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# Esophageal granular cell tumour: A case report and review of literature

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**Abstract**
Granular cell tumours (GCTs) are uncommon tumours of neurogenic origin, thought to originate from Schwann cells. They are most commonly seen in skin, subcutaneous tissues and oral cavity, and are rarely found in gastrointestinal tract (8-10% of cases). Esophageal involvement is seen only in 2% cases. They are slow growing in nature, usually are asymptomatic and detected incidentally during investigation for other gastrointestinal causes. Because of the rarity of GCTs in clinical practice, proper workup and management are essential to avoid the potential morbidity and mortality. Endoscopic submucosal resection is the primary treatment which is done in most cases. Herein, we report a case of 85 year old female who was worked up for dyspepsia and dysphagia and was diagnosed as a case of Granular cell tumour on histopathology.

**Keywords:** Endoscopy, esophagus, granular cell tumour, submucosal resection

## Introduction
Granular cell tumours are rare and uncommon benign soft tissue neoplasms originating in the Schwann cells of the nerve sheath. They were first reported by Abrikossoff in 1926, in a case series comprising of benign tumours of tongue [1]. Subsequent studies revealed that they can occur at any site in the body, most frequently being skin and subcutaneous tissues and less commonly seen in thyroid, nervous system, respiratory system, female urogenital tract and gastrointestinal tract [2, 3]. GCTs involving the gastrointestinal tract account for only 8% of all GCTs, and esophageal involvement are seen in only 2% cases [4]. Since the first report of Granular cell tumour, only 300 esophageal Granular Cell Tumours have been reported in literature [5]. Esophageal GCTs exhibit almost the same clinicopathological characteristics as GCTs of other sites, however, due to rarity and low incidence, there is presently no consensus regarding appropriate esophageal GCT diagnosis and management. This present study is intended with emphasis on clinical features, histopathological findings, diagnosis and management.

## Case Report
An 85 year old female patient, presented to our Gastroenterology Department with complaints of dyspepsia, dysphagia, vomiting and chest pain. The patient was haemodynamically stable. They patient had no significant past/medical history. UGI Endoscopy revealed a 10 mm sized nodular submucosal lesion with central depression in the distal oesophagus, 33cm from upper incisor. The lesion was non-obstructing and non-circumferential. Further investigation using EUS confirmed a 10.2 x 0.5mm, well demarcated, hypoechoic lesion in oesophagus, confined to submucosa, without invasion into the muscularis propria.
Endoscopic mucosal resection was done for the lesion. Endoscopic appearance of the tumour surface was smooth and grayish white. The primary endoscopic diagnosis was Carcinoid Tumour. The microscopic examination revealed a well demarcated submucosal tumour composed of minimally pleomorphic spindle cells having abundant eosinophilic granular cytoplasm and a central nucleus. Mitotic count was very low (<5/50hpf). No necrosis was present. A histopathological diagnosis of Benign Mesenchymal Tumour was made, differentials being leiomyoma, Gastrointestinal Stromal Tumour (GIST) and Granular Cell

tumour. The section was subjected to Immunohistochemistry (IHC) to reach the correct diagnosis. IHC was negative for c-kit, DOG-1 and SMA while it was positive for S100 and PAS-D. Ki-67 was less than 2%. Hence, a final diagnosis of benign esophageal Granular cell tumour was made. The patient was referred to the medical oncologist for follow-up. After two years, no recurrence was noted and the patient was asymptomatic.

**Discussion**

Granular cell tumours (GCTs), also known as Abrikossoff's Tumours or granular cell myoblastomas are uncommon tumours and esophageal GCTs are even rarer. Of all esophageal tumours, only 1% is GCTs. The neoplasm arises from the neural or Schwann cells [1, 6].

Oesophageal Granular Cell Tumours can occur at any age, but more common in 40-60 year old patients with female predilection [7, 8]. It has been reported that they predominantly occur in distal oesophagus (~65%) and commonly present as solitary lesions, but can also occur at multiple sites [4, 8].

Granular Cell Tumours are insidious in nature and usually asymptomatic at diagnosis. Typically, the endoscopic feature is a non pedunculated, grayish white, firm to hard, elevated lesion with a smooth surface. Rarely the surface can be ulcerated [1, 4]. These tumours are usually confined to submucosa, but can involve mucosa and muscularis propria. If muscularis propria is involved, then it is difficult to differentiate it from an intramural leiomyoma, GIST and other benign tumours of oesophagus.

Endoscopic ultrasound (EUS)/ Esophagogastroduodenoscopy (EGD) play an important role in both diagnosis as well as management. They allow evaluation of location, depth of invasion, tumour size and lymphatic involvement. They usually have hypoechoic and homogenous pattern which help to differentiate GCTs from malignant lesions, cysts, inflammatory polyps and lipomas. However, a definitive diagnosis is always made on histopathological examination and Immunohistochemistry (IHC) [8, 9].

Histologically, GCTs characteristically reveal nests and sheets of polygonal or spindle cells with small round nuclei and abundant eosinophilic granular cytoplasm. Histological features mimic oesophageal spindle cell tumours, GIST, leiomyomas and rarely spindle cell variant of squamous carcinomas. The point of differentiation here is N: C ratio, which is usually low in GCTs [10].

IHC plays an important role in the confirmatory diagnosis of GCTs. The positive staining of S100 was first reported in 1986. A complete workup includes PAS, NSE, nestin and CD 68. Negative markers include SMA, desmin, CD 117 and CD 34. Ki-67, a nuclear proliferation associated antigen and a relative marker of cell proliferation, is usually negative in majority of cases. Unlike GCTs, oesophageal leiomyomas are positive for both SMA and desmin, but are negative for S 100. Melanomas and malignant schwanommas are S100 positive, but are also positive for HMB-45 and Vimentin with high Ki67 [11-13].

Malignant oesophageal GCTs are extremely are, less than 2% of cases. Nevertheless, it is important to identify malignant features, because of the potentially aggressive features and bad prognosis [6]. In 1998, Fanburg-Smith *et al* suggested 6 histological criteria for differentiating benign from malignant tumours-Increased N:C ratio, nuclear pleomorphism, vesicular nuclei with prominent nucleoli, tumour necrosis, spindling and increased mitotic activity (more than 2/hpf) [3].

The management of GCTs includes

a) Conservative endoscopic follow up for small asymptomatic oesophageal tumours (≤1cm. in diameter)
b) Endoscopic or surgical removal for lesions that are symptomatic, fast growing or ≥1 cm. in diameter.

Treatment decisions with respect to surgical intervention versus endoscopic resection should be based on characteristics of tumours in each individual and the physician's expertise [4, 5].

**Conclusion**

Esophageal GCTs are uncommon tumours of GIT, often asymptomatic, solitary and diagnosed incidentally in middle aged female patients. EUS is the most beneficial diagnostic tool, which helps in planning appropriate treatment based on depth of invasion. IHC staining of tumour markers aids in the differential diagnosis.

**References**

1. Abrikossoff A. Myomas originating from transversely striated voluntary musculature. Virchows Arch A Pathol Anat Histol. 1926; 260:215-33.
2. Nie L, Xu G, Wu H, Huang Q, Sun Q, Fan X. Granular cell tumour of the esophagus: a clinicopathological study of 31 cases. Int J Clin Exp Pathol. 2014; 7(7):4000-7.
3. Fanburg-Smith JC, Meis-Kindblom JM, Fante R, Kindblom LG. Malignant granular cell tumour of soft tissue: diagnostic criteria and clinicopathological correlation. Am J Surg Pathol. 1998; 22(7):779-94.
4. Johnston J, Helwig EB. Granular cell tumours of the gastrointestinal tract and perianal region: a study of 74 cases. Dig Dis Sci. 1981; 26:807-16.
5. De Rezende L, Lucendo AJ, Alvarez-Arguelles H. Granular cell tumours of the esophagus: report of five cases and review of diagnostic and therapeutic techniques. Dis Esophagus. 2007; 20:436-43.
6. Morrison JG, Gray GF, Jr, Dao AH, Adkns RB, Jr. Granular cell tumours. Am Surg. 1987; 53:156-60.
7. Zhong N, Katzka DA, Smyrk TC, Wang KK, Topazian M. Endoscopic diagnosis and resection of esophageal granular cell tumours. Dis Esophagus. 2011; 24:538-43.
8. Palazzo L, Landi B, Cellier C, Roseau G, Chaussade S, Couturier D, Barbier J Endosonographic features of esophageal granular cell tumours. Endoscopy. 1997; 29:850-53.
9. Voskuil JH, Van Dijk MM, Wagenaar SS, Van Vliet AC, Timmer R, Van Hees PA. Occurrence of esophageal granular cell tumours in The Netherlands between 1988 and 1994. Dig Dis Sci. 2001; 46:1610-4.
10. Goldblum JR, Rice TW, Zuccaro G, Richter JE. Granular cell tumours of the esophagus: a clinical and pathological study of 13 cases. Ann Thorac Surg. 1996; 62:860-5.
11. Parfitt JR, McLean CA, Joseph MG, Streutker CJ, AI-Haddad S, Driman DK. Granular cell tumours of the gastrointestinal tract: expression of nestin and clinicopathological evaluation of 11 patients.

Histopathology. 2006; 48:424-30.
12. Maekawa H, Maekawa T, Yabuki K, Sato K, Tamazaki Y, Kudo K *et al*. Multiple esophagogastric granulae cell tumours. J Gastroenterol. 2003; 38:776-80.
13. Mitomi H, Matsumoto Y, Mori A, Arai N, Ishii K, Tanabe S *et al*. Multifocal granular cell tumours of Thr gastrointestinal tract: Immunohistochemical findings compared with those of solitary tumours. Pathol Int. 2004; 54:47-51.