

CASE REPORT

Gastric Glomus Tumour-A Rare Case Report*Nidhi Bansal¹, Arnav Roychoudhury^{2*}*

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Abstract:

Gastric glomus tumour is a rare benign mesenchymal neoplasm composed of modified smooth muscle cells and neoplastic counter part of perivascular glomus body. Commonest site of glomus tumour being distal extremities (Subungual region of the finger tips, palm wrist and toes). Usually benign, may metastasize to liver and cause death; malignant behavior more likely if > 5 cm. In the gastrointestinal tract, stomach is the most common site of glomus tumors and present as submucosal masses that project into the lumen or out onto the serosa. Rare in solid organs – liver, kidney, Gastrointestinal (GI) tract; common GI site is stomach and are usually single tumor. It comprises of about 1% of all gastric mesenchymal tumours. After the 1st description of gastric glomus tumour in 1951 by Key *et al* only few cases have been reported. The correct assessment of the patient optimizes the chance for an accurate pre operative diagnosis and leads to a correct surgical or endoscopic intervention.

Keywords: Carcinoid, Glomus, Mesenchymal

Introduction:

Gastric Glomus Tumour (GGT) is a rare, usually benign mesenchymal neoplasm composed of modified smooth muscle cells a neoplastic counter part of perivascular glomus body. Common site of glomus tumour is distal extremities (Subungual region of finger tips, palm wrist and toes) [1, 2]. Usually benign, may metastasize to liver and cause death; malignant behavior more likely if > 5 cm. In the Gastrointestinal (GI) tract, glomus tumors are

most commonly found in the stomach, and present as submucosal masses that project into the lumen or out onto the serosa [3]. Rare in solid organs – liver, kidney, gastrointestinal tract; common GI site is stomach and are usually single tumour [4,5]. It comprise about 1% of all gastric mesenchymal tumours. After the 1st description of GGT in 1951 by Key *et al*. [6] only few cases have been reported. The correct assessment of the patient optimizes the chance for an accurate pre operative diagnosis and leads to a correct surgical or endoscopic intervention.

Case Report:

A 49 year old male presented with complaints of pain upper abdomen and fullness since 20 days. No lymphadenopathy, no peritoneal mass and no ascites were clinically present. Liver function test were within normal limits. Hematological parameters were within normal limits.

Computed Tomography (CT) scan abdomen showed thickening of gastric antrum. Laparoscopic distal gastrectomy with gastro jejunostomy was done. Upper GI endoscopy showed a submucosal growth over the antro-pyloric region and a diagnosis of GIST was made. Laparoscopic distal gastrectomy with gastro jejunostomy was done. Post operative course is uneventful and patient discharged in satisfactory condition.

Gross Finding:

Distal gastrectomy specimen was received in the department of histopathology in 10% formalin. On cutting open a submucosal well circumscribed tumour measuring 2.0 x 2.0 x 2.0 cm was seen. Cut surface of the tumour appears grey blue with foci of hemorrhage.

Microscopy (H & E):

It showed a well circumscribed cellular tumour in muscularis propria comprising of sheets and nests of cells with round oval nuclei and clear cytoplasm along with many dilated blood vessels. Minimal atypia, rare mitosis and no necrosis were seen. Overlying mucosa is unremarkable (Fig.1, 2, 3). Diagnosis of benign mesenchymal tumour favoring GGT was made and Immunohistochemistry was advised for confirmation and typing of the tumor. Immunohistochemistry was positive for SMA (4+), and negative for CK, CD117, DOG-1 and Chromogranin.

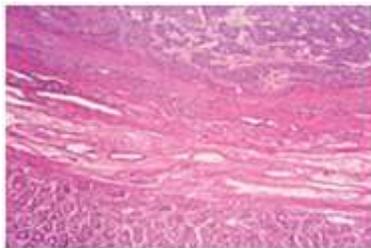


Fig. 1: Normal Gastric Mucosa and Sheets of Tumour Cells (H&E 4X)

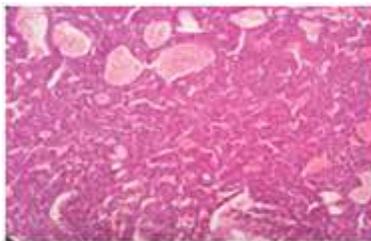


Fig. 2: Trabeculae of Tumour Cells Distributed around Dilated and Ectatic Blood Vessels (H&E 10X)

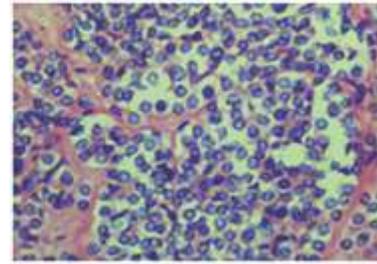


Fig. 3: Tumour Cells are arranged in Sheets and Nests of Cells with Round Oval Nuclei and Clear Cytoplasm along with Many Dilated Blood Vessels (H & E 40X)

Discussion:

GGT is a benign mesenchymal neoplasm arising from the neuromyoarterial glomus. The glomus has also been described as an arteriovenous shunt that may contract or expand [7]. Glomus tumours are commonly observed in the dermis or the subcutis. They have also been described in the bone and joints, skeletal muscle, soft tissue, mediastinum, trachea, kidney, uterus and vagina. The first case of GGT was reported in 1951 by Key *et al.* [6] and since then, few cases have been reported. Vascular tumours of the gastrointestinal tract are rare (accounting for less than 2% of benign tumours), but according to Miettinen *et al.* [3], the frequency of GGT is estimated to be 1% of that of gastrointestinal stromal tumours. Glomus tumours of the stomach have a marked predominance in females [6] though older studies [8] showed nearly equal sex distribution. Moreover, they usually occur in the fifth or sixth decade of life. GGT, rare benign mesenchymal tumour is usually solitary. There is only one case report of multiple GGT [9]. Furthermore, GGT are small and have a greater predilection for the greater curvature of the stomach [8, 13, 14, 16]. The differential diagnosis included epithelioid GIST, paraganglioma and carcinoid tumour. Malignant behavior of the

tumour is difficult to be predicted on histology alone. In gastric tumours, there are no definite predictors of malignancy though they can metastasise. Although glomus tumours of the stomach are usually benign, malignant behavior cannot be excluded. Folpe *et al.* [10] proposed the classification for malignant glomus tumours as: -

- a) deep location and size more than 2 cm or
- b) presence of atypical mitotic figure or
- c) combination of moderate to high nuclear grade and mitotic activity (5 mitoses/50 high-power fields).

It should also be mentioned that the classification criteria have been established for superficial or deep soft tissue glomus tumours. However, due to lack of evidence in the current literature, we suggest that the above mentioned criteria should be used by convention for GGT. Only one case of metastatic GGT has been described [3]. The tumour measured 6.5 cm and on histological analysis mild atypia (1-3 mitoses/HPF) was observed.

Histomorphology of benign GGT is distinctive. Benign glomus tumours consist of small uniform rounded glomus cells that are located in the walls of dilated vessels. The tumour cells have small uniform nuclei, show positive immunoreactivity for smooth muscle actin and are outlined by PAS-positive basement membrane [10]. Glomus tumours are also calponin positive and lack the C-KIT mutation seen with GIST tumours [11].

Immunohistochemistry is essential in the differential diagnosis of glomus tumours. Immunohistochemical staining for actin is negative in gastrointestinal endocrine tumours, but positive in about half of the GISTs. Gastric

epithelioid GISTs are usually positive for C-KIT (CD117) [3]. Leiomyomas and leiomyosarcomas are differentiated from GISTs by positive immunoreactivity for desmin and smooth muscle actin and negative immunoreactivity for C-KIT (CD117) and CD34 [8, 12]. Finally, operative intervention should be carefully planned in cases of submucosal gastric masses. Lymph node metastases were not common. As GGT are mesenchymal tumours with potential malignant behavior, wedge resection with negative margins should be the treatment of choice. Enucleation is not recommended due to the high recurrence rates [15]. GGT should always be included in the differential diagnosis of submucosal gastric lesions, keeping in mind that preoperative investigation of these patients often yields misleading results.

Conclusion:

Preoperative diagnosis of GGT is difficult. Despite their distinct histological appearance, their clinicopathologic, radiology and upper endoscopy features overlap with more common gastric tumours. The diagnostic gold standard for such lesions is the histological examination and the immunohistochemical markers. A multi-faculty medical approach of the patient optimizes the chances for an accurate preoperative diagnosis and leads to a targeted surgical intervention. The correct assessment of the patient optimizes the chance for an accurate pre-operative diagnosis and leads to a correct surgical or endoscopic intervention.

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