



## Case report

# Large tracheoesophageal granular cell tumor: An atypical case presentation, location, and management

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## ABSTRACT

**Introduction:** Granular cell tumors (GCT) are rare tumors that most frequently present in the oral cavity. While some present within the gastrointestinal tract, a GCT near the trachea is an extremely rare occurrence.

**Presentation of case:** A 42-year-old man presented to the Emergency Department after a motor vehicle accident. A computerized tomography (CT) scan revealed an incidental soft tissue 3.2 × 5.5 cm mass anterior to the esophagus and posterior to the trachea with no adjacent lymphadenopathy. The patient denied dyspnea, voice changes, or dysphagia. Due to its size and location, the patient underwent a transcervical excision of the retrotracheal tumor. Tumor cells were positive for CD68, CD163, S100, and SOX10, confirming a GCT.

**Conclusion:** This is a distinctive presentation of a large (5 cm) GCT in the plane between the trachea and esophagus. GCTs are not often on the differential diagnosis of masses that present in this region.

## 1. Introduction

Granular cell tumors (GCT) are primarily benign soft tissue tumors [1]. Originally called myoblastomas by Abrikossoff in 1926 due to suspected myogenic origin, GCTs have become widely accepted to show Schwann cell differentiation [2]. Although relatively rare with an incidence of 0.03% in one review of 410,000 surgical specimens [3], these tumors most frequently present in the head and neck, specifically the tongue and other subsites in the oral cavity. However, 5–11% of GCTs present within the gastrointestinal tract [4]. Here, GCTs have incidence rates of 65% for the esophagus, 20% for the colon, and 9% for the stomach [3]. Even rarer, GCTs may present as primary tracheal masses, with around 44 cases to date [2].

GCTs are characterized by their morphology (large cells with abundant pale granular eosinophilic cytoplasm) and expression of Schwann cell markers (S100 and SOX10) as well as histiocyte markers by immunohistochemistry [5,6]. Treatment varies from observation to surgical excision [2]. Excision is usually recommended for diagnosis and management of large tumors [7]. We report an unusual case of a GCT presenting as a large tracheoesophageal mass in an asymptomatic and otherwise healthy individual. Given that the majority of GCT cases are in

the tongue and other subsites in the oral cavity, the presentation of a GCT near the trachea is an extremely rare occurrence [2]. Most esophageal and tracheal GCTs are small submucosal lesions detected incidentally on bronchoscopy or esophagogastroduodenoscopy, allowing biopsy and removal endoscopically [7]. The large size and submucosal location of the tumor in this patient required a transcervical surgical approach for removal which is exceedingly unusual. This work has been reported in line with the SCARE 2020 criteria [8].

## 2. Presentation of case

A 42-year-old man presented to the Emergency Department after a motor vehicle accident with chest and extremity pain. A computerized tomography (CT) scan did not show injuries related to the motor vehicle accident but did reveal a soft tissue 3.2 × 5.5 cm mass anterior to the upper esophagus and posterior to the trachea with no adjacent lymphadenopathy (Fig. 1A-B). This mass was centered slightly more on the left than the right, between the posterior tracheal wall and the anterior esophagus, inferior to the level of the cricoid, and extending into the chest. Despite narrowing of the airway by the mass, the airway was patent and the patient denied shortness of breath, voice changes,

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dysphagia, or difficulty sleeping, despite a considerable impact of the mass on his airway. The patient was a former smoker for 20 pack-years with the rest of the medical history being insignificant. Magnetic resonance imaging (MRI) scan demonstrated a mass effect on the left thyroid lobe, with the thyroid and esophagus separate from the  $3.1 \times 5.5 \times 4.7$  cm tumor (Fig. 1D-E). The patient then underwent an ultrasound-guided fine-needle aspiration which showed rare clusters of cytologically bland epithelioid cells with granular cytoplasm that were positive for SOX10, S100, and PAS-D, while negative for AE1/3 and CD68. These findings were consistent with a GCT.

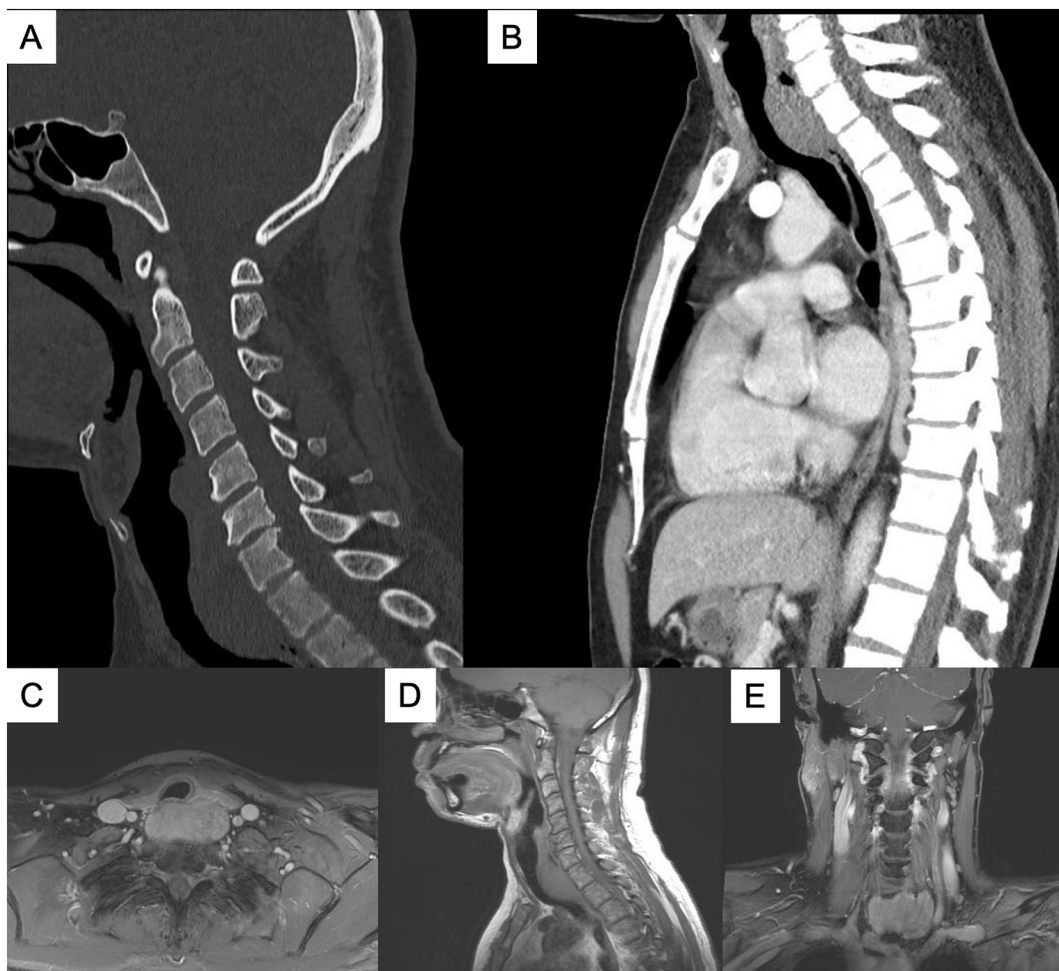
The patient underwent a transcervical excision of the retrotracheal tumor under general anesthesia. A nasogastric tube was placed into the esophagus intraoperatively to assist with identification. Recurrent laryngeal nerve monitoring was used intraoperatively due to the proximity of the mass to the left recurrent laryngeal nerve. The mass was firm and well circumscribed, palpated inferior to the cricoid ring, medial to the carotid artery and posterior to the trachea. Removal required lateral retraction of the omohyoid muscle and rotation of the larynx to the right. During the operation, there was evidence that the mass may have been present for a long time, as it was large in size and intimately involving the posterior tracheal wall and esophageal musculature. There was no violation of the tracheal or esophageal mucosa during the operation and the patient's recovery was uncomplicated. On immediate and 6-month post-operative follow up, the patient had no pain, dysphagia, or voice

changes.

Gross examination of the tumor revealed a  $5.5 \times 4.4 \times 2.8$  cm mass surrounded by a thin layer of connective tissue (Fig. 2A). The cut surface was homogenous and pale yellow-tan without gross necrosis (Fig. 2B). On histologic examination, the tumor was composed of nests of large pale cells with intervening collagenous bands (Fig. 3A). Cells contained abundant granular pale eosinophilic cytoplasm and small ovoid nuclei with occasional wrinkled contours (Fig. 3B). There was no evidence of nuclear pleomorphism, necrosis, or significant mitoses. Tumor cells were variably positive for CD68 (Fig. 3C) and CD163 (Fig. 3D) and strongly positive for S100 (Fig. 3E) and SOX10 (Fig. 3F), confirming granular cell tumor.

### 3. Discussion

GCTs are unusual tumors that most frequently present in head and neck subsites, and rarely within the trachea or esophagus. This case is exceedingly rare in that the GCT presented between the posterior trachea and anterior esophagus with a considerable size (greater than 5 cm). While the mass' intimate involvement with the posterior tracheal wall suggests a tracheal origin, we were unable to definitively determine if this mass was originally of tracheal or esophageal origin as the mucosa of both the esophagus and trachea was completely normal endoscopically.



**Fig. 1.** (A-B) Routine non-contrast sagittal CT images showing a cervical spine with no visualized fluid or mass above the C5 level, a  $3.2 \times 5.5$  cm tumor anterior to the upper esophagus with no adjacent lymphadenopathy, and a mild narrowing of the airway. (C) Axial, (D) sagittal, and (E) coronal MRI with contrast showing a  $3.1 \times 5.4 \times 4.7$  cm tumor in the superior mediastinum/thoracic inlet notable for smooth internal enhancement. (C-E) Also seen is a mass effect on the left thyroid lobe, with the thyroid and esophagus separate from this mass as well as a patent nasopharynx, oropharynx, hypopharynx, and larynx and normal appearing parotid, submandibular, and thyroid glands.



**Fig. 2.** (A) Gross examination revealed a 5.5 cm mass surrounded by a thin layer of connective tissue. (B) The cut surface was homogenous and pale yellow-tan without gross necrosis. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Masses in the tracheoesophageal region have an extensive differential diagnosis. In adults, 90% of tracheal tumors are malignant with squamous cell carcinoma and adenoid cystic carcinoma as the first and second most common respectively [9]. Endobronchial carcinoid tumors, which are the most common primary tracheobronchial tumor in children and adolescents [10], and mucoepidermoid carcinomas, which represent only 0.1–0.2% of thoracic malignancies, are on the differential of tumors in this region. More rare diagnoses include tracheal lipomas, tracheobronchopathia osteochondroplastica, and tracheal leiomyomas [9]. Other benign processes like amyloidosis, hamartomas, and respiratory papillomatosis are also included in the differential diagnosis [10]. We present this case of a tracheoesophageal GCT as evidence that fine needle aspiration of masses in this region can establish a diagnosis and guide appropriate treatment.

GCTs are found throughout the body, but most frequently within the tongue (30%), skin and subcutaneous tissues (30%), breast (15%), and respiratory tract (10%) [11]. The tumors can present multifocally although the majority are benign solitary lesions with malignant variants being extremely rare [11]. GCTs are slow growing, non-tender, generally painless, and usually small lesions averaging 2.2 cm (range 0.45–6 cm) [12,13]. While there are several hypotheses regarding cytologic origin, the most supported idea is that the lesion is a reaction to altered cellular metabolism in the Schwann cell or its progenitor. The

distinctive feature of abundant granular cytoplasm and the presence of S-100 protein also support this idea [11].

GCTs are found in gastrointestinal tract structures 5–11% of the time, with 65% of these residing in or around the esophagus [4]. Overall, around 400 cases of esophageal GCTs have been reported in the literature, roughly comprising 1.7% of all GCTs [7,14]. Most patients with esophageal GCTs are asymptomatic and are most often found incidentally on esophagogastroduodenoscopy. Interestingly, a recent review of 11 esophageal GCT cases found that patients with masses less than 2 cm are more likely to be asymptomatic [15], but this patient did not show symptoms of gastrointestinal reflux disease or dysphagia even with the mass measuring greater than 5 cm and causing compression of both the trachea and esophagus. Treatment for esophageal GCTs vary depending on the size of the mass, but small masses tend to undergo observation [14,16], while larger masses are usually subjected to endoscopic removal.

Since the first reported case of GCT, there have been a limited number of GCTs presenting in the tracheal region of the head and neck with only around 44 cases to date [17,18]. Laryngotracheal GCTs are also quite rare, and often appear as whitish or gray mucosa that resemble vocal cord polyps or granulomas involved in the posterior third of the true vocal cords [19]. Tracheal GCTs have averaged 2.3 cm in size with a clinical presentation of persistent cough, hemoptysis, wheezing, obstructive sleep apnea, and progressive dyspnea [12]. Most patients are asymptomatic, however, and only show signs and symptoms with tumors greater than 2 cm [20]. Tracheal GCTs are commonly found incidentally through CT scans or bronchoscopy, as was the case with this patient. Treatment is similar to esophageal GCTs, with observation recommended for small tumors and endoscopic laser excision for larger tumors.

The most effective management of GCTs is conservative excision [21]. In the case of a malignant GCT, regional lymph node dissection and persistent follow-up are recommended. This particular GCT was not malignant but if it were malignant, one could consider performing a central neck dissection. The effectiveness of chemotherapy for the treatment of these masses has been unproven, and therefore is not a mainstay treatment option [21,22]. Adjuvant radiotherapy may be considered for rare recurrent lesions [21].

#### 4. Conclusion

The case presented is distinctive from others in the literature in that this patient's GCT was located in the plane between the trachea and esophagus and greater than 5 cm. Typical presentation is submucosal and visible endoscopically [12]. In this patient, endoscopic visualization was difficult. Despite the significant size of the mass, the patient did not experience classic symptoms of a tracheal tumor such as shortness of breath, or of an esophageal tumor such as dysphagia or gastrointestinal reflux. The patient also required a transcervical approach for removal due to its size and unusual location, which differs from many approaches for the removal of GCTs cited in the literature [15]. In conclusion, GCTs are Schwann cell derived, rare tumors that are not often considered on the differential diagnosis of tracheoesophageal masses.

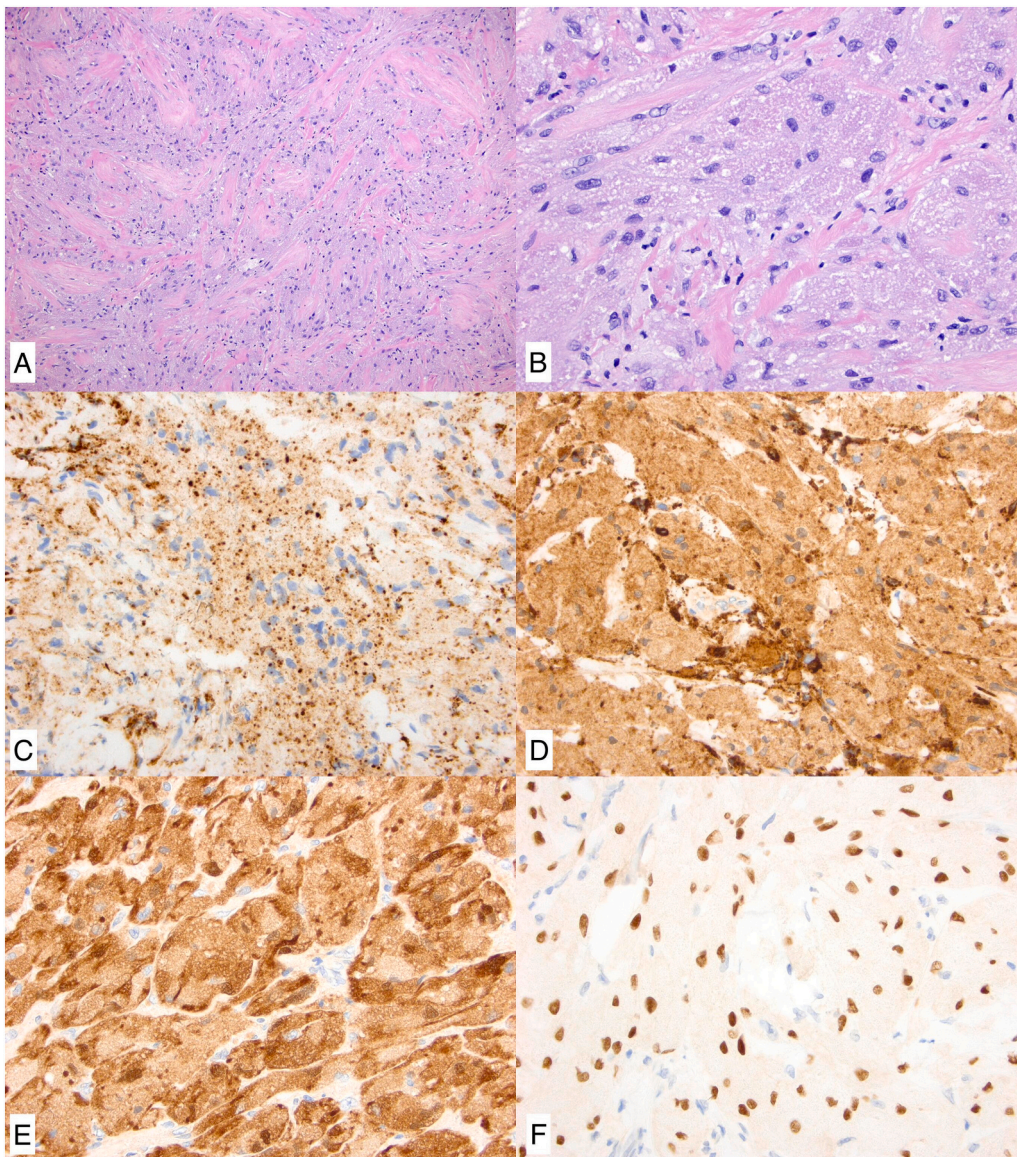
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#### Ethical approval

None to report.





**Fig. 3.** (A) The tumor was composed of nests of large pale cells with intervening collagenous bands. (B) Cells contained abundant granular pale eosinophilic cytoplasm and small ovoid nuclei with occasional wrinkled contours. Tumor cells were variably positive for (C) CD68 and (D) CD163 and strongly positive for (E) S100 and (F) SOX10.

### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Author contributions

Conception & design: All authors.  
 Data Acquisition: All authors.  
 Analysis and interpretation: All authors.  
 Drafting the manuscript: All authors.  
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### Research registration

None to report.

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### Declaration of competing interest

No conflicts of interest.

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