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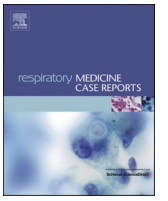

Case report

Granular cell tumor in the pulmonary tree

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A B S T R A C T

Granular cell tumors are often benign growths of the tongue, dermis, and subcutaneous tissues. Rarely, we see production in the pulmonary system. These schwannian and histiocyte origin tumors in the lungs are called pulmonary granulocyte tumors. While granular cell tumors are rare, pulmonary granulocyte tumors are even rarer, with less than eighty-five cases described since the 1930s. Herein, we introduce these rare growths and provide a review of known epidemiological evidence and pathophysiology.

1. Case presentation

A 43-year-old female with history of sickle cell trait and anemia presented with weakness and profound anemia. She has a 30-pack-year smoking history. Past medical history was non-contributory. Physical exam was unremarkable except for tachycardia. Admission labs showed a hemoglobin of 1.5. Chest x-ray (CXR) showed a right apical/upper lung field opacification. CT chest demonstrated a cavitary mass within the apical segment of the right upper lobe (RUL). Bronchoscopy yielded a 95% obstructing mass of the RUL bronchus suspicious for malignancy. Brushings, lavage, and biopsies were performed; biopsies were consistent with benign pulmonary granular cell tumors. She underwent a robotic RUL bronchial sleeve lobectomy with lymph node dissection. Postoperative pathology confirmed the diagnosis and lymph nodes were negative for malignancy.

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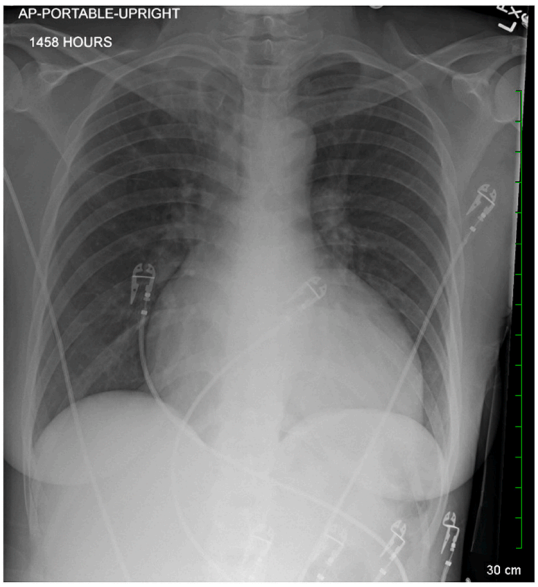
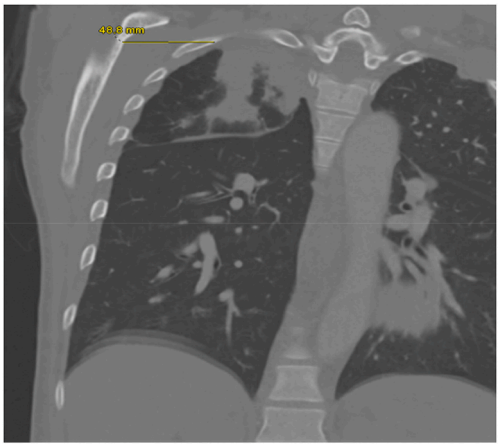
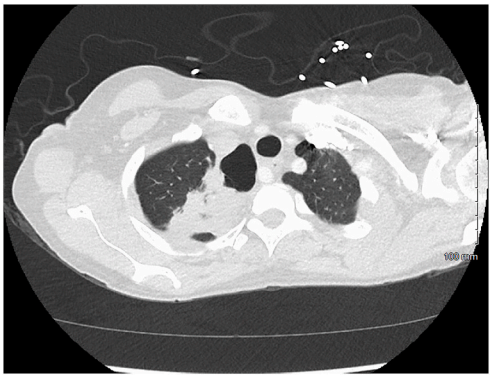
2. Imaging

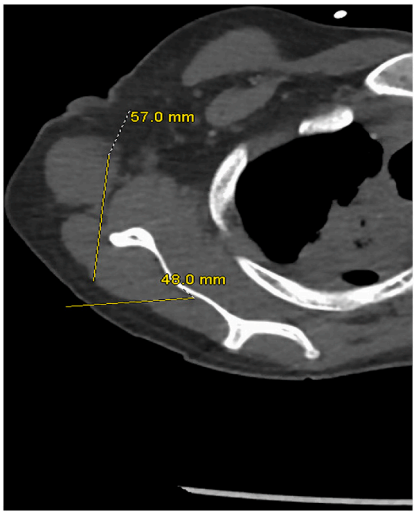
2.1. Chest X-Ray (CXR)



2.2. Chest CT (CT PE)







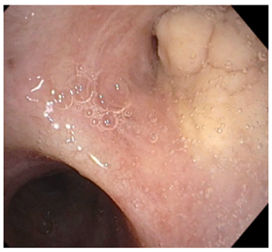
2.3. Pulmonary function tests

PFTs showed mild obstructive lung disease, moderate restrictive pattern, and normal DLCO.

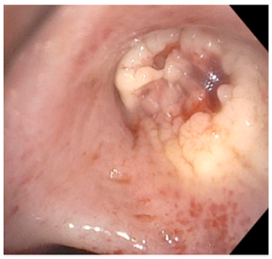
2.4. PET scan

PET CT showed low-grade FDG uptake with non-hypermetabolic right paratracheal lymph nodes.

2.5. Bronchoscopy images



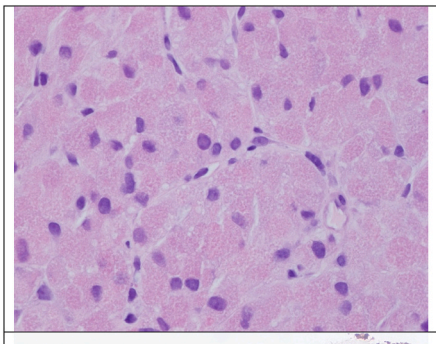
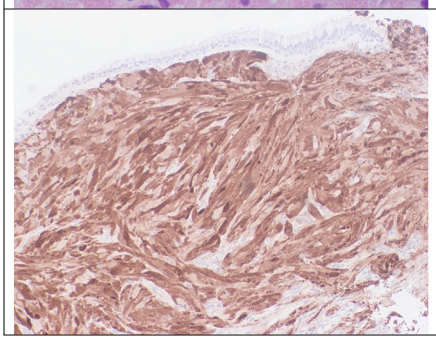
RUL endobronchial mass.



RUL mass, post-biopsy.

2.6. Pathology, histology

Slides with associated descriptions.

	Sheets of polygonal tumor cells with abundant eosinophilic granular cytoplasm and a bland round/oval nucleus are seen in this pulmonary granular cell tumor (H&E, 400X)
	- The cells of the pulmonary granular cell tumor show positive staining with the immunohistochemical stain for S-100 (IHC, 100X)

3. Discussion

Granular cell tumors (GCT) are often benign growths of the tongue, dermis, and subcutaneous tissues, rarely seen in the pulmonary system [1]. When found in the lower respiratory tract, GCTs are referred to as pulmonary GCTs (pGCTs). These rare tumors are most often benign and incidentally noted on radiographic imaging.

Described initially by Abrikossoff in a patient with a tongue lesion in 1926, these schwannian and histiocyte origin tumors are often benign [2,3]. However, an even rarer subset, of less than two percent, are classified as malignant with high metastatic, recurrence, and mortality rates [4]. Most of these GCTs are found in younger women between twenty to forty years of age [5]. pGCTs are exceedingly rare, in juxtaposition to already rare GCTs, with less than eighty-five cases described since the 1930s [6–8]. Rarely, a studied pGCT has demonstrated malignancy.

pGCTs are often incidentally found during work-up of lower-respiratory pathology. Initial presentation typically includes dyspnea, wheezing, cough, atelectatic changes, obstructive pneumonia, and hemoptysis [9,10]. Histologic presentation varies between benign and malignant pGCTs. Benign pGCTs consist of oval, eosinophilic granular cytoplasm and small nucleoli without mitotic activity, hemorrhage, or necrosis [11,12]. Malignant pGCTs however, consist of prominent nucleoli with apparent necrosis and hemorrhage, with evidence of mitotic activity [6]. Literature review for therapeutic plans suggest patient-centered regimens dependent upon both the patient and tumor background and characteristics.

Currently, there are no clear therapeutic plans for those patients diagnosed with pGCTs. Deavers *et al.* recognized endoscopic removal, laser, and resection as possible treatments, with surgical resection yielding the highest cure rate with uncommon episodes of recurrence [11,13]. Studies outlining risks versus benefits in small, non-obstructive tumors have not been performed to this date. Removal of a pGCT does not suggest a good prognosis, unfortunately. McSwain *et al.* note an approximate 13% association between GCTs and neoplastic processes [14,15], although the specific correlation to pGCTs has not been well studied. In addition, some studies have reported a link between pGCTs and non-neoplastic processes, such as sarcoidosis [11].

The mechanism behind the development of pGCTs will be drawn from further understanding of the underlying genetics. Possibly, these benign pGCTs are on the continuum of genetic mutations in the driver proteins (ASXL1, Notch2, and PARP4). Unfortunately, the true pathophysiologic mechanism will take time to resolve given its absolute rarity and lack of data.

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