Multifocal granular cell tumor of the esophagus: A case report

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A 35-year-old male patient who complained of dyspepsia underwent an upper gastrointestinal endoscopy which revealed two yellowish, smooth-surfaced polypoid lesions located at the 36. cm (10 mm) and 38. cm (7 mm) of the esophagus. The lesions were diagnosed as granular cell tumor (GCT) on histologic examination. Endoscopic snare polypectomy was performed after submucosal injection of adrenalin and isotonic saline for large one and two polyps were removed successfully without any complication. Granular cell tumors are rare tumors of neural origin usually located in the dermis and subcutis of the thoracal skin and tongue. The involvement of the gastrointestinal tract accounts for 5% to 9% of the cases, one third of all occurring in the esophagus. Within the esophagus the most common location seems to be distal. Although they are usually asymptomatic and measuring less than 2 cm in greatest diameter, the most common complaints are dysphagia, abdominal pain and chest pain if symptoms do occur. In the literature, there is no agreement in the therapy of esophageal GCT despite the risk of malignancy, although a rare occurrence. In the English literature, the second case of a multifocal GCT of the esophagus treated successfully by endoscopic resection is presented here. [Turk J Cancer 2002;32(3):116-122]

Key words: Granular cell tumor, esophagus, multifocal, endoscopic resection, therapy

Granular cell tumors (GCTs) are rare tumors of neural origin usually located in the dermis and subcutis of the thoracal skin and tongue. They are usually found as single, painless nodules in those areas. The involvement of gastrointestinal tract occurs in 5-9% of cases, mainly in the esophagus (1,2). Approximately 200 cases of esophageal GCT were reported in the English literature. GCT involves usually the distal part of the esophagus as a single lesion in an asymptomatic manner. In the literature, there is no agreement in the therapy of esophageal GCT despite the risk of malignancy, although a rare
occurrence. We present here a multifocal GCT of the esophagus treated successfully by endoscopic resection.

Case Report

A 35-year-old male was referred to us with the diagnosis of esophageal polypoid lesions found at endoscopy performed due to his dyspeptic complaints during previous 6 months. There was no history of dysphagia, weight loss, reflux episodes, long-term drug usage or a known major illness. Biopsies of lesions were reported as granular cell tumor.

Physical examination and routine biochemical tests revealed no abnormality. At computed tomography, there was no lymphadenopathy or esophageal wall thickness. Repeated endoscopy revealed two polypoid lesions located at the 36. cm (10 mm) and 38. cm (7 mm) of the esophagus which were yellowish in color and with a smooth surface without any bleeding or ulcer (Figures 1 and 2).

During endoscopic examination a mild duodenitis and Helicobacter pylori negative antral gastritis were detected as additional findings. We performed polypectomy with snare after submucosal injection of adrenalin and isotonic NaCl (6 cc) for large one and two polyps were removed successfully without any complication at the same session (Figure 3).

Fig 1. The first esophageal polypoid lesion at the 36.cm (10 mm in width) which were yellowish in color, with a smooth surface and without any bleeding
Fig 2. Removal of the second polyp (at the 38 cm) with a snare

Fig 3. The appearance of esophagus just after polypectomy without any complication
Fig 4. Polypectomy specimen shows a polypoid lesion composed of granular cells covered by hyperplastic squamous epithelium mimicking squamous cell carcinoma (H&E, x115)

Fig 5. Granular cells with anti-S-100 positivity on immunohistochemical examination (ABC, x460)
The patient was put on lansoprazole 30 mg bid and sucralfate 15 cc bid for 10 days. Approximately one month after polypectomy, he underwent third endoscopy which showed a small scar at the distal polyp localization while a smooth, normal appearing mucosa at the proximal site. Pathologic examination of excisional biopsy was also granular tumor with pseudoepithelomatous changes mimicking squamous cell carcinoma (Figures 4 and 5). The patient remained asymptomatic and the last endoscopy revealed no abnormality at 1 year.

Discussion

The granular cell tumor is usually a benign entity which is rarely diagnosed without a histologic examination. The first reported case occurred in the tongue and was defined as granular cell myoblastoma in 1926 by Abrikosoff, who believed in its muscular origin (3). Recent findings have suggested that this tumor is of schwannian origin, while it characteristically reacts with S100 and neuron-specific enolase, both Schwann cell markers (4).

Malignant transformation is rare in the context of GCTs. There are fewer than 30 reported cases of malignant granular cell tumor (1-3%) including various parts of the body (5). Only 8 of malignant cases (4%) arised from esophagus in a total of approximately 200 reported cases of esophageal granular cell tumor in the literature (6-8). The involvement of the gastrointestinal tract accounts for 5% to 9% of the cases, one third of all occurring in the esophagus. Within the esophagus the most common location seems to be distal. Although they are usually asymptomatic and measuring less than 2 cm in greatest diameter, the most common chief complaints are dysphagia, abdominal pain and chest pain if symptoms do occur.

Biopsy of smooth, submucosal lesion is frequently unsatisfactory and non-diagnostic in 50% of the cases when it demonstrates only fragments of covering squamous epithelium. However, the opposite is also true when some degree of acanthotic hyperplasia of the epithelium overlying the tumor is present, which could be incorrectly interpreted as a well-differentiated squamous carcinoma (8,9). So there must be a good cooperation between the endoscopist and the pathologist to correctly diagnose this rare entity. Moreover, Palazzo (9) described the endosonographic features of esophageal GCTs as tumors less than 2 cm in diameter in 95% of cases, arising in the inner layers in 95% of cases usually as second echo-poor layer or third echo-rich layer in a minority. He found only one transmural malignant infiltration among 21 cases of esophageal GCTs.

About 90% of esophageal GCT cases appear as single tumor while two or more lesions were found in remaining 10% (8). There are approximately 30 reported cases of multifocal granular cell tumors involving the esophagus. Most multifocal GCTs are synchronous, while only a minority of cases are metachronous. Although it can occur at any age, the average age of patients at diagnosis is 45 years, without a sex predominance. Most multiple GCT of the esophagus involved the distal part decreasing in frequency towards proximally (10-12).

In the literature, there is no agreement in the therapy of esophageal GCT despite the risk of malignancy although rare. Orlowska et al. (8) in the largest
series about esophageal GCT suggested a conservative approach after reviewing the literature and recommended surgical or endoscopic therapy only for large or symptomatic lesions. The traditional procedure was represented by surgical local excision, but new therapeutic options, all with successful results including laser, snare polypectomy, injection of dehydrated alcohol and multiple biopsies with injection of polidocanol (10,13-16) have been proposed. Laser treatment is an expensive technique and not always available. Excision and debulking by biopsy forceps is cheaper and more readily available but perfect removal could not be obtained in all cases and may necessitate multiple endoscopic and histologic examinations. The diathermy snare polypectomy is an effective and largely used procedure throughout the world for gastrointestinal polyps with some modifications in special cases (17,18). Due to potential risk of perforation in the therapy of submucosal tumors like GCTs, submucosal injection and endosonographic examination should be done before polypectomy especially for larger (>2 cm) tumors. Esaki et al. (10) described a patient with four synchronous esophageal GCT treated by snare polypectomy. Herein, we have presented 2nd time in the literature the endoscopic removal of multifocal esophageal GCTs by the same method.

In conclusion, GCT should be kept in mind as a diagnosis of esophageal polypoid lesions in a middle-aged patient, which may be single or 2-3 in number, yellowish, smooth surfaced nodule(s) and usually as asymptomatic presentation. Complete endoscopic excision seems as a logical way in small sized lesions, but endoscopic USG may be performed if available for large lesions before polypectomy.

References


