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SCIENTIFIC LETTERS

Granular cell tumor of the esophagus[☆]



Tumor de células granulares del esófago

In 1931, Abrikossoff described the first case of granular cell tumor (GCT) in the esophagus. These tumors can develop in any part of the body.¹ Eight percent of GCTs are found in the gastrointestinal tract and the esophagus is the most frequently affected organ.² The majority present as solitary lesions, but 10% of the cases are multifocal.² We present herein the case of a 21-year-old man, referred for endoscopy due to gastroesophageal reflux symptoms of one-year progression and dysphagia and retrosternal pain of recent onset. During the endoscopy, a yellow, hard, mobile, 15 mm lesion with a nodular aspect was seen at the middle esophagus (fig. 1). Diagnosis from the histopathologic analysis was a GCT of the esophagus and immunohistochemical staining showed protein S-100, CD56, and CD68 expression (fig. 2). The patient was re-evaluated through endoscopy that revealed a residual lesion of 4 mm in diameter. Endoscopic ultrasound (EUS) identified a hypoechoic lesion that completely depended on the esophageal mucosa. Mucsectomy was performed with the band ligation technique with no complications.

GCT is a white-to-gray lesion representing 0.0019-0.3% of all tumors. Most GCTs are solitary, as in our patient, but 10-15% can be multiple and pink or yellow in color.¹⁻³ Even though we found the tumor at the middle third of the esophagus, the most frequent location is at the distal third of the esophagus. Esophageal GCTs are located at the distal third, middle third, and proximal third in 65, 20, and 15% of cases, respectively.⁴

The origin and nature of a subepithelial tumor cannot be determined through endoscopy alone, and GCTs are no exception. EUS is currently the most accurate imaging technique for determining the gastrointestinal wall layer from which the tumor originates. This information, combined with the echogenicity of the study, helps make the differential diagnosis and evaluate the probability of endoscopic resection. In our patient, the endoscopic ultrasound showed a hypoechoic lesion that depended entirely on the mucosa, but it is important to point out that through EUS,

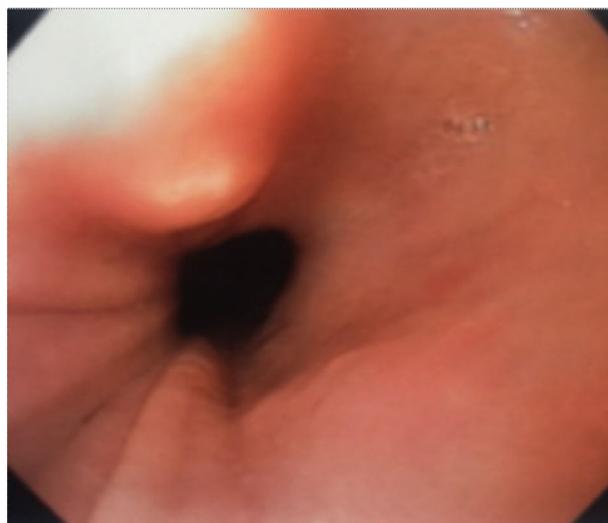


Figure 1 15 mm yellow esophageal lesion.

GCTs usually look like hypoechoic, homogeneous lesions with smooth margins that are predominantly located in the mucosa or submucosa. Nevertheless, they can also be found in the muscular or subserous layers.² On the other hand, even though the echographic pattern can clearly distinguish these tumors from other lesions or lipoma (the latter originates in the submucosa and is hyperechoic), the hypoechoic lesions located in the submucosa are prone to be poorly classified, because that layer of the gastrointestinal wall can give rise to different types of tumors with similar ultrasonographic characteristics. Therefore, the final diagnosis requires histologic analysis.²

Histologically, GCTs are composed of polygonal large cells that contain abundant eosinophilic granules, as observed in our patient.² In addition, they stain positive for the S-100 protein, suggesting a neural cell origin.^{2,5}

Although GCTs are usually benign neoplasias, malignant potential has been described in 4% of the lesions, especially in those larger than 4 cm.² The histologic malignancy criteria include tumor necrosis, fusiform cells, enlarged nucleoli, increased mitotic activity, augmented nuclear and cytoplasmic radii, and pleomorphism. The diagnosis of malignancy is made with the presence of 3 of these criteria.⁶

Resection in our patient was carried out through mucsectomy with the band ligation technique, but it can also be performed through other techniques, such as: endoscopic snare resection of the mucosa and endoscopic dissection of the submucosa.¹ Mucsectomy in any of its variants is useful when the GCT is confined to the mucosa. However, GCTs

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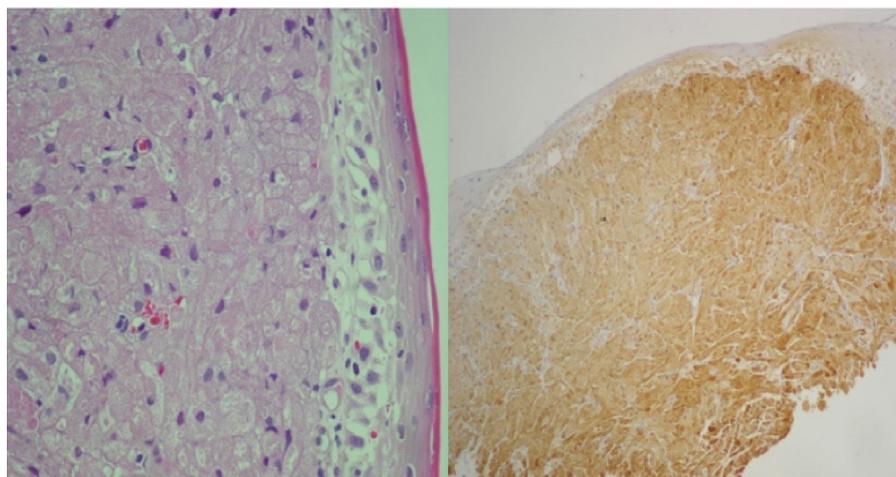


Figure 2 On the left, hematoxylin and eosin stain show abundant eosinophilic granules. On the right, immunohistochemistry shows S-100 protein expression.

frequently originate in or invade the submucosa, resulting in incomplete lesion resection. In such cases, endoscopic dissection of the submucosa is a safe and effective alternative that enables complete resection in 92.9% of the cases, when there is experience with the technique.⁷

In short, GCTs can have different aspects at endoscopy and they require histologic analysis with immunohistochemistry for diagnosis. The layer of origin and the appropriate technique to employ are established through EUS.

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Conflict of interest

The authors declare that there is no conflict of interest.

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