

papillary appearance. The difference between the two lies in respect to mucin secretion which is common in IPMN but is found in only one third of IPMN-B cases.^{2,3}

IPMN-B can be graded based on cholangiography and the management protocol is decided accordingly. Our case was cholangiographic type IIB (intrahepatic polypoid or cystic neoplasia with involvement of extrahepatic bile duct); in this situation, an aggressive resection is advised.⁴

The survival of patients with IPNB reduces with progression from low-grade dysplasia to invasive carcinoma. The recurrence rate at 5 years in benign IPNBs has been reported to be nearly 20%; this rises to 60% in malignant cases. Most recurrences are locoregional.⁵

Curative resection is the major treatment and bodes well for long-term survival, especially in patients with early-stage IPNB.

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Multimodality imaging of gastric glomus tumor presenting with upper GI bleed

Glomus tumors are rare tumors of mesenchymal origin. They are composed of modified smooth muscle cells and originate from perivascular glomus bodies.¹ Most are located in the subungual regions. They are rarely found in the gastrointestinal tract where they mostly occur in the stomach.² The presentation may vary from non-specific abdominal discomfort to life threatening upper gastrointestinal haemorrhage.³ Recognition of imaging features helps differentiate this potentially malignant entity from other submucosal tumors.

Case Report

A 35 year old female presented with upper gastrointestinal bleeding. On examination she had pallor and epigastric tenderness. Her blood investigations were unremarkable except for haemoglobin of 7.9gm/dl.

Ultrasound examination revealed a well-defined hypoechoic lesion, in the pre-pyloric region of stomach with a small dimple in the centre of lesion (**Figure 1**) Barium meal study showed a well-defined smooth margined filling defect at the greater curvature in the pre-pyloric region, with obtuse margins and a central dot of barium pooling suggestive of ulceration (**Figure 2**). CT scan revealed a hypodense, polypoid lesion, measuring 4x3 cm in the pre-pyloric region at the greater curvature. There was enhancement in the arterial phase with persistent progressive enhancement in the portal venous phase (**Figure 3**). MRI showed a well-defined submucosal

T1W hypointense and T2W slightly hyperintense lesion with intense enhancement after gadolinium administration (**Figure 4**).

Based on the submucosal location, contrast enhancement and central ulceration a provisional diagnosis of gastrointestinal stromal tumor was made with the differentials of carcinoid, haemangioma and glomus tumor.

Anterectomy with Billroth 1 reconstruction was performed. Histopathology showed that the tumor was arising from the submucosa (**Figure 5**). There were numerous dilated thin walled vascular channels. Sheets of round to oval tumor cells (glomus cells) containing clear cytoplasm were found. Hemangiopericytoma like areas were seen within the tumor with focal epithelioid differentiation of glomus cells and infrequent mitoses. The serosa was free of tumor. Immunohistochemistry showed that the tumor cells were positive for SMA, vimentin and caldesmon and were negative for synaptophysin, chromogranin, CD 34, CD117. Mib - 1 index was < 1%. Based on these findings a diagnosis of glomus tumor was made.

Discussion

Glomus tumors originate in the neuromyoarterial glomus, a normal arteriovenous shunt abundantly supplied with nerve fibres which has a temperature regulating function.⁴

Glomus tumors were first described by Barre and Masson in 1924. They can be found anywhere in the body like bones, joints, skeletal muscles, trachea, kidney, uterus and vagina. The classic location is the subungual region of a digit⁵ followed by palm, wrist and toes. Rarely they are found in gastrointestinal tract where these commonly occur in the stomach as a solitary submucosal nodule in the pre-pyloric region and less commonly in the small intestine.²

Glomus tumors are less common than GIST, with a ratio of 1:100.⁶ Most of the reported cases are localised to the greater curvature side of the pre-pyloric region of stomach.

Endoscopic ultrasound (EUS) findings suggest that these originate from the third and fourth layer of the stomach based on endoscopic classification.^{2,3,7} These

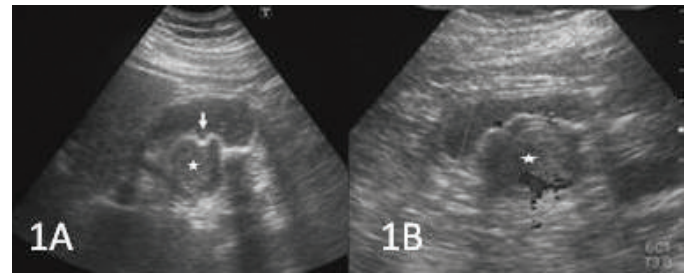


Figure 1: (A and B) Ultrasound scan shows a well-defined submucosal, hypoechoic lesion in the pre-pyloric region of the stomach (white star) with overlying echogenic mucosa showing a small central dimpling suggesting ulceration (white arrow).

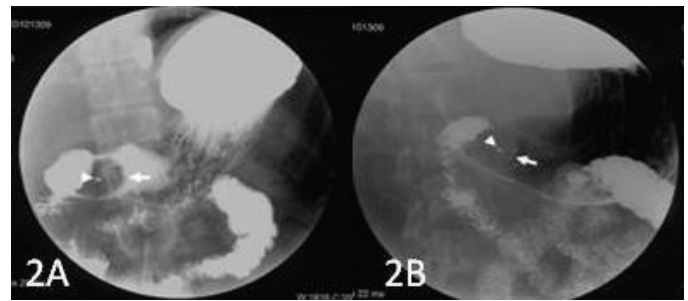


Figure 2: (A and B) Double contrast barium meal film in supine position showing a well-defined sharply margined submucosal lesion in the pre-pyloric region with a (white arrow) central dot of barium pooling suggesting ulceration. (arrow head).

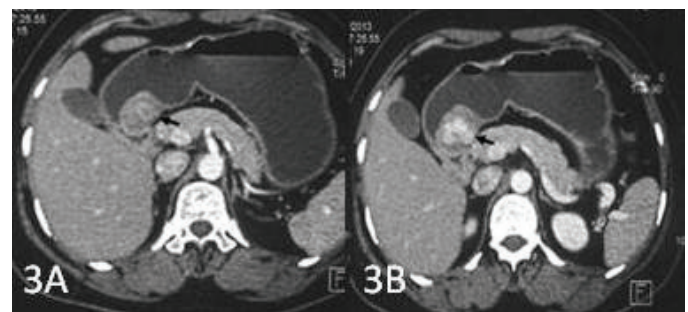


Figure 3: (A and B) Contrast enhanced computed tomography images in axial planes showing a well defined lesion along the greater curvature side of pre-pyloric region of stomach (black arrow) with intact overlying muscularis and mucosal layer, predominantly endophytic growth pattern, with progressive enhancement on arterial and portal venous phases. No tumor calcification or intra-lesional low attenuation area is seen. The surrounding fat planes are clear.

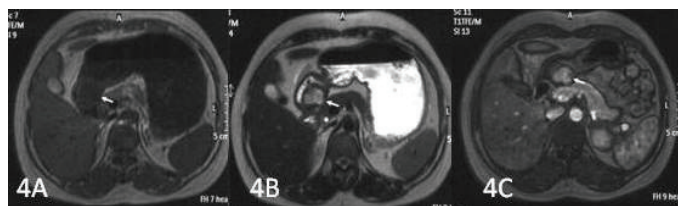


Figure 4: (A, B and C) MRI abdomen. Axial images showing a well-defined submucosal lesion which is hypointense on T1W images, hyperintense on T2W images and show intense enhancement on post gadolinium administration (white arrow). The mucosal layer is hyperintense on T1W and hypointense on T2W images with subtle enhancement on post contrast images.

lesions may be heterogeneous owing to haemorrhage and calcification, and can be confused with GIST or leiomyosarcoma. On power Doppler sonography, they are hypervascular.⁷ On CT, they appear as well-circumscribed submucosal masses and may contain flecks of calcifications. Contrast enhanced study shows intense enhancement in arterial phase persisting in the portal venous phase.^{2,3,7} MRI findings, as first described by Kao-Lang Liu in 2005² are that the tumor is slightly hypointense on T1-weighted images, slightly hyperintense on T2-weighted images and exhibits persistent enhancement after gadopentetate dimeglumine administration.

Other submucosal enhancing tumors include GIST, schwannomas, ectopic pancreatic rests, haemangiomas, carcinoid tumor, and melanoma metastasis. Pre-operative differentiation of gastric submucosal tumors is important to distinguish them from more malignant counterparts for proper surgical management.

The most common malignant counterpart of a glomus tumor is GIST. However GIST's more frequently show cystic change with heterogeneous enhancement, ulceration, necrosis, cavitation, and communication with bowel. Mean age of presentation is 40-70 years and the distribution among both the sexes is almost equal. Lymph node metastasis is less common.²

Definitive diagnosis is possible only on histopathology and immunohistochemistry. Glomus tumors show uniform sized glomus cells located in the walls of dilated vessels with immunoreactivity for smooth muscle actin and calponin, and lack c-KIT (CD 117) mutation (positive in GIST).⁷

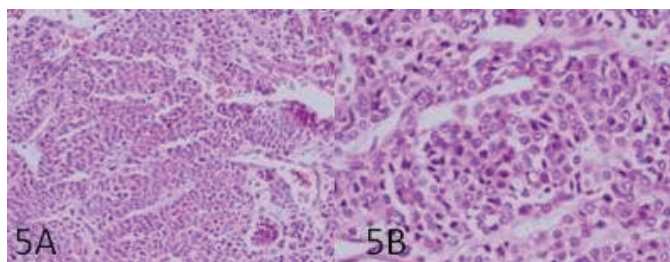


Figure 5: (A and B) Histopathology slide showing perivascular arrangement of tumor cells with round nuclei and low mitotic index. The section shows vascular channels lined by glomus cells admixed with eosinophilic myoid spindle to oval cells. (HE10,40X)

As gastric glomus tumors have potential malignant behaviour, enucleation is not recommended and wedge resection with negative margins should be the treatment of choice.⁷ In case of antral tumors, antrectomy with gastrojejunostomy should be attempted as wedge resection would lead to significant gastric deformity.

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