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CASE REPORT

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Case report sustained by literature review on the major approaches of the granular esophageal cell tumor

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Abstract: Background: Granular Cell Tumors (GCT) are rare benign neoplasms with an estimated prevalence of 0,4% in retrospective series. About 8% of GCT are located in the gastrointestinal tract, having the esophagus as the main affected area. Malignancy was reported in 2-4% of cases and there are no well established treatment protocols. Case summary: It was present an oligo-symptomatic female patient with diagnosis of GCT presented on the endoscopy. Complementary investigation was based on endoscopic ultrasound (EUS). The treatment consisted of mucosectomy and endoscopic variceal ligation, evolving with a favorable outcome. Conclusion: Despite disagreement on protocols for treatment of GCT, mucosectomy appears to be an effective option.

Keywords: Granular cell tumor. Esophagus. Benign neoplasm. Endoscopic mucosal resection.

Introduction

Granular Cell Tumors (GCTs) are benign neoplasms, derived from Schwann Cells located at the submucosa [1]. It's real prevalence is unknown. However, a retrospective series showed that GCTs represent 0.4% of all esophageal benign tumours [2]. It is predicted that 8% of GCTs are located in the gastrointestinal segment, with ⅔ of cases presented on the esophagus. Moreover, the malignancy rate is estimated at 2-4% [3].

Series and case reports describe the lesion as an incidental find in upper gastrointestinal endoscopies prescribed for other diagnostic hypotheses [2]. In addition, it is discussed the etiopathogenesis of the disease, as well as diagnostic methods and differences on the therapeutic approaches benign tumours [4]. Although, there are no well-established treatment protocols benign tumours [2].

The present case report consists of a case of a patient with dyspeptic complaints, diagnosed with GCT. Furthermore, a review of the literature on etiopathogenesis, diagnostic investigation, treatment and management of this type of neoplasm.

Methods

The present study is a case report whose bibliographic research used the descriptors (MeSH Terms) Granular cell tumor. Esophagus. Benign neoplasm. Endoscopic mucosal resection. The research was carried out through the study of digital articles and virtual books attached to the PubMed, Embase, Ovid, Cochrane Library, Web Of Science, ScienceDirect Journals, Scopus, academic Google, in database platforms such as Scientific Electronic Library Online (SCIELO), PubMed, and in scientific repositories, following the rules of systematic review – PRISMA. Transparent reporting of systematic reviews and meta-analyzes, Available in: https://www.prisma-statement.org/. Accessed on 12/23/2020. Used as the main data sources, the most relevant works were selected for the theme for synthesis and presentation of information, excluding references that diverged from the purposes covered here.

Case Report

The present study was elaborated according to the rules of CARE case report. Available in: https://www.care-statement.org/ Accessed on 12/23/2020.

Patient Information

Female, 49 years old, unemployed. Came to our service to perform diagnostic endoscopy and colonoscopy due to intermittent bloating and postprandial regurgitation. Personal history of hypertension and diabetes mellitus and no (significant) family history. No previous abdominal surgery.

Clinical Findings

Physical examination without particularities, except for being overweight.

Timeline

The patient looks for assistance due to intermittent postprandial bloating and regurgitation, without relieving factors. The symptoms had started a month ago and were accompanied by nausea and early satiety. Other signs and symptoms, such as weight loss, bleeding and dysphagia were not reported.

Diagnostic Assessment

Laboratory test results showed no significant abnormality: Hb 124 g/L, Hct 35%, WBC 8.6 x 109/mm3, Platelets 230 x 109/mm3, PT 37.8, INR 1.1 and PTT 26.3 s. An upper gastrointestinal endoscopy was performed, which showed a yellowish-white nodular lesion with subepithelial appearance and fibroelastic consistency at the distal esophagus, measuring approximately 6mm (Figure 1). Colonoscopy identified a sessile polyp in the ascending colon. Biopsies of the esophageal lesion and colonic polyp were performed, revealing a tumor of granular esophageal cells (Figure 2) and low-grade tubular adenoma, respectively.

Figure 1 Echoendoscopic image of the granular cell tumor.

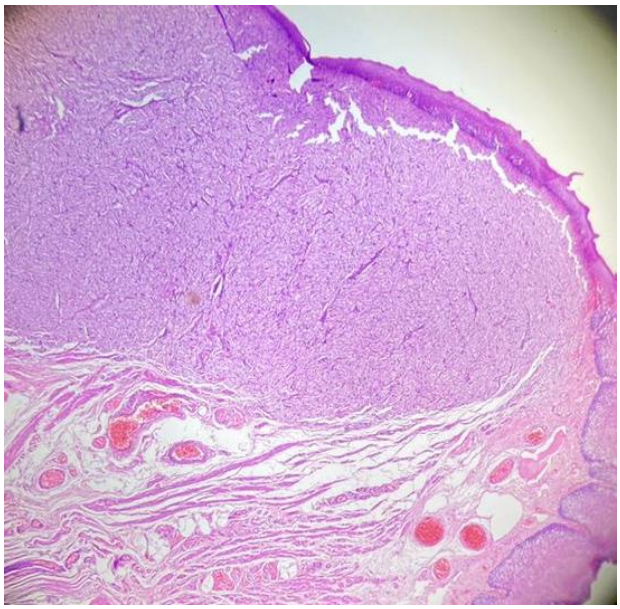

Figure 2 Histopathological image of the granular cell tumor.

In a joint decision between the patient and medical team, we opted for resecting the esophageal lesion. In consequence, a complementary exam for its staging was requested in another institution, due to unavailability in our service. The diagnosis of subepithelial lesion was confirmed with the echoendoscopy. The lesion was described as oval, hypoechoic, homogeneous, of approximately 5.6 mm x 1.6 mm, with precise limits and regular contours. Inserted in the deep mucosa, it had a discrete layer rejection in the submucosa and there was no evidence of impairment of regional lymph nodes. Whereas it was a small lesion, benign in nature, with no invasion of deep layers, a good prognosis for the patient could be inferred.

Therapeutic Intervention

It was chosen for the endoscopic mucosectomy with the aid of an elastic band under sedation. A frontal view endoscope and a single channel made it possible to identify the lesion in the distal third of the esophagus. Followed by the use of an elastic band, which was triggered involving the entire lesion and the resection, made with a diathermic loop in Blend 1 mode/power of 30W. The piece was then captured with the same gripping and externalization loop throughout the mouth, which revealed to be free of macroscopic lateral margins.

Proton Pump Inhibitor (Omeprazole 40mg/day) was prescribed for daily use along with symptomatics as needed. The patient was discharged on the same day. The submucosal injection of saline solution was

not necessary once the capture of the lesion with free margins using the elastic band kit was successful (Supplementary Video https://youtu.be/j09aK6Iyy08).

Follow-up and Outcomes

One month after the intervention, the patient remained asymptomatic. The anatomopathological study confirmed the diagnosis of granular cell tumor located in the submucosa. The tumor consisted of uniform cells of abundant and granular eosinophilic cytoplasm, with small nuclei and homogeneous chromatin. Furthermore, the lesion presented with free lateral and deep margins, affirming the absence of malignancy. The procedure used, although complex, when performed by experienced practitioners has a great tolerance for patients due to its low-invasive characteristic and uniqueness. No adverse effects have been reported.

Patient Perspective

The tumor diagnosis initially caused a negative impression of the clinical picture even more in a patient with an anxious profile, however, the endoscopy team explained Initially the tumor diagnosis caused a negative impression in the prognosis, especially in a patient with an anxious profile. However, the endoscopy team explained in detail all the therapeutic options, as well as the need for complementary diagnosis with an exam not available in the present institution.

All decisions were made by mutual agreement, respecting the patient's autonomy and the resection was performed without complications. The follow-up was made by telephone contact and she was satisfied with the result.

Discussion and Literature Revision

The patient is within the epidemiological group most affected by esophageal granular cell tumors and the diagnosis, as pointed out by the literature, was made accidentally. It is known that, although small, there is a risk of growth and malignancy development [1]. In addition, it is evident the difficulty of carrying out highly complex tests through the public health system in the present country, as well as the obstacles concerning the follow-up of those cases. Thus, in accordance with the patient's will, we opted for the endoscopic resection. Granular cell tumor (GCT) is a rare type of tumor. Its character of insidious onset sometimes leads to negligence or wrong diagnosis of the disease.

GCTs were first described by Abrikossoff in 1926. He reported this type of lesion in different parts of the body, most commonly in the oral cavity, skin and subcutaneous tissue, and less frequently affecting the breasts, thyroid, respiratory tract, biliary segment, female genital tract, nervous system and the gastrointestinal tract as a whole [1]. Approximately 8% of GCTs develop in the gastrointestinal tract, regularly in the esophagus, involved in one to two thirds of the cases [2-4]. Esophageal GCTs more commonly affect the distal esophagus. Was reported that most of the GCTs were found in the distal esophagus (⅔), and only 20% and 15% in the middle and proximal esophagus, respectively [5]. The esophagus, GCTs are usually found incidentally during diagnostic endoscopy exams. It appears as a small nodule or plaque with a gray-white to yellowish color [6]. It is also generally restricted to the submucosal layer of the esophagus. However, it was found that only 26% of patients have this typical aspect [7]. The esophageal GCTs can also be present as red or gray-white. Rarely, ulcerated tumors are seen on the mucosal surface. The most widely accepted theory is that it has a neurogenic origin, derived from Schwann cells, component of the submucosal neural plexus in the esophagus. A malignant potential has been reported in 2 to 4% of cases, which increases with the tumor size [3].

GCTs are predominant in females and occur mainly during middle age, matching the patient's profile. Patients with lesions smaller than 2 cm are more likely to be asymptomatic. If symptomatic, they may show symptoms that mimic gastroesophageal reflux disease. These tumors are usually found incidentally during EGD [8].

EUS is the best procedure for assessing upper gastrointestinal submucosal lesions. It should be performed on all patients with the diagnosis of esophageal GCT, as the size of the tumor and the degree of invasion are important to define the treatment method [8]. EUS and endoscopic features together can distinguish GCT from lipomas, esophageal cysts and inflammatory polyps. Typical ultrasound characteristics were observed in 95-100% of cases, and histopathology confirmed the diagnosis of GCT for all 21 lesions after their removal [9]. In another study, it was found that the EUS identified 13 of the 14 cases of GCT before being confirmed histologically. These studies highlight the importance of EUS in the diagnosis of esophageal GCT [10].

Biopsy with standard forceps is sufficient to achieve the diagnosis in 50-83% of cases [1,11].

Endoscopic mucosal resection (EMR), EUS-guided biopsy and EUS-guided fine needle aspiration can also be used. By endoscopy and EUS, GCTs are difficult to distinguish from other submucosal tumors, for example leiomyomas or gastrointestinal stromal tumors. Therefore, histopathological analysis is essential [11].

A malignant potential has been reported in 2% to 4% of esophageal GCTs, especially if they are bigger than 1 cm [12]. The histological criteria for malignancy proposed by Fanburg-Smith are still debatable among pathologists, being metastasis the only malignancy criterion with unanimous agreement. Regarding GCTs with less than 1 cm, reports in the literature suggest conservative surveillance with endoscopy and EUS.

In general, several potentially less invasive endoscopic approaches are available to address GCTs, including endoscopic band ligation and diathermy loop. In addition, EMR is also viable in cases confined to the mucosa [13].

Others studies recommended the use of EMR for lesions with less than 2 cm in diameter; alternatively, endoscopic resection of the submucosal tunnel can be used for tumors with a diameter between 2 and 3 cm. Surgical removal with traditional open surgery or video-assisted thoracoscopic surgery was indicated for tumors with a high suspicion of malignancy, lesions originating from muscularis propria or, in some cases, with diameter between 2 and 3 cm [1,3]. Surgical removal with traditional open surgery or thoracoscopic surgery was recommended for tumors with a high suspicion of malignancy, tumors originating from the muscles themselves, or tumors larger than 3 cm in diameter. The criteria suggested for EMR are Tumor size less than <2 cm, absence of connection to the muscle itself and no evidence of anatomopathological malignancy [9].

These techniques can sometimes lead to unsatisfactory results, with incomplete or unclear resection status, especially in lesions that exceed the mucosa. As most GCTs show a submucosal manifestation, an ESD consists of an anatomically accurate resection method, once it performs a circumferential incision of the mucosa around the tumor, followed by the dissection of the submucosa [2]. Thus, with this approach, en bloc resection, a reliable histopathological diagnosis is more likely. In the case of a submucosal esophageal GCT, an ESD is also a suitable treatment [1].

Final Considerations

As it was shown, in the present study, the patient is within the epidemiological group most affected by esophageal granular cell tumors. Her diagnosis was established with a diagnostic ultrasound exam and an EUS. The less conservative approach was justified by the limit of growth risk and malignant potential for injury, in addition to the limitations of performing EDA series in the scope of the country's public health system. Regarding the treatment itself, it was decided to perform the EMR with the aid of elastic band, due to the small characteristic of the lesion (6 mm) and the restriction to the mucosa, as viewed previously on the EUS. The procedure was carried out successfully, without complications, and a sample was sent for anatomopathological study, confirming the accuracy of the diagnosis.

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Ethics Approval
Not required

Informed consent
Written consent was obtained from the patient

Conflict of interest
The authors declare no conflict of interest.

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