

Endoscopic Resection of a Granular Cell Tumor: A Case Report

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ABSTRACT

Background: Granular cell tumors (GCTs) are benign soft tissue tumors that are thought to originate from Schwann cells. They are relatively rare, and presentation in the gastrointestinal tract is even more uncommon.

Case report: In this paper, we present a 26-year-old female patient who was incidentally diagnosed with an esophageal GCT. The tumor was initially detected by endoscopy, evaluated by endoscopic ultrasonography (EUS), then resected using an endoscopic mucosal resection technique.

Keywords: Endoscopic, Granular Cell Tumor.

INTRODUCTION

Granular cell tumors are rare, generally benign tumor of neural origin (Schwann cell tumors) that can occur at any site of the body but are usually found on the tongue, subcutaneous tissues, and breast. Located in gastrointestinal tract is uncommon, and most commonly presented in the esophagus.

GCTs were first described in the tongue by Abrikossoff in 1926 and were previously called granular cell myoblastomas or Abrikossoff's tumors. Esophageal GCTs, however, were first reported in 1931. ⁽¹⁻³⁾ Large GCTs (>1 cm) can lead to dysphagia. Because most GCTs have a low risk of malignancy (<2%), and most patients are asymptomatic, the management approach is controversial. ⁽⁴⁾

While the prevalence of esophageal GCTs is unclear, there is a study that showed that these tumors account for less than 1% of all benign esophageal tumors, and most patients were diagnosed by upper esophagogastroduodenoscopy (EGD) while going under the procedure for other indications. ⁽⁵⁾

In this report, we present a case of a 26-year-old female who was found to have an esophageal GCT while being screened for endoscopic abnormalities before intragastric balloon placement.

CASE REPORT

A 26-year-old obese female, not known to have any chronic conditions, presented to the endoscopy unit for assessment before possible placement of an intragastric balloon. The patient had no history of dysphagia, abdominal pain, weight loss nor any other gastrointestinal complaints. She also denied alcohol consumption, smoking, and drugs.

The patient was vitally stable before the procedure, and her physical examination was unremarkable.

Her laboratory findings a hemoglobin level of 11.7 g/dL, hematocrit of 35.5%, white cell count (WBC) of $5.8 \times 10^9/L$, and a platelet count of $330 \times 10^9/L$. Her

international normalized ratio (INR) was 1.08, random blood sugar (RBS) level 4.37 mmol/L, urea 2.16 mmol/L, and creatinine 78.65 mmol/L.

When EGD was done, it showed a medium size lesion around (1 cm), yellow-colored, non-obstructing, non-circumferential lesion in the middle third of the esophagus (26 cm from the incisors) (Figure-1). No other lesions or masses were found in the stomach nor duodenum, but a diffuse nodular mucosa was found in the antrum and body of the stomach. Histopathology of the gastric biopsies showed evidence of *Helicobacter pylori*.

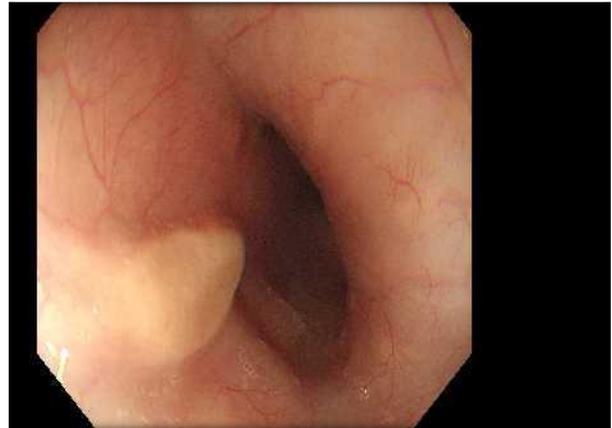


Figure1: Endoscopic picture of around 1 cm, yellow colored, non-obstructing, non-circumferential lesion in the middle third of the esophagus.

The EGD was followed by an upper gastrointestinal endoscopic ultrasound. The esophageal mass that was found earlier was isoechoic, and there was sonographic evidence suggesting local invasion into the deep mucosa (layer 2) (Figure-2). Otherwise, there were no significant endosonographic abnormalities in the stomach, examined duodenum, liver nor pancreas

After close endoscopic and sonographic examination of the lesion, preparations for mucosal resection (EMR) under chromoscopic magnification were started. First, methylene blue was used to mark the borders of the lesion and was injected along with saline to raise it. Mucosal resection with a snare was then performed. A 10 mm area was resected, and resection and retrieval were completed with no bleeding nor other immediate complications.



Figure 2: Endoscopic Ultrasound showed isoechoic lesion in the second layer with sonographic evidence suggesting local invasion into the deep mucosa.

The specimen was sent for histopathological evaluation which showed a partly circumscribed sub-epithelial nodular lesion composed of sheets of uniform epithelioid cells with abundant granular cytoplasm and small nuclei. S100 immunostain showed diffuse positivity. Consistent with a likelihood of granular cell tumor. However, there was no necrosis, nuclear polymorphism, nor significant mitotic activity (Figure-3, 4).

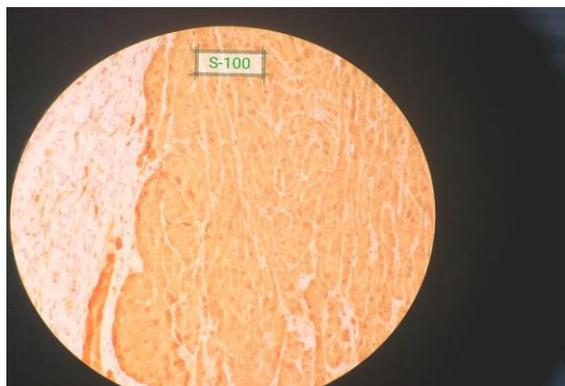


Figure 3 : S100 immunostain showed diffuse positivity.

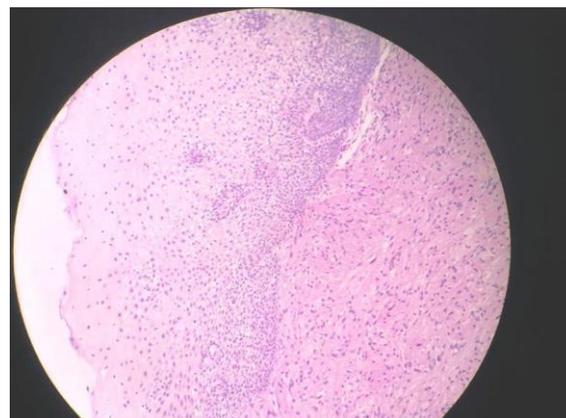


Figure 4: Histologically the lesion composed of sheets of uniform epithelioid cells with abundant granular cytoplasm and small nuclei.

EGD was repeated 10 months after resection and showed a completely normal endoscopic examination of the esophagus.

DISCUSSION

Esophageal GCTs are rare neoplasms that originate from the nervous system. Until now, controversy is still surrounding their diagnosis and treatment, and no clear recommendations have been proposed in recent years.

Most of the reported cases of esophageal GCTs are found incidentally upon EGD screening for other indications such as assessment before bariatric procedures, as was the case with our patient. ⁽⁶⁾ By endoscopy, GCTs look yellowish or gray to white color lesions ⁽⁷⁾, and frequently elevated flat lesion with a smooth surface, which commonly involve the submucosa. Similar to leiomyomas, GCTs can involve the mucosa and muscularis propria. However, rarely larger tumors can be associated with necrosis and ulceration. ⁽⁸⁾

On endoscopic ultrasonography, GCTs arise from the submucosal layer (layer-2) and usually appear as isoechoic lesions. This differentiates them from other benign submucosal esophageal tumors such as leiomyoma, which appear more hyperechoic. Therefore, the use of high frequency ultrasound probes is useful to distinguish between these two conditions. In this context, Dong Kim *et al* (2010) published a series that consisted of 41 patients with GCTs or submucosal leiomyomas and underwent EUS before histologic confirmation. The authors concluded that high-frequency EUS probe helps

to differentiate esophageal GCTs from submucosal leiomyomas.⁽⁹⁾

Because of the rarity of GCTs, there are no clear international guidelines of how to approach these kind of tumors either conservatively, endoscopically, or surgically based on the size of tumor and symptoms. Patients with tumors less than 1 cm in diameter can be treated by conservative approach with periodic endoscopic follow-up if no evidence of malignancy seen on biopsy. In cases where the tumor is greater than 1 cm, and there is no sonographic evidence of invasion of the muscularis propria,⁽¹⁰⁾ it can be removed by endoscopic mucosal resection (EMR) or endoscopic submucosal dissection (ESD). Otherwise, surgical options should be considered in the advanced malignant lesion and when there are any contraindications for ESD/EMR.

We followed up our patient clinically and reexamined her endoscopically 10 months after resection. As stated above, the patient remained well and

asymptomatic with no evidence of recurrence by endoscopy. Similarly, Ramin Niknam *et al* (2017) published a report of an esophageal GCT sized 10-20 mm, and treated by endoscopic resection. They concluded that endoscopic treatment can be safely scheduled for the tumors with size 10–20 mm.⁽⁶⁾ Nevertheless, consensus recommendations and guidelines are lacking on the treatment and follow-up of this kind of tumors.

Ethics approval and consent to participate

The patient provided written informed consent to accept all endoscopic procedures, after receiving information regarding the treatment, risks and benefits.

Consent for publication

Written informed consent was received from the patient for publication of the data in this case report.

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