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Gastric schwannoma : a case report

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ABSTRACT

We report the case of an 81-year-old female patient in whom a submucosal tumor of the stomach was incidentally discovered, labeled at the outset as a gastrointestinal stromal tumor (GIST), as it was the most frequent and most "obvious" histologic type. However, the pathologist's conclusion was completely different. This experience prompts us to keep in mind the other histologic types of submucosal tumors of the stomach and the contribution of immunohistochemistry to distinguish between them.

Keywords : Schwannoma, gastrointestinal stromal tumors, immunohistochemistry, surgical resection.

INTRODUCTION

Gastric schwannomas (GS) are rare mesenchymal tumors that arise from Schwann cells of the nerve sheath of Auerbach's plexus or less frequently from Meissner's plexus. Most are benign and asymptomatic. The main differential diagnosis of GS is the gastrointestinal stromal tumors (GISTs) that have much clinical, histological and demographic presentation similarities, the contribution of immunohistochemistry is essential to remove confusion. The risk of malignant transformation is exceptional. Surgical resection is the treatment of choice for gastric schwannomas.

MATERIAL AND METHODS

CASE REPORT :

An 81-year-old female patient, with no particular pathological history, admitted to the cardiology department for deep vein thrombosis of the right lower limb. A CT scan performed during her hospitalization fortuitously revealed an oblong and well-defined mass, measuring 7 centimeters,

moderately enhanced, developed in the wall of the antrum and protruded into the gastric lumen suggesting a stromal tumor (figure 1); the CT scan revealed also an asymptomatic gallbladder stone. On the CT scan data, an upper endoscopy was performed to visualize the mass and perform biopsies for pathological examination, the endoscopy objectified a submucosal mass at the level of the greater curvature of the stomac, the gastric mucosa was normal and biopsies of the lesion have been carried out. Histological study of biopsies noted a tumor proliferation consisting of elongated cells with spindle-shaped anisokaryotic nuclei, sometimes hyperchromatic with rare mitosis, associated to polymorphic inflammatory elements. The immunohistochemistry was negative for anti-DOG-1 and c-Kit. The pathologist concluded in a GIST, although DOG-1 and c-Kit were negative.

Surgical resection was decided and performed by laparotomy, the exploration of the abdomen noted a well-limited solid tumor, developed in the antrum along the greater curvature, moreover, there was no liver metastasis, neither peritoneal carcinoma nor suspicious lymph nodes. We performed an antrectomy using 2 linear staplers (figures 2, 3, 4), digestive continuity was realized by a manual end-to-side gastroentrostomy. Postoperative follow-up was simple and histological study of the resection specimen (figures 5, 6) shown tumor proliferation made by short crisscrossed and entangled bundles, bordered by spindle-shaped cells with elongated nuclei, slightly hyperchromatic. The mitoses were rare (3 mitoses / 10 fields). The stroma was small and fibro-inflammatory. The immunohistochemistry was strongly positive for the S100 protein, focally positive for GFAP and negative for c-Kit, DOG-1 and CD 34, which made it possible to retain the diagnosis of gastric schwannoma instead of the GIST initially mentioned.

Figure 1 : CT scan : stomach (yellow arrow), tumor (red arrow).

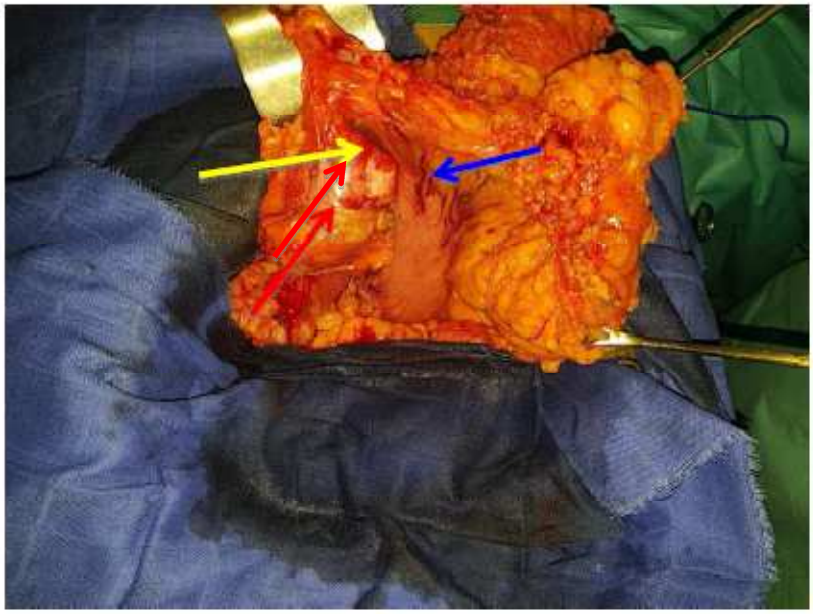

Figure 2: Posterior wall of the stomach (blue arrow), posterior wall of 1st portion of duodenum (red arrow), tumor (yellow arrow).

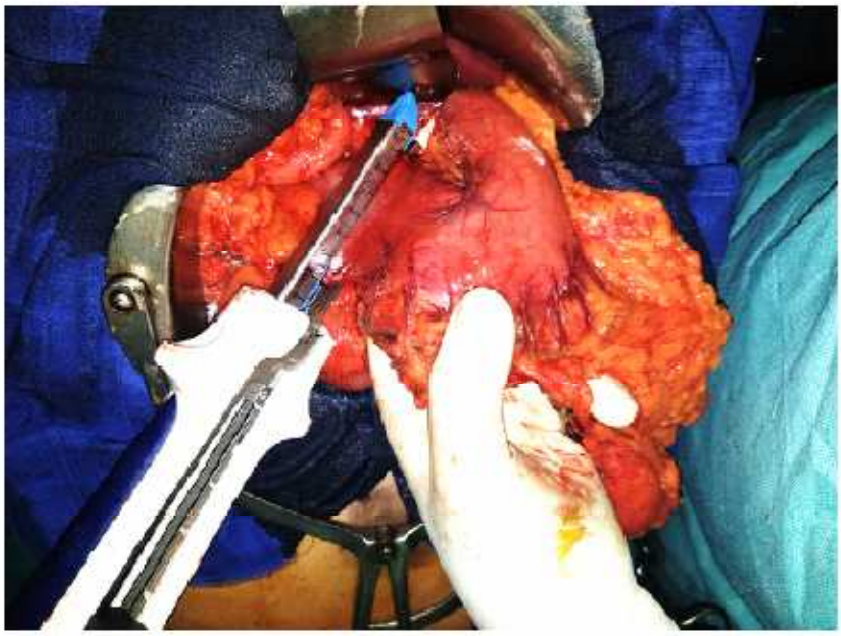

Figure 3 : section of the duodenum downstream of the tumor

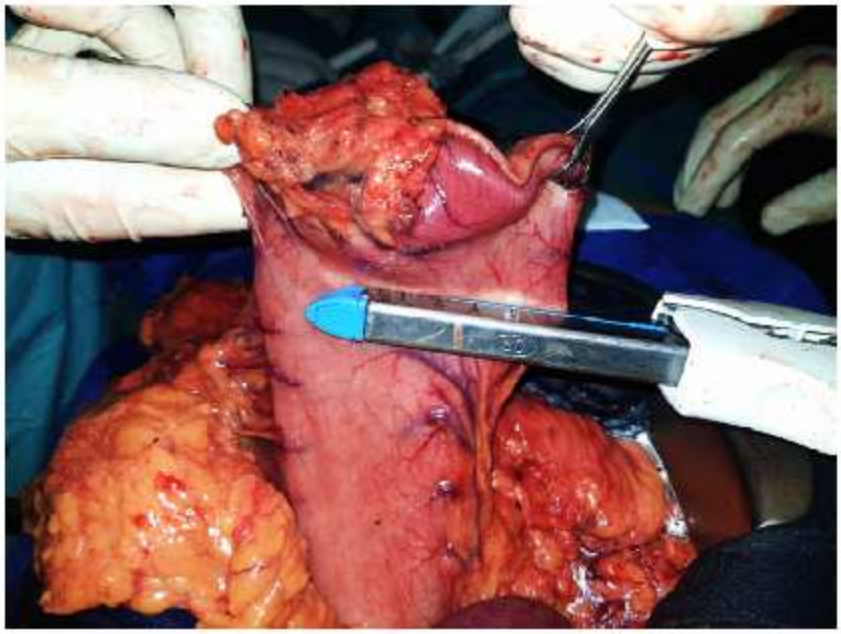

Figure 4 :section of the stomach upstream of the tumor

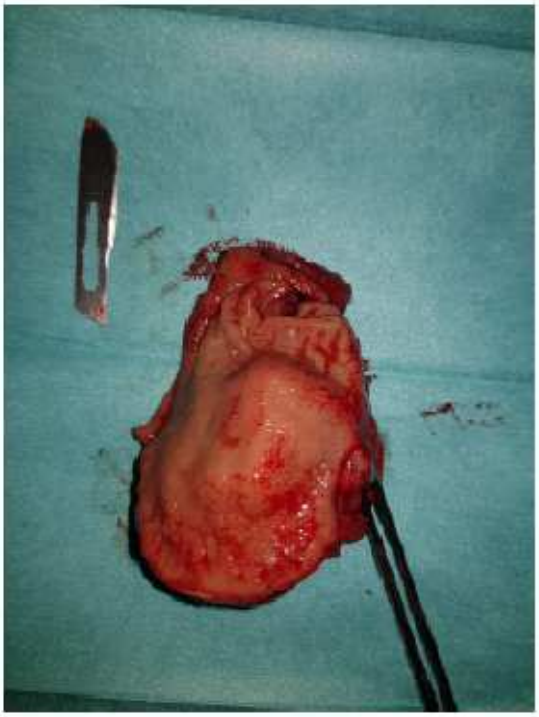

Figure 5 : resection specimen showing the normal aspect of the mucosa raised by the tumor

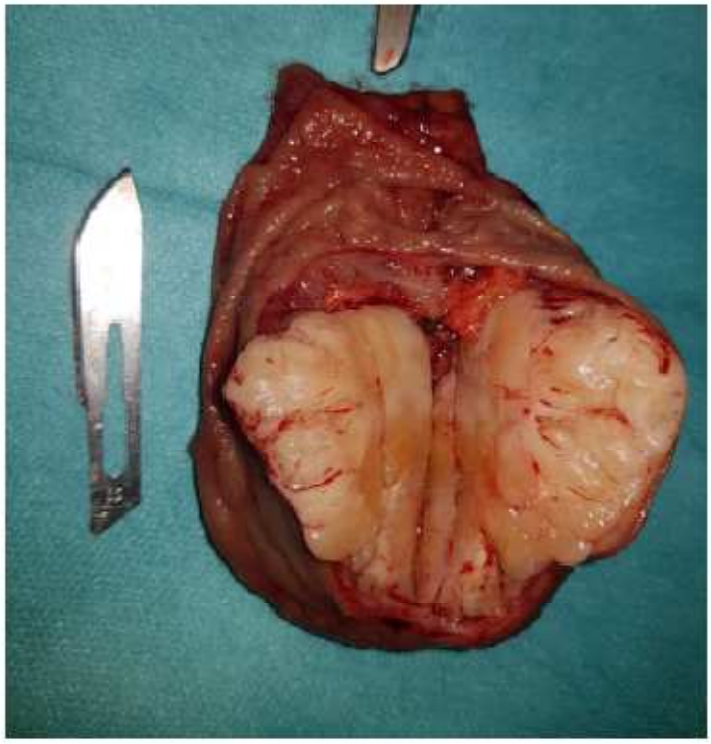

Figure 6 : the aspect of the tumor after the opening of the specimen

RESULTS AND DISCUSSION

GS are extremely rare, only 300 cases approximately have been reported in the literature(1). They represent 0.2% of all gastric tumors(2), 6.3% of gastric mesenchymal tumors, and 4% of all benign gastric tumors(3,4). They are thought to arise from the sheath of Auerbach's plexus or, less commonly from Meissner's plexus. GS tend to appear as single lesions and mostly arise along the lesser curvature.They occur in middle age to late adulthood with a peak in the sixth decade of life. Female predominance has been reported with a male to female ratio of 1: 2.64. Their diameter varies between 0.8 and 15.5 cm with the average of 4.69 cm (5–7).

GS are usually asymptomatic, mostly found incidentally during endoscopy or abdominal imaging exams for other reasons which it was the case of our patient. Symptomatic forms are typically manifested by abdominal pain or discomfort followed by upper gastrointestinal bleeding and less frequently with a palpable abdominal mass, poor appetite, dyspepsia, weight loss, nausea or vomiting (1,5,7).

The main differential diagnosis of GS is GISTs. Since the treatment and prognosis are totally different between these tumors, the correct diagnosis of GS should be done before surgery(8).

In addition to specifying the exact location of the lesions, upper gastrointestinal endoscopy frequently shows protruding submucosal masses with ulcerated mucosa in some cases (5,7). Endoscopic biopsies have limited usefulness since false negatives can occur when the mucosa of the lesion is intact(5). In these cases, fine-needle aspiration biopsy guided by endoscopic ultrasound (EUS) is a reliable diagnostic method (7,9).

On computed tomography (CT), GS usually shows a well-defined, oval, and iso-to mildly hypoattenuated mass compared with the liver on plain CT images, and a homogenous enhancement

pattern on enhanced CT images(10,11). GS rarely develop degenerative changes, such as necrosis and calcification unlike GISTs (12–14).

On magnetic resonance imaging (MRI), GS appear as strongly enhancing tumors, having low to medium signal intensity on T1 weighted images and high signal intensity on T2 weighted images(15).

However, the above-mentioned radiological signs are not specific and can be confused with GISTs even with positron emission tomography with 18- fluorodeoxyglucose (PET-FDG), since they present as hypermetabolic (7,16).

The definitive diagnosis is based on histology and immunohistochemistry. GS grossly appear as solid tumors, generally well-demarcated, without invasion. Most are found in the body, where they vary from small, dome-shaped mucosal nodules to large fungating tumors. Roughly half show central ulceration. Most are confined to the muscularis propria. Their cut surface is yellow- white to tan, glistening, often trabeculated, without hemorrhage, necrosis, or cystic change(17). Histologically, gastric schwannoma shows a fascicular arrangement with spindle-shaped nuclei. The immunohistochemistry is positive for S-100 protein, vimentin and glial fibrillary acidic protein, CD34 positive or negative, and negative for smooth muscle actin, desmin, DOG-1 and c-Kit (CD117); the latter positive in GIST(18,19). It is important to distinguish gastric schwannomas from malignant tumors with positivity for S-100 such as clear cell gastrointestinal sarcoma and metastatic melanoma(6).

GS can rarely progress into malignant tumors of the peripheral nerve sheath (MTPNS), which are characterized by a greater number of mitotic index, presence of necrosis and nuclear atypia(1,5,6). Most of malignant GS reported in literature were associated with neurofibromatosis (20). Surgical resection is the treatment of choice for GS and can be performed both by laparotomy and laparoscopy. Wedge resection, subtotal or total gastrectomy can be done depending on the location and size of the tumor. Lymphadenectomy is not usually performed unless enlarged lymph nodes are seen, since gastric schwannoma rarely metastasizes to lymph nodes(1,5,6). Recurrences are only observed after incomplete resection(20).

CONCLUSION

The rarity of GS and the non-specificity of clinical and imaging signs make that are rarely evoked preoperatively and often confused with other mesenchymal tumors especially GISTs, our observation was a real example. Surgical resection is the treatment of choice and definitive diagnosis is often established on histological and immunohistochemical study of the specimen.

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