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Case Report

Esophageal Gastrointestinal Stromal Tumour: A Case Report

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Abstract: Gastrointestinal stromal tumours (GISTs) represent 0.1-3% of all neoplasms of the digestive tract and only in 1-2% of cases they arise in the esophagus. The most frequent clinical manifestations of esophageal GISTs (E-GISTs) are dysphagia, atypical chest pain, cough or gastrointestinal bleeding. Preoperative study is made by endoscopy, echo-endoscopy, biopsy and computed tomography or magnetic resonance imaging. The preoperative diagnosis is difficult because only histological examination can differentiate them from other esophageal tumours (as leiomyoma, schwannoma and leiomyosarcoma) and rarely a biopsy is performed at a submucosal well-circumscribed esophageal mass. Surgery is the first choice approach for localized and resectable cases. Thoracoscopic or laparoscopic enucleation is sufficient for small-sized and well-capsulated tumours, instead, esophagectomy should be considered in all other cases. Imatinib is the drug of choice for pharmacologic treatment in advanced disease. We report our anecdotal experience of a 63-year old male patient presented at our Unit complaining of dysphagia and underwent transhiatal laparoscopic enucleation of E-GIST.

Keywords: Gastrointestinal Stromal Tumours, Esophageal Neoplasms, Dysphagia, Laparoscopy

1. Introduction

The concept of "gastrointestinal stromal tumour" (GIST) was first introduced in 1983 by Mazur and Clark to describe gastrointestinal non-epithelial neoplasms different from other non-epithelial tumours. [1]

The GISTs are mesenchymal malignancies arising in the gastrointestinal tract from the esophagus to the rectum and account for 0.1-3% of all gastrointestinal neoplasms. [1-4]

They occur in the different tracts with different frequencies: 60% in the stomach, 20–25% in the small bowel, 5% in the rectum and less frequently in the colon, esophagus, appendix, mesentery, omentum and retroperitoneum. [1, 2]

Histologically, the GISTs originate from the interstitial Cajal cells (ICC) that are also called "pacemaker of the gastrointestinal tract" because they are located in the walls of the digestive tract and coordinate their automatic contractions to move ingested food and liquids. At histological examination, the GISTs differ from other gastrointestinal mesenchymal tumours for the expression of CD117 and CD34 and the lack of microscopic evidence of smooth muscle. [1, 5]

In the present paper, we report our anecdotal experience about a case of esofageal GIST (E-GIST), a very rare condition whose management remains still speculative in Medical Literature.

2. Case Report

A 63-year-old male was admitted to our Unit complaining of a 5 months history of dysphagia.

The following instrumental examinations were made: esophagogastroduodenoscopy, chest computed tomography with contrast enhancement, echoendoscopy, esophageal radiography and manometry (Figure 1).

It resulted an intramural esophageal mass (sized 3.5 cm x 3.5 cm) originating from the second layer of esophageal wall (muscularis mucosa) causing a stricture just above the cardia without mucosal involvement. The diagnostic hypothesis was leiomyoma.

Laparoscopic transhiatal extramucosal enucleation was performed as described in the following paragraph. The postoperative hospital stay lasted 4 days and was uneventful; at day 2, after a meal of water-soluble contrast excluding perforations, the patient started to eat.

Histological and immunohistochemical study yielded the following results (Figure 2): GIST with low-grade malignancy (mitotic index <4 mitosis / 50HPG); proliferative index: MIB-1 <4%; CD117: positive; CD34: positive; S100: negative; smooth muscle actin: negative; KI74: positive.

Patient did not undergo chemotherapy because surgery was locally radical and there were no systemic metastases. After 24 months he was disease free.

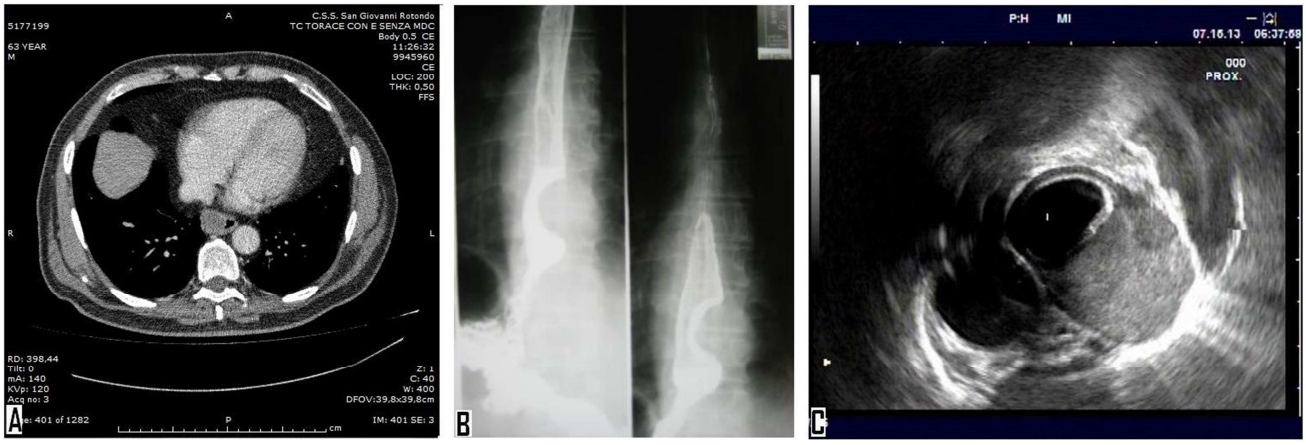

Figure 1. *Preoperative imaging evaluation of E-GIST: A. Transversal sections of chest computed tomography. B. Esophageal radiography (barium meal). C. Echoendoscopy.*

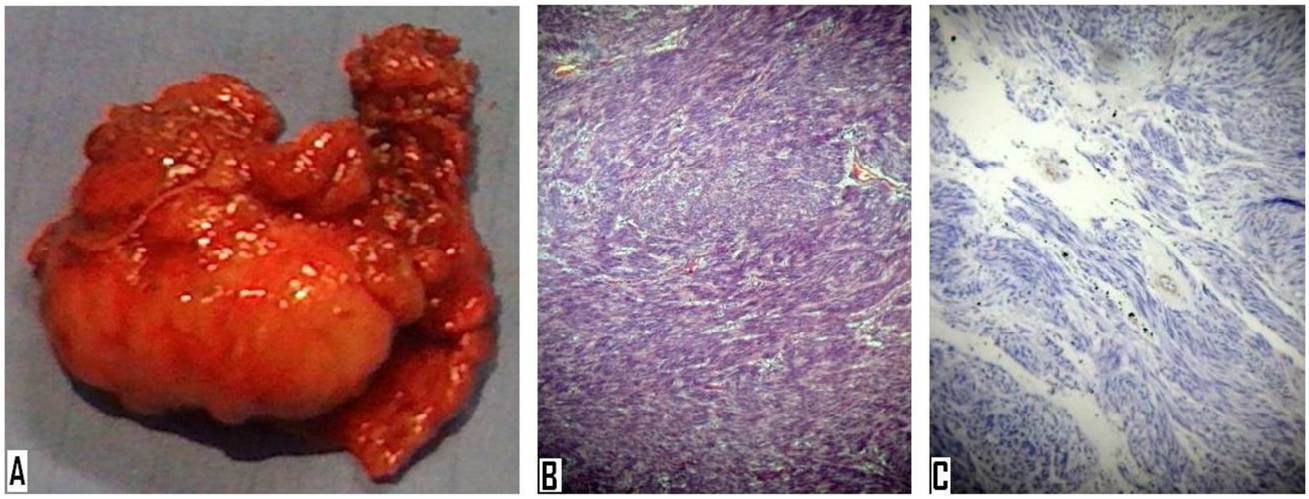

Figure 2. *Surgical specimen and histological study of E-GIST: A. Macroscopic view of the surgical specimen. B. Microscopic view at immunohistochemical study: research of CD117 was positive. C. Microscopic view at immunohistochemical study: research of smooth muscle actin was negative.*

3. Surgical Strategy

The patient signed a detailed informed consent for surgery and was prepared for doing an esophagectomy if the tumour could not be laparoscopically enucleated.

Position of patient. The patient was lying on his back on the surgical table and his legs were abducted.

The surgeon stood in between the patient's legs, the first assistant on his right and the second assistant on his left.

Pneumoperitoneum. A 12 mmHg pneumoperitoneum was inducted by a Veress needle.

Position of trocars. Five trocars were placed in the following way:

The first (11mm) was located to the right of the umbilicus.

The second (11mm) was introduced to the left of the umbilicus.

The third (5 mm) was inserted 3-4 cm below the costal arch

on the right midclavicular line.

The fourth (5 mm) was placed 3-4 cm below the costal arch on the left midclavicular line.

The fifth (5mm) was introduced on left of midline, just below the xiphoid process.

Dissection. By the transhiatal access, it was exposed the anterior and lateral wall of the distal esophagus with the GIST mass. The tumour was not stubbornly adhering to the mucosa so, after the identification of the cleavage plane, it was performed an extramucosal enucleation by blunt dissection utilising a swab mounted on a clamp and taking care to avoid the tumour rupture and the mucosal perforation. A Dor fundoplication was done to protect the wide extramucosal myotomy.

Conclusion. An intraoperative endoscopy confirmed the correct enucleation and the absence of mucosal perforation. After a thorough haemostasis, it was placed a drainage tube near the esophageal hiatus that was removed at postoperative day 2 after a meal of water-soluble contrast.

4. Discussion

The esophageal GISTs (E-GISTs) account for fewer than 2% of all GISTs and their most frequent clinical manifestations are dysphagia, atypical chest pain, cough or gastrointestinal bleeding. [2-4]

The preoperative evaluation is made by endoscopy, echo-endoscopy, biopsy, computed tomography with contrast enhancement and magnetic resonance imaging. [3, 4, 6]

The preoperative diagnosis remains a challenge because E-GISTs share similar clinical, endoscopic and radiographic characteristics with leiomyoma, schwannoma and leiomyosarcoma.

Only histological examination can differentiate them, but rarely a biopsy is performed at an esophageal submucosal mass with endoscopic and echo-endoscopic features of a benign well-circumscribed lesion.

In fact, most of these lesions are leiomyomas and the scarring after biopsy can make difficult the subsequent enucleation. [3, 4]

The standard approach for localized E-GISTs is surgery; the debate about enucleation or esophagectomy remains still open and related to their biologic behaviour. [3, 4]

The biologic potential of these neoplasms is evaluated by tumour size and mitotic index (or mitotic rate). [7]

Tumour size includes 4 groups: very low- (< 2 cm), low- (2-5 cm), intermediate- (5-10 cm), and high-risk (> 10 cm) GISTs. The number of mitoses within 50 high-power fields (HPFs) includes 3 groups: low (< 5), intermediate (5 to 10), or high (> 10) mitotic rate. [7]

Thoracoscopic or laparoscopic enucleation without preoperative biopsy is sufficient for small-sized, confined to the esophageal muscle layer, well-capsulated tumours without mucosal lesions and with low mitotic rate. Preoperative biopsy and esophagectomy should be considered in all other cases. [4, 7, 8, 9]

Even in case of small-sized E-GISTs, an esophagectomy becomes mandatory if there is irregular border, high or intermediate mitotic rate (at histological examination after enucleation or biopsy), inhomogeneous internal echo, heterogeneous enhancement with contrast media, increasing in size. [7, 8, 10]

The prevention of tumour rupture during surgery is an important prognostic factor. [4, 7]

The complete lymphadenectomy does not impact the prognosis of GISTs because of their low local and lymphatic diffusion rate regardless of the size and mitotic index. [7]

The drug of choice for GISTs treatment is Imatinib mesylate; its effectiveness in the treatment of metastatic disease is well documented, instead, its advantages in adjuvant and neoadjuvant therapy remain speculative. [1, 4, 5]

The prognosis of resectable E-GISTs is variable and depends on the size and mitotic index; the overall survival at 5 years after E-GIST diagnosis was reported to be 14%. [1, 3]

Concerning our case, some considerations can be formulate.

In spite of all modern tools for diagnostic imaging, it is always difficult to achieve a preoperative diagnosis of a rare disease without histology. In fact, although our patient presented with dysphagia, a typical symptom of E-GIST, at a first approach other pathologies were hypothesized because of its rarity. The preoperative suspect was a leiomyoma and only postoperative histological examination gave us the correct diagnosis.

Given the small sized mass (3.5 cmx3.5 cm), the regular margins at echo-endoscopy and the absence of mucosal lesions, we chose to attempt a laparoscopic/thoracoscopic enucleation instead of an esofagectomy.

At first, it persisted the doubt to approach the mass thoracoscopically or laparoscopically. Given the proximity of the tumour to the hiatus and the wide experience of our center on surgery of the distal esophagus (for the treatment of gastroesophageal reflux and achalasia), it was considered easier a laparoscopic transhiatal approach, and this preoperative intuition resulted postoperatively correct.

Considering the histological features of a non-invasive tumour, the finding of a low mitotic index (< 4 mitotic / 50HPG), according to medical literature, we excluded to perform an esophagectomy at a later time as radical treatment and avoided chemotherapy. After 2 years the patient is without recurrence.

5. Conclusion

To sum up, E-GIST is a rare condition with many topics still debated about its management.

In fact, the preoperative diagnostic workup, the surgical approach, the extent of resection, the drug therapy remain still speculative. Our anecdotal experience demonstrates the minimally invasive surgery approach to small sized E-GIST is oncologically safe, as long as the mass is completely removed without rupture, the mitotic activity is low, the microscopic margins are negative.

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