CASE REPORT

GASTRIC SCHWANNOMA: CASE REPORT AND LITERATURE REVIEW

ABSTRACT

Aims: Here we present a case of Gastric Outlet Obstruction (GOO) with suspected Gastrointestinal Stromal Tumor (GIST) diagnosed as Gastric Schwannoma (GS) with post-operative histopathology.
Presentation of the case: 31-year-old female presented with complaints of epigastric lump for past 2 years with symptoms of partial GOO for 2 months. Pre-operative investigations were suggestive of a Sub-Mucosal Tumor (SMT), possibly GIST. Intra-operatively an exophytic, 10x10 cm tumor was seen arising from the antrum of stomach. A Distal Gastrectomy with reconstruction in the form of Roux-en-Y Gastro-Jejunostomy was done. Post-operative histopathology was suggestive of GS. The tumor cells were negative for CD-117 and CD-34. They tested positive for S-100.
Discussion: Schwannomas are rare, benign, slow growing tumors of the Gastrointestinal Tract (GIT). In the GIT they are most commonly seen originating from the nerve plexus in the submucosa of stomach. They comprise only 0.2% of all gastric tumors with noted female preponderance. They present with non-specific complaints and lack discrete characteristics on imaging which makes it difficult to distinguish them from other SMT's such as leiomyoma and GIST. Histopathology with IHC for S-100 protein remains the gold-standard for diagnosis.
Conclusion: Not all SMT's arising in the stomach are GIST's. GS although a rare entity entails a much better prognosis without need of any peri-operative adjuvant or neoadjuvant therapy.

Keywords: Stomach, Gastric Schwannoma, Submucosal tumors

1. INTRODUCTION

Schwannomas are tumors arising from Schwann cells that form the myelin sheath around peripheral nerves. Although schwannomas can develop anywhere along the course of peripheral nerves, they most commonly occur in the head and neck region, and are rarely found within the GIT [1]. Within the GIT they are most commonly found in the stomach [2].
GS arise from the Schwann cells forming the submucosal neural plexus in the stomach. They account for about 0.2% of all gastric tumors [2]. Their occurrence is relatively rare with limited evidence available in English literature. They are predominantly benign tumors and bear an excellent prognosis following surgical resection.
With limited differentiating features of these tumors from other SMT and low prevalence of GS a preoperative diagnosis is rarely made or rather they are often misdiagnosed as GIST, a more common SMT arising from the stomach.

We present here a rare case of a 31-year-old female who underwent laparotomy for resection of a suspected antral GIST pre-operatively but diagnosed to be GS with a pathology report.

2. PRESENTATION OF THE CASE

A 31-year-old female consulted in our OPD with complaints of awareness of lump in the abdomen for the past two years, with dull aching epigastric pain and occasional episodes of retention vomiting for the past two months.

On examination she had a firm, nontender, well defined 6x7cm lump in the epigastrium and right hypochondrium with a smooth surface moving with respiration.

Ultrasound (USG) Abdomen revealed a hypoechoic lesion of size 85x71x83 mm in the right hypochondrium originating from the gastric pylorus, closely abutting the 2nd part of duodenum. Upper GI endoscopy revealed a bulge in the stomach with a smooth intact overlying mucosa causing significant luminal narrowing. Scope was not negotiable beyond the constriction. A preoperative Contrast Enhanced Computed Tomography (CECT) Abdomen (figure 1) was done to visualize the extent of the tumor revealed a 6.5x7.8x7.3 cm exophytic homogenously enhancing mass lesion in antro-pyloric region causing significant luminal narrowing with maintained fat planes with the anterior abdominal wall, segment VI of liver and hepatic flexure laterally and the major vessels posteriorly with multiple enlarged peri gastric lymph nodes. An USG guided percutaneous Fine Needle Aspiration Cytology (FNAC) done before the patient presented to us revealed large spindle shaped cells with high mitotic activity. However, validation of the same couldn't be done in our setup. With all the above findings, diagnosis of Gastric GIST was made and she was taken up for surgery.

On laparotomy, an exophytic mass of size 10x10 cm was noted arising from the antrum of the stomach with multiple enlarged peri-gastric lymph nodes (figure 2). An intra-operative frozen section of the lymph node was sent in view of a possible gastric lymphoma and was eventually reported as reactive hyperplasia. Distal gastrectomy was performed with en-bloc removal of tumor along with the enlarged lymph nodes with reconstruction by Roux-en-Y Gastrojejunostomy.

She had an uneventful post-operative course and was discharged by post-operative day 7 on routine oral diet.

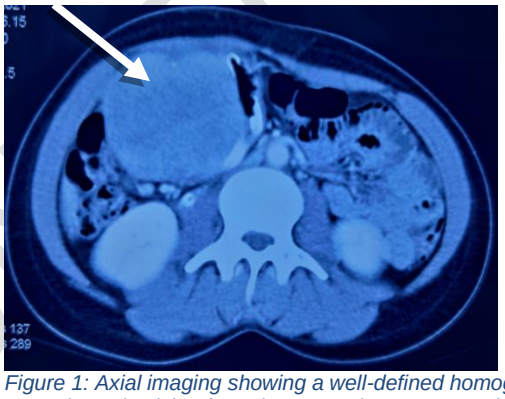

Figure 1: Axial imaging showing a well-defined homogenous mass (arrow) arising from the stomach antrum causing luminal narrowing with enlarged peri gastric lymph nodes

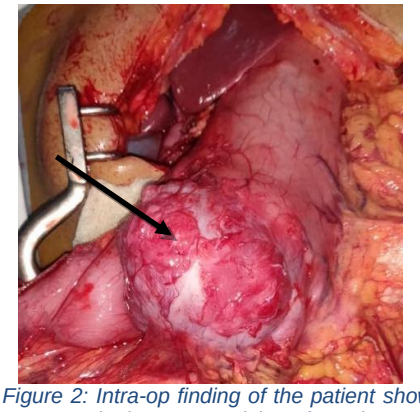

Figure 2: Intra-op finding of the patient showing an exophytic mass arising from the antrum of stomach (arrow)

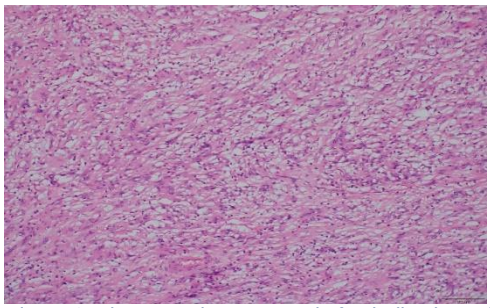

Figure 3: The tumor is composed of spindle shaped cells with bland nuclei (Hematoxylin and Eosin, X200)

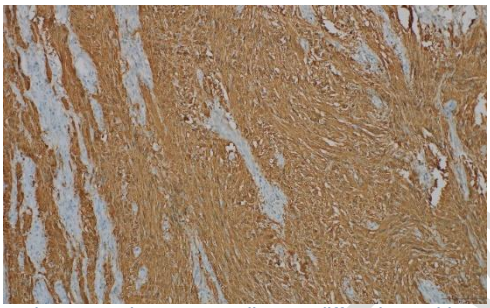

Figure 4: The tumor cells are diffusely positive for S100 (Immunohistochemistry, X200)

Pathology:

- Gross: White in colour, firm in consistency with areas of whorling without any area of necrosis or hemorrhage.
- Macroscopic: Tumor protruding into gastric lumen but not invading the mucosa. Overlying mucosa normal.
- Microscopic: Spindle cell shaped tumor with admixed lymphocytes involving muscularis propria reaching up to the submucosa with surrounding lymphoid aggregates (Figure 3). The tumor cells are showing mild atypia, with vesicular chromatin, inconspicuous nuclei and indistinct cytoplasmic borders. Mitotic rate was low (1-2/3 mm2). Occasional verocay bodies were also seen.
- IHC (Figure 5):

CD-117	CD-34	DOG-1	S-100	Desmin	Vimentin
-	-	NT	+	NT	NT

NT= Not Tested

So, with the findings on histopathology and immunohistochemistry the diagnosis of Gastric Schwannoma was made.
She is doing well 6 months post-operatively without any recurrence.

3. DISCUSSION

Daimaru et al first described Gastric Schwannoma in 1988 as a single nodular mass arising from the submucosa associated with excavation of overlying mucosa. These tumors occurred more commonly in the female gender and are usually present in the 5th or 6th decades [3]. Most cases are detected incidentally or presented with non-specific complaints. In the stomach they are most commonly found in the body (50%) followed by antrum (32%) and lastly fundal (18%) [4]. Our case presented in the 4th decade with features of Gastric Outlet Obstruction (GOO) secondary to the mass arising in the antro-pyloric region.
These tumors presenting as submucosal masses with non-diagnostic clinical or imaging features and a spindle cell histology often leads to a misdiagnosis of the more common GIST. Currently, Immunohistochemistry (IHC) of a tissue sample remains the only definitive differentiating finding to definitively diagnose GS pre-operatively [5], but carries the risk of tumor seeding during retrieval of tissue sample. Recent studies have evaluated the role of EUS in differentiating the SMT's. Jung et al reported the homogenous nature of the tumor with marginal halo differentiates GS from GIST [6–9]. However, large-scale studies with comparative analysis are lacking to validate these results.

In contrast to GIST, GS is a predominantly benign tumor with rare instances of malignant transformation or local invasion reported in available literature [10–12]. The cornerstone in their management lies in en-bloc excision of the tumor and bears an excellent prognosis [8].
CECT is the most readily available noninvasive investigation in current medical practice. It therefore becomes imperative to look out for differentiating features on these scans. GIST is associated with a heterogeneous architecture with peripheral areas of enhancement corresponding to viable tumor and central hypodense region suggestive of hemorrhage/necrosis/cyst formation within the tumor [13]. Whereas, GS is usually a homogenously enhancing well encapsulated tumor which may stem secondary to the fact that GS is a much slow growing tumor compared to GIST. Additionally GS is more commonly associated with reactive lymph node hyperplasia. [7,14]. However, much overlap occurs between these findings and no definitive differentiating features are yet known.
The typical features of gastric schwannoma are firm homogenous tumors without areas of necrosis or hemorrhage showing spindle cells arranged in bundles or whorls with a peripheral lymphoid cutoff and may or may not have germinal center formation consistent with the findings of our case [15]. Other than presence of cuff of lymphoid aggregates the histological features also are insufficient to diagnosis GS without IHC staining which on staining positive for S-100 ultimately concludes the final diagnosis.

4. CONCLUSION

This case proves that not every submucosal tumor in the stomach is GIST and even though GS is a rare tumor it should be kept as a differential diagnosis unless proven otherwise and a post-operative histopathology should be followed for further planning of treatment.

CONSENT

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

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