Case study

OESOPHEAL GRANULAR CELL TUMOR: A UNCOMMUN CASE REPORT.

Abstract:

Granulor cell tumors (GCT) formally known as Abrikossofftumors, are rare and usually benign tumors, most frequently occur in the skin and subcutaneous tissues but can also be found in the gastro-intestinal (GI) tract especially in the oesophagus (60% of cases).We discuss a case of a young woman with asthma who presented an acid reflux and was fortuitouslydiagnosed with oesophageal granular cell tumor (GCT) after esophagogastroduodenoscopy (EGD) and endoscopic ultrasound (EUS).We discuss clinical, endoscopic and histopathological aspects of this case with the different criteria of diagnosis.

Keys words: Oesophealgranular cell tumor, oesophagus, diagnosis,

Introduction:

Granulor cell tumors(GCT) formally known as Abrikossofftumors , are rare and usually benign tumors, most frequently occur in the skin and subcutaneous tissues but can also be found in the gastro-intestinal (GI) tract especially in the oesophagus (60% of cases) [1].

Oesophageal GCTs are subepithelialtumors, accounting for approximately 1% of all GCTs [2].They are usually asymptomatic and accidentally discovered by endoscopic examination for symptoms not associated with tumor such as reflux acid. The Diagnosis is histological and treatment is controversial.

In order to explore the clinicopathological characteristics of the tumor,we describe a case of a young patient diagnosed accidentally by Oesophageal GCT after releasing an upper gastrointestinal endoscopy for acid refux.

CASE PRESENTATION:

A 32-year-old patient with medical history of asthma presented to our department of gastro-enterology for intermittent regurgitation and complained of acid reflux for 2 years, sometimes complicated with vomiting. Upper gastrointestinal endoscopy has found fortuitouslywhite sub-mucosal lesion measured approximately 4mm, in the distal oesophagus at 35 cm from incisors and a superficial gastritis (active phase). The diagnosis as GCT's was realised by standard histology with hematoxylin and eosin after biopsy of the lesion.

The histologyshowed a mesenchymal tumour proliferation composed of sheets of polygonal cells with abundant eosinophilic and granular cytoplasm with a small, round and centrally hyperchromatic nucleus. The epithelium overlying this proliferation is acanthosic showing pseudo epitheliomatoushyperplasia.The diagnosis was confirmed by immunohistochemistry showing a strong staining for S100 protein.

The physical examination including oro-pharyngeal examination found no abnormalities and no other localisation of the tumor.Therefore, the patient underwent a CT scan that came normal.

Endoscopic ultrasonography revealed a low echo, homogenous and oval lesion intramural, sub-epithelial, and within the deep mucosa in layer 2 with well-defined borders (figure 2).

We reassured the patient that a granular cell tumor is typically a benign mucosal tumor with a low risk of recurrence and malignant degeneration. A follow-up EUS at 3 months showed no significant changes and the patient became asymptomatic for her reflux acid after treatment.

Figure 1: endoscopy demonstrated a white sub-mucosal lesion

at 35cm from incisors

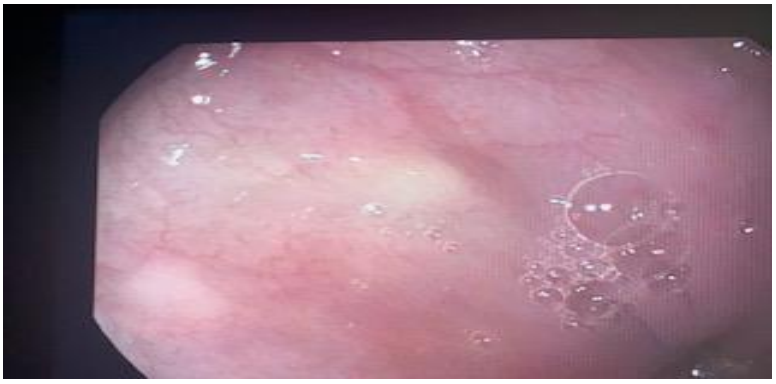


Figure 2 :sonographic image of the lesion on EUS measured 4.4mmx2.4mm

DISCUSSION

Granular cell tumorswere first described by Abrikossoff in 1931 and there have been only approximately 300 cases discussed in the literature [3-4]. Theyare rare tumorsand usually found in the skin and subcutaneous tissues [4]. Several detailed reviews found that approximately 1% – 8 % of all GCTs occur in the gastrointestinal tract [5]. Of all esophagealtumors, only 1% are GCTs [6].Oesophageal CGT is considered the most common site of occurrence with a predilection for the distal part (60% of the cases) [6].In fact, in a study led by Yongsheng Shi and all (Experience with Esophageal Granular Cell Tumors: Clinical and Endoscopic Analysis of 22 Cases) it was reported that approximately 65% of esophageal GCTs are located in the distal part of the esophagus, 20% in the middle part of the esophagus, and 15% in the proximal part of the esophagus [7]. However, another report claimed that the most common location of esophageal GCTs is the middle third of the esophagus [8]. In our case the lesion was detected in the distalesophagus.

This case is unique because the patient presented at an atypical young age with an otherwise typical presentation of GCT. Although GCT can occur at any age, it is more common between the fourth and sixth decades of life [8]. Our patient was only 32 years old. GCT is more common in women than men by a ratio of 2-3:1 [8]. It is also more common in African woman than white patients in a 3:1 ratio [8]. These characteristics of gender and race align with our patient's demographic. GCTs are often asymptomatic, but the most common presenting symptom is dysphagia [1].

The clinical manifestations of CGT depend on the size of the lesion. In fact, patients with small lesions less than 20 mm in diameter are usually asymptomatic and the lesions are found incidentally during endoscopy or radiography (it's the situation of our patient)[9]. However, patients with greater diameter lesions can present with symptoms such as dysphagia in the first place or with other less common symptoms like the gastro-esophageal reflux disease, dyspepsia, chest pain, cough or nausea [9].

Endoscopically, GCTs are typically yellow-white in color, firm, with a negative pillow sign [10]. This patient's findings on endoscopy were consistent with this presentation, and are seen in Figure 1. They are also typically hypoechoic with smooth margins arising from the second or third layer of the distal esophagus [11]. The patient's lesion was found in the second layer within the deep mucosa of the distal of the esophagus, as seen in Figure 2

Figure 3:histopathologic examination revealed large polygonal cells that contain eosinophilic granules with low nuclear-cytoplasmic ratios.Coloration with hematoxylin and eosin stain, original magnificationx100.

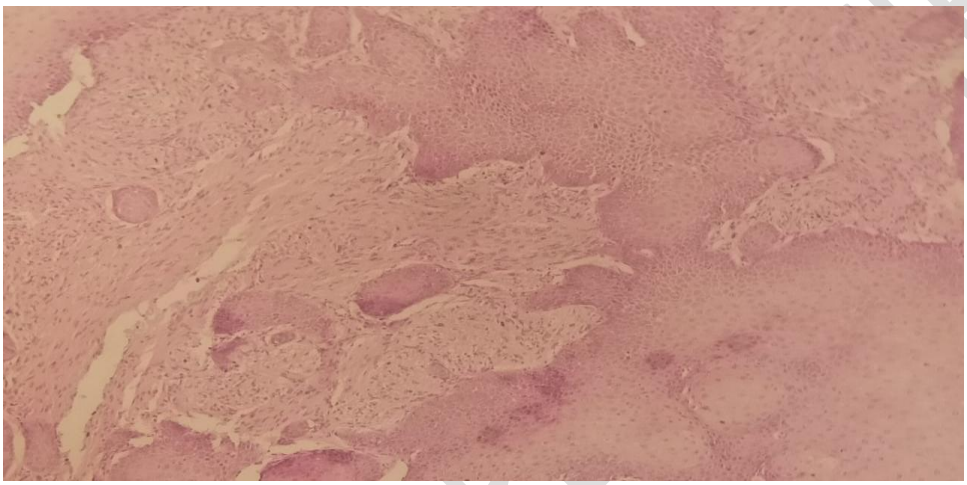
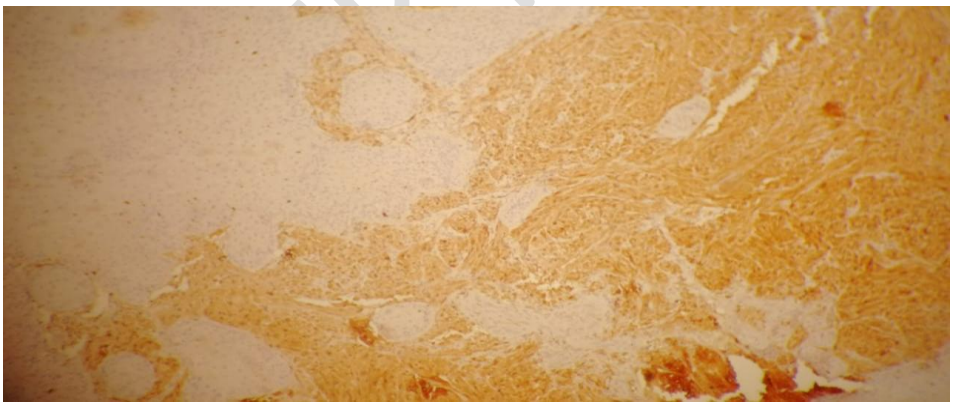


Figure 4: Immunohistochemistry shows a strong staining for S100, original magnification x 100.



Histologically, GCTs have large polygonal cells that often contain eosinophilic granules with low nuclear-cytoplasmic ratios [11,12]. Our patient had identification of GCT on histology with all of the above characteristics, and immunohistochemically, the tumor cells stained positive for S100.
It would staining identify also PAS, neuron-specific enolase, and nestin [13].
GCTs are normally benign, but there have been descriptions of malignant potential in the literature in 4% of lesions, generally with those lesions over 4 cm [13-14]. Fanburg-Smith identified six criteria to clarify malignant potential [15]. The six histologic criteria are necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity, high nuclear to cytoplasmic ratio, and pleomorphism [15]. With these criteria in mind, unresected lesions that are asymptomatic and under on 1 cm should

be monitored with EGD or EUS every one to two years for an increase in size [1]. If the lesions are symptomatic or over 1 cm, endoscopic mucosal resection or endoscopic sub-mucosal dissection is recommended [1].

CONCLUSION

Esophageal GCTs are uncommon tumors of the GI tract that are often diagnosed incidentally in middle-aged patients. The utility of EUS is invaluable for determining lesion size, origin, borders, and echogenic structure. Although GCTs are typically asymptomatic with an insidious clinical course, approximately 2% of GCTs are malignant upon histopathologic examination. If resection is not released for the small lesion, a follow up with monitoring by endoscopy must be conducted.

REFERENCES

1. Benchekroun Z, Akammar A, Bennani H, et al. Atypical esophageal granular cell tumor: Case report. *Radiol Case Rep*. 2021;16(12):3995-3999. Published 2021 Oct 22. doi:10.1016/j.radcr.2021.09.040.
2. Christopher PR, Kingsley PA, Singh Bedi H, Singh Kwatra K, Rathore S, Das KC. Large Mid-esophageal granular cell tumor: benign versus malignant. Rare Tumors. 2015;7:76–78.
3. De Rezende L, Lucendo AJ, Alvarez-Argüelles H. Granular cell tumors of the esophagus: report of five cases and review of diagnostic and therapeutic techniques. Dis Esophagus. 2007;20(5):436-43. doi: 10.1111/j.1442-2050.2007.00692.x. PMID: 17760659.
4. Zhang, M, Sun Z, Zou X: Esophageal granular cell tumor: clinical, endoscopic and histological features of 19 cases. Oncology letters. 2014, 8:551-555. 10.3892/ol.2014.2152
5. 123–7 PMID: 26306145; PMCID: PMC4543730. doi:10.4251/wjgo.v7.i8.123. [7] Ordóñez NG. Granular cell tumor: a review and update. AdvAnatPathol 1999; 6:186–203 PMID: 10410172.
6. McSwain GR, Colpitts R, Kreutner A, O'Brien PH, Spicer S. Granular cell myoblastoma. SurgGynecolObstet 1980; 150:703–10 PMID: 6245475. [9] Orlowska J, Pachlewski J, Gugulski A
7. Orlowska J, Pachlewski J, Gugulski A, Butruk E. A con-servative approach to granular cell tumors of the esophagus: four case reports and literature review. Am J Gastroenterol 1993;88:311–15.
8. De Rezende L, Lucendo AJ, Alvarez-Arguelles H. Granular cell tumors of the esophagus: report of five cases and review of diagnostic and therapeutic techniques. Dis Esophagus 2007;20:436–43.
9. Wang HQ, Liu AJ. Esophageal granular cell tumors: Case report and literature review. *World J GastrointestOncol* 2015; 7(8): 123-127 [PMID: 26306145 DOI: 10.4251/wjgo.v7.i8.123],
10. Chen WS, Zheng XL, Jin L, Pan XJ, Ye MF. Novel diagnosis and treatment of esophageal granular cell tumor: report of 14 cases and review of the literature. Ann ThoracSurg. 2014;97:296–302.
11. Zhong N, Katzka DA, Smyrk TC, Wang KK, Topazian M. Endoscopic diagnosis and resection of esophageal granular cell tumors. Dis Esophagus. 2011;24:538–543.
12. Radaelli F, Minoli G: Granular cell tumors of the gastrointestinal tract. Questions and answers . J GastroenterolHepatol. 2009, 5:798-800.
13. Souza LCA, Pinto TDA, Cavalcanti HOF, et al. Esophagealschwannoma: case report and epidemiological, clinical, surgical and immunopathological analysis. Int J Surg Case Rep. 2019;55:69–75.
14. Narra SL, Tombazzi C, Datta V, Ismail MK. Granular cell tumor of the esophagus: report of five cases and review of the literature. Am J Med Sci. 2008 May;335(5):338-341. doi: 10.1097/MAJ. 0b013e3181568197.

15. Fanburg-Smith JC, Meis-Kindblom JM, Fante R, Kindblom LG. Malignant granular cell tumor of soft tissue: diagnostic criteria and clinicopathologic correlation. Am J SurgPathol. 1998 Jul; 22(7):779-794.