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Case Report

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GRANULAR CELL TUMOUR OF THE OESOPHAGUS

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ABSTRACT

Granular cell tumours (GCTs) are rare and usually benign neoplasms of neural origin that are thought to derive from Schwann cells.[1] Although GCTs can occur at many sites in the body, approximately 6% of these tumours arise in the oesophagus.[3] Because GCTs are rare, there is little guidance regarding the most appropriate investigation and management of these tumours. Endoscopy is the gold standard diagnostic investigation for GCT of the oesophagus. Endoscopic ultrasound (EUS) can be helpful in differentiating between GCTs and other pathology. The spectrum of management ranges from surveillance to surgical resection. This case report describes a case of an oesophageal granular cell tumour and a review of the current literature regarding these rare tumours is undertaken.

KEYWORDS: Granular cell tumour, Abrikossoff's tumour, Malignant granular cell tumours

INTRODUCTION

Granular cell tumours (GCTs) are rare and usually benign neoplasms of neural origin that are thought to derive from Schwann cells.[1] They are also referred to as 'Abrikossoff's tumour' after the Russian pathologist Alexei Abrikossoff who first described them as "myoblastenmyome" (myoblastoma) in 1926.[2] GCTs usually lie within the sub-mucosal layer, but can involve the mucosa and muscularis propria.[2]

These tumours can occur at many sites in the body, but most often arise from the skin. Approximately 6% of all GCTs arise in the oesophagus.[3] As GCTs are rare; there is little guidance on the most appropriate investigation and management of these tumours.

In this case report we present a case of an oesophageal granular cell tumour managed by surveillance and we undertake a review of the current literature regarding these rare tumours.

CASE REPORT

A 57year old male, with a background history of alcohol dependency, chronic pancreatitis and a known oesophageal GCT, presented with a 1day history of sudden onset, severe epigastric pain associated with odynophagia. Oesophago-gastro-duodenoscopy (OGD) was performed and a 13mm round, protruding, sub-epithelial mass was noted 31cm from the incisors (Figure 1). Grade D oesophagitis was reported distal to the mass and a 4cm hiatus hernia was also present.

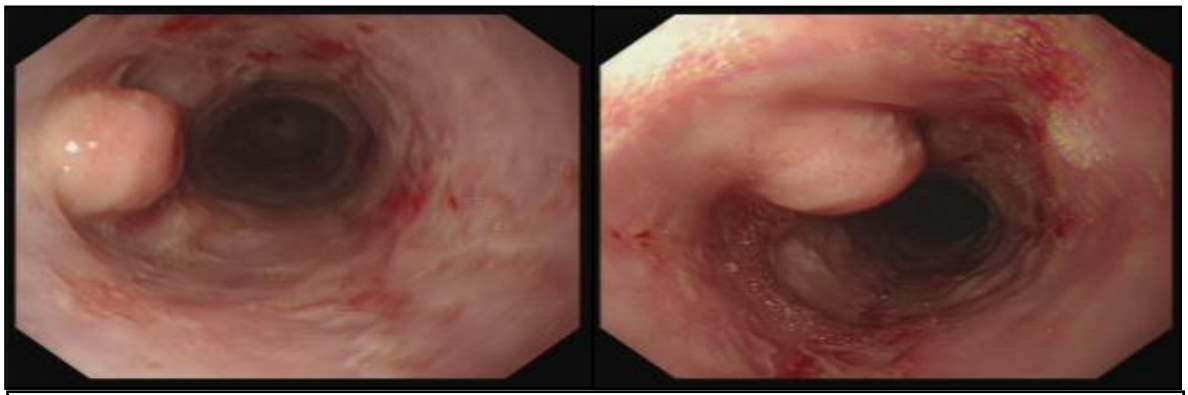

Figure 1: OGD images of the patient's smooth, sub-epithelial mass in the mid-to-distal oesophaghus. Grade D oesophagitis is also evident distally.

The biopsy report from the oesophageal tissue demonstrated rounded, cytologically benign cells with 'striking' granular cytoplasm. Immuno-histochemical staining for S-100 protein was positive, consistent with the patients' previous diagnosis of gastrointestinal GCT.

Since this patient's initial diagnosis of oesophageal GCT in 2007, he had been managed by surveillance OGD with repeated biopsies, that consistently showed granular cytoplasm with overlying hyperplasia and on one occasion demonstrated underlying proliferation of eosinophilic cells, consistent with a histological diagnosis of GCT. On all occasions there was no histological evidence to suggest malignancy.

Computed tomography was performed to further characterise the mass, and this confirmed mild oesophageal wall thickening and mucosal enhancement. Incidentally, an abnormal heterogeneously enhancing mass was noted in the right deltoid region, which was reported as suspicious of muscle metastasis. MRI was recommended as the imaging modality of choice to assess this possible metastatic muscle lesion. Unfortunately the patient died from unrelated causes before MRI or other further investigation of the muscle lesion could be carried out.

DISCUSSION

There are approximately 250 reported cases of oesophageal GCT.[4] A study of 31 cases found oesophageal GCTs to be diagnosed usually in middle age (median age 49) with the majority in the mid to distal oesophagus, as was the case with our patient.[5] In two separate case series of patients diagnosed with oesophageal GCTs, the most common indication for endoscopy was epigastric pain/discomfort, dysphagia and heartburn, with dysphagia being more common in tumours ≥1.5cm.[6,7]

Endoscopy is the gold standard diagnostic investigation for GCT of the oesophagus, where they typically appear as sessile, white-to-grey lesions with a smooth surface (Figure 1).[4] During upper GI endoscopy GCTs may however be mistaken for lipomas, leiomyomatas or cysts. EUS can be helpful to differentiate between GCTs and other pathologies and usually demonstrates a hypoechoic lesion with a solid pattern.[8] EUS is particularly useful in defining depth of tumour involvement to aid planning of further management including endoscopic or surgical resection.

A definitive diagnosis of GCT is made on pathological and immunohistochemical analysis. Histopathology typically demonstrates nests of large polygonal cells with abundant, eosinophilic, granular cytoplasm and small, dark nuclei.[9] Immunohistochemical staining for S-100 is thought to be diagnostic for GCT, but they also stain for CD68, inhibin-α and PGP 9.5 (Figure 2).[1]

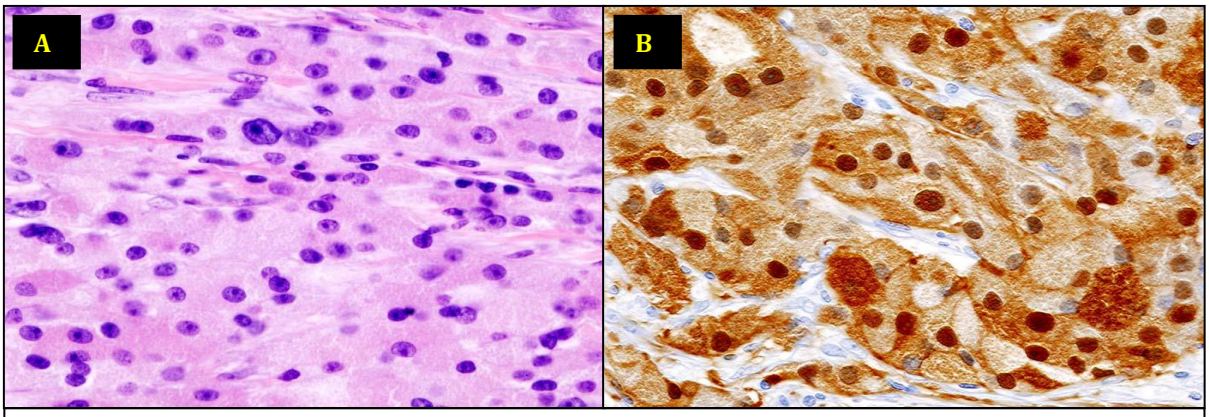

Figure 2: A - Histopathological appearance of granular cell tumour, and B - Immunohistochemical staining for S-100.
[Image credit: Wikimedia Commons]

Although extremely rare, malignant GCTs have a 5 year survival rate of less than 35%.[10] Fanburg-Smith et al. describe six histological features that suggest malignancy in GCTs; these are 1. necrosis, 2. spindling, 3. vesicular nuclei with large nucleoli, 4. increased mitotic activity, 5. high nuclear to cytoplasmic ratio and 6. pleomorphism[11]. In addition 50% or more of cells expressing p53 and Ki67 proliferative index of 10-50% was related to malignancy.

Biopsy in our patient demonstrated no histological evidence suggestive of malignancy, however, there are reports of clinically metastatic oesophageal GCTs that do not exhibit histological features of malignancy.[12]

There is no definitive and agreed management strategy for oesophageal GCTs. The spectrum of management ranges from surveillance to surgical resection.

A review by Rezende et al. discusses the treatment options.[13] Endoscopic surveillance with repeated

biopsies is recommended for small, histologically benign GCTs with mild to no symptoms. Endoscopic treatment is recommended for larger GCTs of 10-20mm, which are symptomatic, histologically benign and do not invade into the muscularis propria.

Surgical management is reserved for malignant or invasive GCTs (beyond muscularis propria) or where there is a contraindication to endoscopic management. Endoscopic mucosal resection (EMR) and endoscopic submucosal dissection (ESD) have both shown to be safe and effective options for GCTs, with ESD being carried out for submucosal GCTs.[14,15]

CONCLUSION

Granular cell tumours are rare, usually benign tumours of neural origin thought to derive from Schwann cells.

6% of GCTs arise in the oesophagus.[3] Endoscopy is the gold standard diagnostic investigation for GCTs of the oesophagus.[4] EUS is also useful particularly for defining depth of tumour involvement to aid in planning of further management. A definitive diagnosis of GCT is made on pathological and immunohistochemical analysis.

Endoscopic surveillance with repeated biopsies is recommended for small, histologically benign GCTs, endoscopic treatment (EMR and ESD) is recommended for larger GCTs of 10-20mm, which are symptomatic, histologically benign and do not invade into the muscularis propria, and surgical resection is reserved for malignant or invasive GCTs or where there is a contraindication to endoscopic management.

Grant

None.

CONFLICTS OF INTEREST

We the authors of this case report have no conflicts of interest to declare.

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