Granular cell tumors (GCTs) are a rare entity that can affect the whole human body. Concerning the esophagus, in 2% to 4% of cases, a malignant potential has been reported, which increases with size. That is why resection should be pursued, especially in lesions >1 cm. Because most GCTs show a submucosal manifestation, endoscopic submucosal dissection (ESD) is a favorable anatomically convincing method because it involves circumferential incision of the mucosa surrounding the tumor, followed by dissection of the submucosa. This approach makes en bloc resection more likely. The endoscopic management of a submucosal esophageal GCT by ESD is demonstrated here.

First described by Abrikossoff in 1926, GCTs are a benign entity with malignant potential that evolve from the Schwann cells of the nervous system. Principally, they can occur at any site of the human body. About 2% of GCTs are in the esophagus; other parts of the GI system are rarely involved. Owing to the rising availability and use of EGD, the discovery rate of GCTs has increased. Normally, patients with GCTs are asymptomatic, and most lesions are detected incidentally.

Figure 1. Gastroesophageal junction with granular cell tumor (star).

Figure 2. EUS view of the esophageal granular cell tumor showing a well-defined tumor (calipers) strictly limited to the submucosa.

Figure 3. The target lesion is circularly marked.
By endoscopy and EUS, GCTs are difficult to distinguish from other submucosal tumors, for instance leiomyomas or GI stromal tumors. Therefore, sufficient histopathologic analysis is required. Typically, GCTs show polygonal or round cells in compact nests with pyknotic nuclei surrounded by eosinophilic cytoplasm. Usually, immunohistochemical analysis gives strongly positive results for S100, neuron-specific enolase, Nestin, and several myelin proteins with negative staining for smooth muscle actin, Desmin, CD117, and CD34 inter alia.

In 2% to 4% of esophageal GCTs, a malignant potential has been reported, especially when they are >1 cm. Thus, resection should be pursued. In GCTs <1 cm, literature reports suggest conservative surveillance with endoscopy and EUS.

The technique of ESD was initially developed in Japan in the 1990s for the en bloc resection of flat gastric neoplasias. Hereofore, these lesions could endoscopically be resected only by a piecemeal technique by use of classic EMR, which often resulted in an unclear completeness of resection (R0). In the meantime, ESD has also been increasingly used in the esophagus and for colorectal lesions. Nowadays, it can be considered a standard treatment, especially for flat and laterally spreading lesions >2 cm.

After the suggestive area is detected, ESD begins by marking with coagulation dots with a safety margin of approximately 5 mm. This is followed by the submucosal injection of saline solution. With the ESD knife (in the case described here, the HybridKnife; Erbe,
Tuebingen, Germany), the lesion is then cut (eg, circumferentially) and dissected from the muscularis in the submucosal level. For this purpose, a conical or straight transparent ESD cap is used. For the lesions mentioned, ESD offers a reliable and oncologically convincing method.15

CASE REPORT

A 49-year-old woman with reflux disease underwent EGD, which revealed a 2-cm centrally ulcerating mass 35 cm from the incisors contacting the cardia (Fig. 1; Video 1, available online at www.VideoGIE.org). EUS showed a submucosal limitation (Fig. 2). Pathologic examination of a transmucosal biopsy specimen identified a GCT.

Because the lesion involved the submucosa, we decided to conduct ESD. It was performed with the patient under general anesthesia. For this purpose, a conical or straight transparent ESD cap is used. For the lesions mentioned, ESD offers a reliable and oncologically convincing method.15

Pathologic examination confirmed a submucosal GCT resected completely in sano (R0) (Fig. 5). No postprocedural adverse events were observed. Four weeks later, EGD showed a regular scar without signs of stenosis (Fig. 6).

DISCUSSION

In esophageal GCTs, a malignant potential of 2% to 4% is reported, which increases with size. That is why, especially in GCTs >1 cm, resection should be pursued.2,7 In general, several potentially less invasive endoscopic approaches are available to address GCTs, including endoscopic band ligation and diathermy loop.2,16 Also, EMR is feasible in cases confined to the mucosa;17 however, these techniques often lead to unsatisfactory results, with incomplete or unclear state of resection, particularly in lesions exceeding the mucosa.2

Because most GCTs show a submucosal manifestation, ESD offers an anatomically precise resection method because it involves circumferential incision of the mucosa surrounding the tumor, followed by dissection of the submucosa. Thus, by this approach, en bloc resection with a reliable histopathologic diagnosis becomes more likely.2 If an esophageal GCT is limited to the submucosal layer, ESD is an adequate treatment.

DISCLOSURE

All authors disclosed no financial relationships relevant to this publication.

Abbreviations: ESD, endoscopic submucosal dissection; GCT, granular cell tumor.

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