
Granular Cell Tumor Of The Esophagus: An Unusual Cause Of Dysphagia And Management Dilemma

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Summary

We report an uncommon case of dysphagia caused by a granular cell tumor in a 38 year old black female. Previously documented granular cell tumors are reported as being small and treated endoscopically. This is probably the largest reported in literature and possibly the first documented in the West African subregion. She required excision via thoracotomy and end to end anastomosis of the residual esophagus. Some management dilemmas are also discussed. We conclude that granular cell tumor of the esophagus should be considered in any patient presenting with dysphagia associated with a large esophageal mass.

Introduction

In our environment, the common causes of dysphagia include caustic strictures, achalasia and malignant strictures of the esophagus. Since these diagnoses are often initially considered, other rarer causes are often overlooked. These include benign tumors of the esophagus such as leiomyomas, gastrointestinal stromal tumors, schwannomas, lipomas and granular cell tumors. Of these, leiomyomas are the commonest accounting for more than 80% of cases (1).

Granular cell tumors, referred to as granular cell myoblastomas previously, because they were thought to arise only from skeletal tissue, are even more uncommon. Probably, only about 200 cases have been reported in world literature (2). These tumors are also known to occur in other sites of the body including the skin, breast, respiratory tract and biliary system, with less than 10% of cases occurring in the gastrointestinal tract. The esophagus is the most common site of gastrointestinal involvement (1).

There are still uncertainties as to the actual malignant potential of this tumor as well as the mode for optimal management.

Case Report

We report on a case of a patient presenting with dysphagia due to a granular cell tumor of the esophagus and review the relevant literature.

In May 2008, a 38 year old female elementary school teach-

er presented with 2 years of progressive dysphagia which had become total in the last 2 months with associated weight loss. She had neither ingested a corrosive substance nor had previous esophageal instrumentation. Physical examination revealed a wasted lady with a weight of 45kg and height of 1.65m. Her BMI was 16.53kg/m².

Barium swallow showed proximal and mid esophageal dilatation and a filling defect in the mid-lower esophagus with smooth outlines suggestive of extraluminal compression by a mediastinal mass. (Fig 1)

Contrast chest computerized tomogram revealed a circumferential intramural mass lesion in the mid-distal esophagus with proximal dilatation but no mediastinal lymph nodes. At upper gastrointestinal endoscopy, a non dilatable esophageal stenosis was encountered at 30cm making it impossible to intubate the stomach. The mucosa was however smooth and intact. Esophageal mucosal biopsies were obtained and histology was reported as normal with no malignant cells seen. An impression of esophageal leiomyoma was made and she was planned for excision via a right thoracotomy. At surgery, a 5.5cm x 3.5cm x 2cm creamy whitish mass was seen extending from just inferior to the arch of the azygos vein and completely encircling the esophagus. A partial segmental esophagectomy was performed and end to end anastomosis was achieved.

Histology confirmed the diagnosis of Granular Cell Tumor with no evidence of malignancy (FIG 2). She was dis-

CASE REPORT

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FIG1. Barium swallow showing large esophageal mass

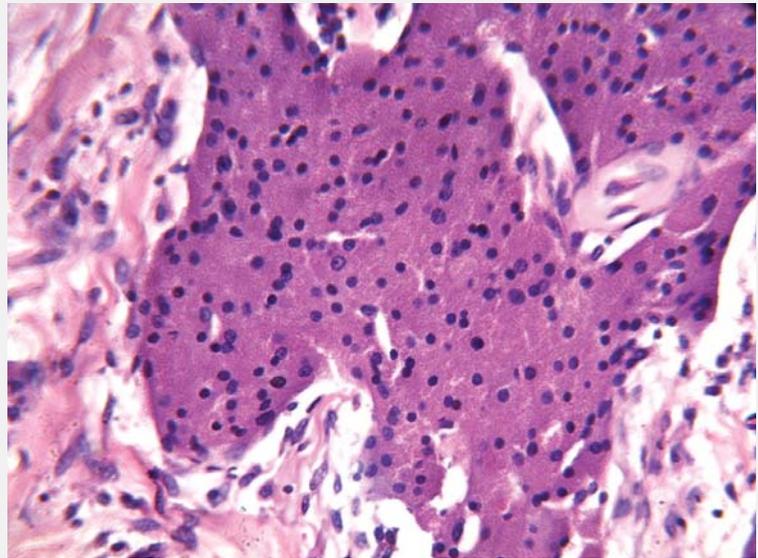


FIG 2. Photomicrograph showing granular cells in esophageal wall

charged home on the 12th post operative day able to tolerate semisolids. She continues to do well on follow up.

Discussion

Granular cell tumors are benign tumors reported to be very uncommon, more so in the gastrointestinal tract. They were first described in the tongue by Abrikossoff in 1926 and have subsequently been described virtually in most organs. Incidentally, it was also Abrikossoff who first described granular cell tumors of the esophagus in 1931. Since then, only about 200 cases have been described worldwide.(2) There remain questions on its malignant potential as well as what constitutes adequate management. This is the first report to our knowledge in the West African subregion.

Although some studies report that a large percentage of patients are symptomatic, these symptoms are mostly not related to the tumor when the diagnosis is made during endoscopy for other problems. Often, these tumors are discovered incidentally. Our patient was very symptomatic due to the huge obstructive effect of the tumor which completely occluded the lumen of the esophagus. It is also interesting to note that this is probably the largest esophageal granular cell tumor ever reported in literature. Most tumors are less than 1cm in its widest dimension making endoscopic polypectomy a viable option.

It is pertinent to note that despite the size of the tumor, it was not possible to make a diagnosis by endoscopic biopsy. This could probably be as a result of superficial mucosal biopsies taken which would not represent the tumor. As it is virtually impossible to make a histological diagnosis prior to endoscopy, the decision to perform an endoscopic biopsy could be debated as this is said to be contraindicated in leiomyomas, for example, because this involves disrupting the mucosal layer and risking secondary infection, bleeding and perforation (3). There are also suggestions that prior biopsy may cause mucosal adhesions and make subsequent enucleation of tumor difficult (4). Endoscopic biopsies, on the other hand, are not contraindicated in cases of granular cell tumors. Therefore, unless there are clear differences in the clinical presentation of these benign tumors to allow for distinction, endoscopic biopsies may be performed inadvertently as was in this case (1). Unfortunately, such differences are not clear cut and no preoperative investigative modality can convincingly differentiate the two except by histology (3).

We therefore recommend that, to forestall such unnecessary and potentially hazardous procedures, endoscopic biopsies should not be performed routinely in these diagnostic dilemmas especially when the mucosa appears normal, except there are indications that the lesion may be mitotic,

as the lesion would subsequently be subjected to histology after surgical excision.

Although there is a shift to more conservative therapeutic approaches, it must be mentioned that this is only possible when the tumors are of a small size as are the majority of less than 1cm. It has been proposed that symptomatology, large size and atypical endoscopic ultrasound findings should alert a physician to the potential for malignancy. (2)The malignant potential of this benign tumor still remains unknown. This will ultimately determine the extent of excision to be carried out or whether these tumors can be left and observed especially when asymptomatic. Overall, there have been only 3 documented cases of malignant esophageal granular cell tumor (2).

We recommend open thoracotomy for excision of such large tumors (>5cm). Despite the size of the tumor excised, it was still possible to achieve intrathoracic end to end anastomosis of the remaining esophagus without undue tension after careful mobilization. The limit of segmental resection and end to end anastomosis of the esophagus possible for benign lesions is not well defined in literature. Short segment resection and esophagogastrostomy are prone to significant gastroesophageal reflux with concerns of intrathoracic leakage of anastomotic lines as well as possible poor blood supply. Hence, it is mostly discouraged

(5). However, though the blood supply of the esophagus is segmental, there still exists a rich plexus of capillaries in the submucosa which supply further than the level of the segment. In addition, the esophagus derives further blood supply from other intrathoracic structures. This may account for the good healing of the anastomosis in this case.

We conclude that granular cell tumor of the esophagus should be considered in any patient presenting with dysphagia associated with a large esophageal mass.

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