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A Rare Case Of Endobronchial Granular Cell tumour: Case Report

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Abstract

Granular cell tumour (GCT) is a rare neoplasm of Schwannian phenotype with hallmark abundant granular cytoplasm which stains positively for S100 immunostain. More than 90% of the cases were benign and slow-growing with a high recurrence rate. This neoplasm was described in almost all sites of the body and uncommonly in the tracheobronchial tree.[1]

We hereby report a rare case of endobronchial benign GCT in a middle-aged male. He presented with cough and dyspnea. No other significant history. Bronchoscopy showed a submucosal endobronchial polyp in the left main bronchus. Histologically, a circumscribed submucosal neoplasm with large, polygonal cells were seen exhibiting abundant eosinophilic granular cytoplasm. This favoured a diagnosis of benign granular cell tumour.

This is one of the few case reports of endobronchial GCTs published. Histomorphology with IHC is diagnostic for GCT. However care should be taken to rule out other common lesions. Complete surgical excision is curative. Although they tend to recur locally, they have a good prognosis with close follow-up and monitoring.

Keywords: Granular cell tumour; Endobronchial tumour; Benign Schwannian tumour

Introduction

Granular cell tumour (GCT) is a mesenchymal neoplasm of neuroectodermal origin, recognized as a distinct entity by Abrikossoff in 1926. The morphologic hallmark of this tumour is abundant acidophilic granular cytoplasm due to lysosomal accumulation. Although most of the cases were benign, malignancy was also reported very rarely and posed a difficulty in diagnosis. It commonly presents in middle age group and has a wide range of distribution. This neoplasm usually arises in skin, soft tissue, head and neck and uncommonly involves airways.[1] When localized to unusual sites, it poses a diagnostic difficulty. Our report describes a case of a 43-year-old male with endobronchial symptomatic benign GCT and reviews clinicopathological features and differential diagnosis of this entity.

Case Report

A 43-year-old male presented with cough, dull chest pain and dyspnea on exertion for the past 20 days. No history of associated fever or hemoptysis. No known co-morbidities/ significant family history or past history was seen. General physical examination, vitals and systemic examination showed no abnormalities. The initial workup was done elsewhere. A CT scan was done which revealed a left-sided endobronchial lesion.

The patient was referred to our hospital for further evaluation. Screening bronchoscopy showed a polypoidal submucosal endobronchial tumour occluding >50% of the lumen of the left main bronchus. Bronchoscopic tumour excision was done

and was sent for HPE. We received multiple pieces of mucosa-covered grey-white soft tissue bits together weighing about 0.5g. The cut surface showed a homogenous tan-white appearance.

Histology showed portions of respiratory mucosa with a circumscribed submucosal neoplasm. The neoplastic cells were large, polygonal and were arranged in sheets. The cells exhibited a small round nucleus, dense chromatin, inconspicuous nucleoli and abundant eosinophilic cytoplasm with granules. Focal squamous metaplasia and pseudoepitheliomatous hyperplasia were noted. Necrosis/ atypia/ lymphovascular invasion are not seen. The granules stained positively for periodic acid Schiff (PAS). A diagnosis of a benign granular cell tumour was reported. Immunohistochemistry (IHC) for S-100 marker was suggested for further evaluation. Photomicrographs of this case are depicted in Figures 1-3.

Discussion

GCT is a rare tumour of Schwannian phenotype origin.[1] The incidence of endobronchial GCTs has shown an uptrend in recent times.[2] Agewise distribution of GCTs showed a peak incidence in the fourth to sixth decade of life, but can also present in younger age groups. A few studies suggest a female preponderance (approximately 50-60%).[3] GCTs can occur in various anatomical sites with a preponderance to the head and neck region (45%-65%). Tracheobronchial localization is very rare and accounts for 6%-10% of GCTs. The bronchus is the second most common site of thoracic GCT (46.9%), following the oesophagus.[4] McSwain *et al* stated that 13% of the GCTs are associated with other neoplasms, commonly small cell and non-small cell lung carcinomas. When pulmonary GCTs are >1cm, associated pulmonary neoplasms should be suspected.[5] Literature reviews across the world documented approximately 2% malignant GCTs (MGCT).[1]

Owing to their indolent nature, the majority of the cases were detected incidentally. The most common symptoms of pulmonary GCTs are cough, breathlessness, chest pain and hemoptysis. Radiology studies may show nodule, signs of pneumonia, bronchiectasis or atelectasis. Bronchoscopic, histomorphological analysis coupled with IHC provides a definitive diagnosis. Tumours less than 8mm were amenable to endobronchial therapeutic procedures. Tumours of greater size require complete surgical resection. Studies state that GCTs are associated with mutations of ASXL1, Notch2, PARP4 and P1K3CA. A targeted therapy with P13K/AKT/mTOR inhibitors can be used for treatment in such cases.[1,6] A higher rate of recurrence was seen in cases treated bronchoscopically. Due to the high recurrence rates of these tumours, an annual follow-up for at least 5 years is recommended.[7]

GCTs can mimic a variety of neoplastic and non-neoplastic lesions. Differential diagnoses of benign GCTs are fibroxanthoma, Rosai- Dorfman disease and differentials for malignant GCTs include malignant peripheral soft tissue sarcoma, alveolar soft tissue sarcoma, malignant fibrous histiocytoma, leiomyosarcoma, PEComa, angiosarcoma, and melanoma. Pseudoepitheliomatous hyperplasia was noted in 50-65% of the cases with GCT.[7] A superficial biopsy in such cases can lead to a confusion of squamous cell carcinoma. Although core biopsy can be diagnostic, care should be taken to rule out other common lesions before finalising a diagnosis of GCT, since various lesions (eg- angiosarcoma, leiomyosarcoma and pleomorphic liposarcoma) can exhibit granular cell change focally. In morphologically doubtful cases immunohistochemistry analyses can be done. The granules stain positively for antibodies against S-100, vimentin, actin or neuron-specific enolase. A minority of the cases could be S-100 negative. In such cases, they stain positively for CD68, NK1C3 and alpha-1 antitrypsin.[1]

Benign GCT has an excellent prognosis. They tend to recur locally, especially when excised incompletely. Malignant GCTs follow an aggressive course with upregulation of p53 and Ki67 proliferation index. Poor prognostic factors are tumour size >8mm, elderly age group, metastasis, Ki-67 index >10% and p53 immunoreactivity.[1]

Conclusion

This is one of the few published case reports of endobronchial GCTs. In summary, GCTs are neuroectodermal tumours, rarely arising in tracheobronchial locations and can present with obstructive symptoms. Most of the GCTs are benign and slow-growing. Since radiological findings cannot

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differentiate this from other malignant neoplasms, they are rarely diagnosed pre-operatively. Histomorphology with IHC is diagnostic. Due to its tendency to recur, a complete excision with follow-up is advised.

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Figure 1: Photomicrograph depicting granular cell tumour arranged in sheets (x10, original magnification).

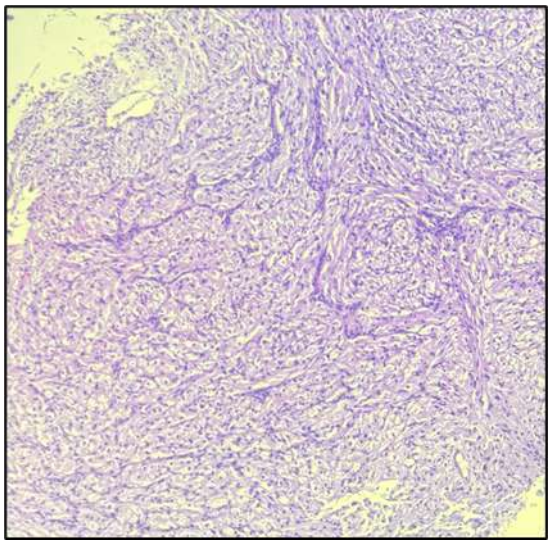

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Figure 2: Histomorphology of granular cell tumour with epithelium showing focal squamous metaplasia (x10, original magnification).

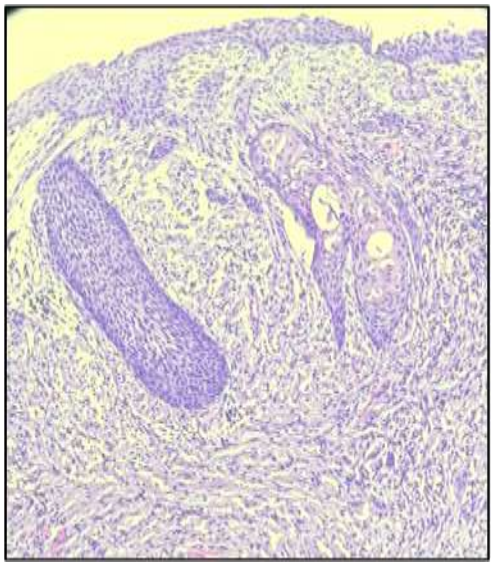
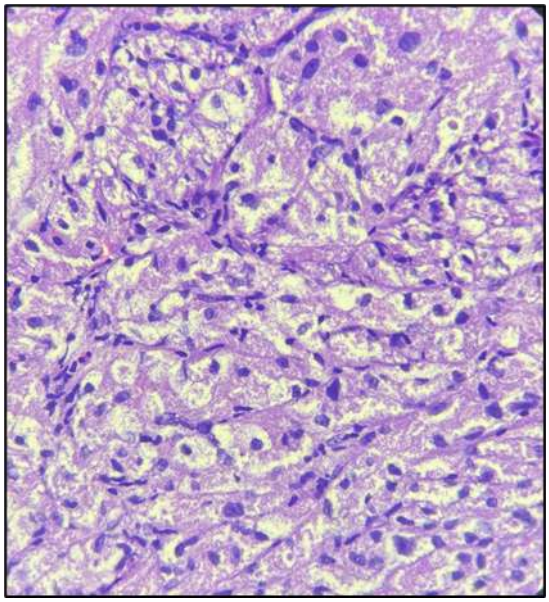


Figure 3: Photomicrography portraying sheets of cells exhibiting round nuclei with abundant granular cytoplasm with ill-defined cell borders.



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