassi Sophia[1], Bencheikh Razika[1], Benbouzid M A[1], Oujilal Abdelilah[1], Essakalli Leila[1]

[1]ENT Department, Hospital of Speciality of Rabat, University Hospital Avicenne, Rabat, Morocco

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***Corresponding author:** Ait Taleb Oum'Hand Hajar

Abstract **Case Report**

This clinical case describes an unusual case of an uncommon benign neoplasm named Abrikossoff's tumour or granular cell tumour (GCT) that generally occurs in the fourth and the sixth decades of life. Its location at the hypopharyngeal area is extremely unusual. Here we describe a rare case of 58-year-old female who presented with history of dysphagia followed by dysphonia then a dyspnea that had been gradually worsening.

Keywords: Granular cell tumour, Abrikossoff's tumour, hypopharyngeal location.

INTRODUCTION

Granular cell tumor (GCT) or tumor of Abrikossoff is a rare entity first described by Abrikosssoff in 1926 [1, 2]. It is a rare lesion probably arising from Schwann cells [3], it is more common between the fourth and the sixth decades of life.

Diagnosis requires histopathological examination and immunohistochemistry. Here, we report a case of GCT involving the hypopharynx treated by a successful surgery.

CASE REPORT

A 58-year-old female patient, with no medical or surgical history, who presented with a 08-month history of dysphagia followed by dysphonia then a dyspnea that had been gradually worsening for 04 months. The patient was admitted to our department due to inspiratory dyspnea with stridor, supraclavicular retractions which led us to request an emergency CT scan (Figure-1). The computerized tomography (CT) scan was indicative of a relatively well-defined homogeneous oval mass with regular margins that goes down from C3 to D2 and buds in the laryngeal vestibule(red star). The mass was ~50 × 42 × 79mm. isodense to the muscle, infiltrating the left piriform sinus(red arrow) buds in the laryngeal vestibule, encompasses cricoid and arytenoid cartilages without obvious lysis and ruptures the posterior wall of the first two tracheal rings with stenosis of the tracheal lumen(blue arrow). There was no associated cervical lymphadenomegaly.

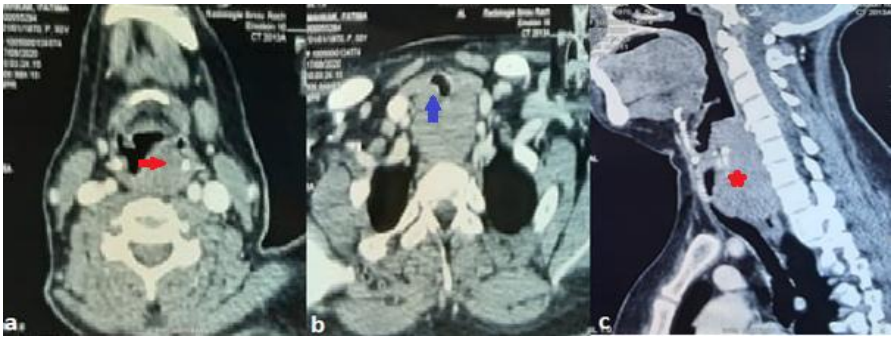


Fig-1: Contrast enhanced axial (a,b) and Sagittal reconstruction (c) showing the mass involving overflowing into the laryngeal lumen and reaching the trachea with stenosis of its lumen(blue arrow)

We performed an emergency tracheotomy then an examination by laryngeal nasofibroscopy which highlighted a submucosal process, filling the entire retro-cricoid space, taking the left piriform sinus and budding in the glottic floor preventing its visualization especially on the left side without reaching the epiglottis.

Suspension laryngoscopy was performed confirming the submucosal aspect of the process which takes the esophageal lumen, the retro-crico-arytenoid space as well as the left piriform sinus with a bulging at the level of the glottic floor which remains difficult to assess. Several biopsies were performed in order to be able to determine the histological type of this mass.

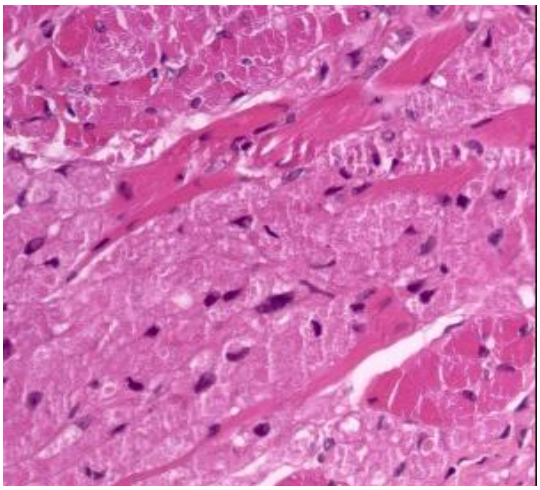


Fig-2 : Proliferation in large cell sheets with eosinophilic cytoplasm, sometimes round nuclei sometimes elongated hyper-chromatic and anisokaryotic nucleus with a crushed character (HE ×400)

Histopathological examination showed a tumor proliferation made up of layers and spans. Tumor cells have an eosinophilic cytoplasm with sometimes round nuclei sometimes elongated hyper-chromatic and anisokaryotic nucleus with a crushed character. These elements were developed in an abundant desmoplastic collagen fibrous stroma. Immunohistochemical study was done. The immunohistochemistry showed positive antiCD56, positive antiPS100, positive anti NSE, positive anti CD68 and positive anti Vimentin. Ki-67 Proliferation Index was estimated at 10%. On the basis of these findings, a final diagnosis of granular cell tumor "Abrikossoff tumor**"** was established.

T1, T2 and T1 FAT SAT weighted axial and coronal sequences with and without contrast MRI was done to ascertain the anatomical extent of the tumor and to study the possibility of a surgical treatment.

The RMI showed a roughly oval tumor process of 54× 45×88 mm, well limited in intermediate T1 signal, hypo-intense in T2, heterogeneous and containing hypersignal areas in T2 (Figure-2).

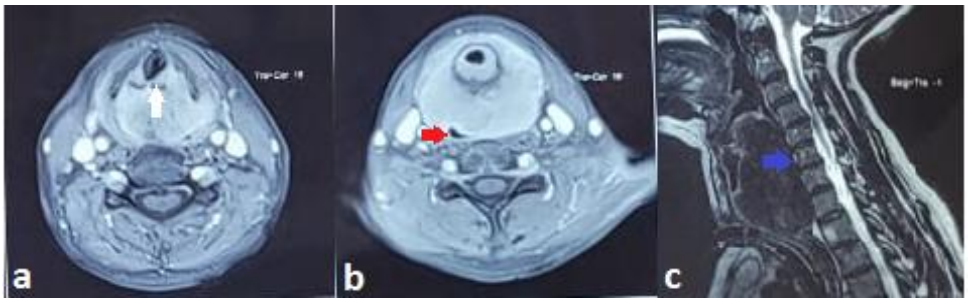


Fig-3: Post contrast T1 axial (a,b) and T2 Sagittal reconstruction (c) showing a large tumor that infiltrates glottic floor(a), pushes the esophagus to the right (red arrow) and extends posteriorly to infiltrates para-vertebral muscles(blue arrow)

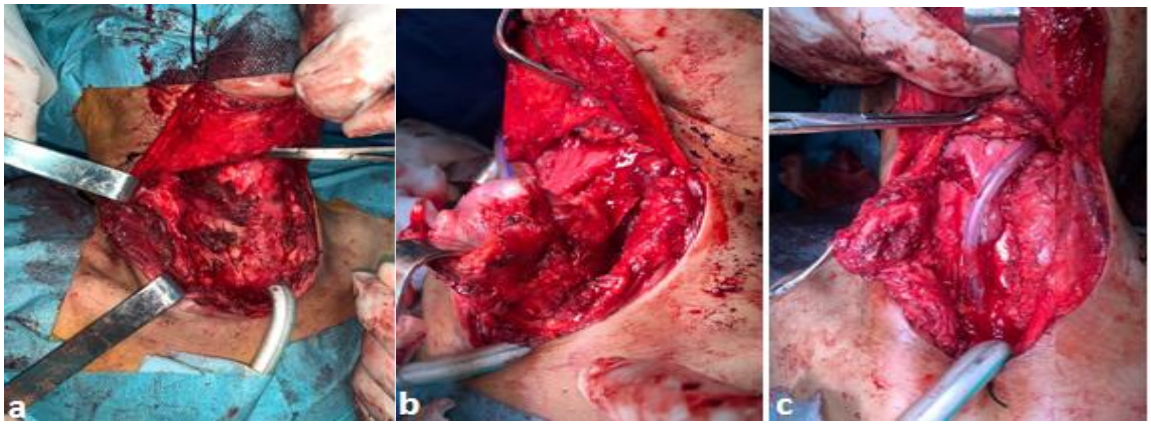
This process buds into the laryngeal lumen and narrows it and infiltrates the para-laryngeal spaces (Fig-2a white arrow). It pushes back the esophagus to the right and obstructs its lumen (Fig-2b red arrow).

The mass infiltrates the longus colli muscles with intimate contact with the anterior surface of the vertebral bodies from C4 to D2 without any obvious bone lysis (blue arrow).

CT scans of the thorax and abdomen did not reveal any involvement of other sites. A multidisciplinary consultation meeting was carried out directly after the histopathological results and MRI. It was decided to operate on the patient given the high chances of resectability: benign histological type of the tumor, the permeability of the vascular axes and the non-lysed aspect of the vertebral bodies.

A feeding jejunostomy was put in place to compensate for the patient's total dysphagia and to guarantee nutritional and above all protein intake in order to ensure better postoperative recovery.

Under general anesthesia an open neck procedure was performed starting with U type incision straddling the tracheostomy, followed by a detachment of the musculocutaneous flap (Fig-4(a)). Supra and infra hyoid muscles were released and a total pharyngo-laryngectomy was performed removing the entire tumor mass (Fig-4(e)) with a resection of the larynx while preserving the maximum of pharyngeal mucosa (Fig-4(c)). Indeed dissection in the posterior plane was more difficult because of firm adherence to the prevertebral muscles (Fig-4(b)) .Thyroid lobes were gently separated from the trachea, cricoid cartilage and inferior constrictor and the closure of pharyngeal mucosa was done by simple and separates stitches on a nasogastric tube (Fig-4(d)). It was judged that pharyngeal reconstruction was unnecessary.



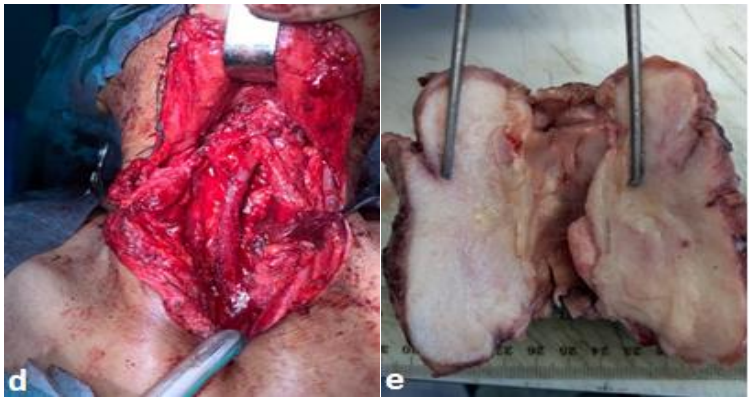


Fig-4: (a) Lifting of the skin flap including the superficial cervical fascia; (b) Release of the mucous membrane of the piriform sinus (including the mass) from the cartilage of the thyroid wing (c) complete excision of the tumor after the dissection in the avascular plane between the esophagus and the trachea (d) Closure of pharyngeal mucosa by simple and separates stitches on a nasogastric tube (e) surgical specimen after excision.

Postoperatively, the patient remained for 10 days under PPI(proton pump inhibitor) and exclusive feeding by jejunostomy, before the ablation of the nasogastric tube, a test with methylene blue was carried out which did not show the presence of pharyngostoma. A barium swallow test was also done and it didn't show any anomaly and six months follow up was uneventful.

DISCUSSION

Granular cell tumor (GCT), also called Abrikossoff's tumor is a rare tumor (0.019-0.03% of all tumors) and greater common in black patients [1].

It is often benign and it occurs at any age with a peak in frequency between fourth and sixth decades [2, 3]. The sex-ratio male/ female is ½ [4, 5].

GCT from Schwann cells is supported by the presence of myelinated nerve bundles in more than 50% of cases [6].

GCT sits mainly in the mucous skin, it is predominantly found in the head and neck region (50% of all cases), lingual localization is the most common followed by floor of the mouth and palate [7].

It is a small tumor, usually nodular, smooth, granular or warty, polypoid or sessile, variable in color (pink or yellowish) [8, 9]. Tumor growth is slow and often asymptomatic [8]. Symptoms reported for hypopharyngeal GCT included dysphagia, dysphagia associated with mild dysphonia and dyspnea, and a foreign body sensation in the throat [10, 11]. In terms of imaging, MRI is the best complementary examination because it directs a possible deep biopsy and does the extension assessment, especially in malignant forms [12]. In the literature GCTs are classically iso-intense in T1 and hypointense in T2 [13], which is consistent with our experience.

Diagnosis requires histopathological examination and immunohistochemistry. Microscopically, it is characterized by a proliferation of cells of large, polygonal or spindle-shaped, grouped into syncytial-looking cluster, separated by a hail stroma with an abundant cytoplasm loaded with fine eosinophilic granulations. The nuclei are small, round or oval, often central, hyperchromatic and regular, without atypia or mitosis [14, 15]. The tumor is covered with an epithelium normal or verrucous hyperplastic especially in mucosal locations [16, 17]. It often infiltrates the underlying striae muscle. Tumor cells are almost always positive for neurogenic markers: S100 protein (100% of cases) and NSE (90% of cases). Vimentin immunostaining is positive in 70% of cases, but in a variable way [14].

The recommended treatment in GCT is wide surgical excision with a favorable evolution in case of complete resection [8]. Due to the rare occurrence of GCT, optimal therapy is challenging particularly when the tumor is localized in the hypopharyngeal area. In our case the tumor was very extensive with an infiltration of the larynx, para-laryngeal spaces and longus colli muscles with intimate contact with the anterior surface of the vertebral bodies which required a total laryngectomy removing the entire tumor with a definitive tracheostomy.

It seems that when the tumor involving surgical margin recurrences were noted in 8% to 12% of GCT [16]. However the effectiveness of radiotherapy and chemotherapy is not proven [17].

CONCLUSION

The GCTs are unifocal and benign tumors in the majority of cases, hypopharyngeal location is an extremely rare entity. Usually the lesions are less than 2cm which allows an endoscopic resection unlike our clinical case where surgery was essential.

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