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# Glomus Tumour in Duodenum: A Pericytic Tumour at Unusual Location

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## 1. Abstract

**1.1. Introduction:** Glomus tumour is a benign neoplasm composed of perivascular modified smooth muscle cells of the normal glomus body. Rarely, it has been reported in the gastrointestinal tract.

**1.2. Case Report:** We present here a case of glomus tumour originated from duodenum, which was found incidentally in imaging. Histopathology examination and immunohistochemical studies confirmed the presence of glomus tumour. This article illustrates a case of glomus tumour at duodenum, with discussion on its differential diagnosis and management approach.

**1.3. Conclusion:** The pathologist needs to be aware of the histology features of glomus tumour. The definitive diagnosis relies on histological assessment. Base on the typical morphology and the help of immunohistochemical study, glomus tumour can usually be confidently diagnosed.

## 2. Introduction

Glomus tumour is a benign neoplasm composed of perivascular modified smooth muscle cells of the normal glomus body. Most of them occur as a soft tissue neoplasm in the distal extremities, such as the subungual region, hand, wrist, and foot [1]. Rarely, it has been reported in the gastrointestinal tract, in which the vast majority arise within the stomach [2]. Glomus tumour arising from the intestine is exceedingly rare, with only a few case reports available in English literature [3-10]. Herein we describe a case of glomus tumour arising in the duodenum.

## 3. Case

A 70 years old Chinese man, with history of colonic diverticulosis, presented with episodes of fresh per-rectal bleeding and haemoglobin drop from 10.6 g/dL to 7.8 g/dL. Proctoscopy showed minimal internal piles. Colonoscopy showed multiple diverticula over entire colon without any mucosal lesion. A diagnosis of diverticular bleeding was made. However, Computed Tomography (CT) scan of the abdomen and pelvis showed an incidental finding of a 1.8cm roundish lesion at the anterior wall of duodenal bulb, in additional to diffuse diverticulosis of colon. The lesion showed early intense arterial enhancement which persisted into portal venous and later phases, and a tiny focus of internal calcification (figures 1 and 2).

Subsequent endoscopic ultrasound was performed to further delineate the lesion, which showed a 2 cm hypo-echoic lesion at the first part of duodenum. It appeared to be arising from muscularis propria layer of duodenal wall with increasing doppler signal and internal calcification (figure 3).

The clinical and radiological impression at that moment was a duodenal gastrointestinal stromal tumour, with other differential diagnoses including neuroendocrine tumour, haemangioma and glomus tumour. The lesion was later excised via laparoscopic approach and sent for histopathological examination.

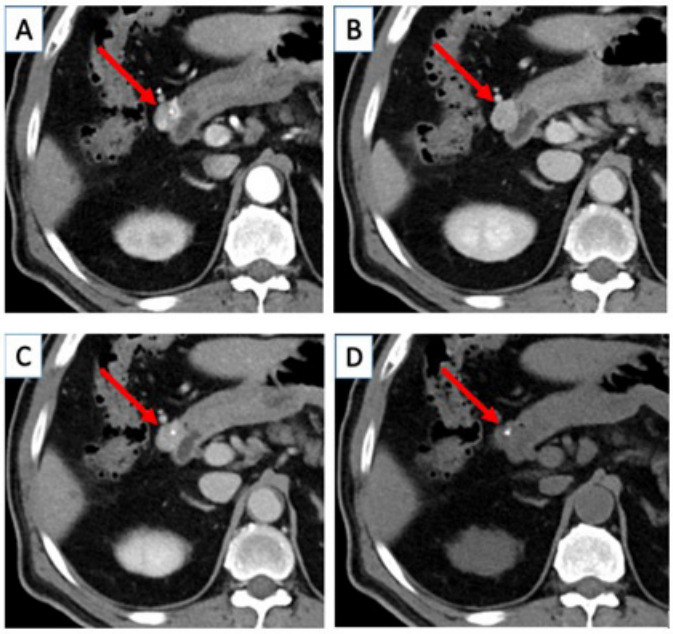



**Figure 1:** Axial contrast-enhanced computed tomography scan showed the duodenal lesion with early intense enhancement in arterial phase (A), persisted enhancement in portal venous phase (B) and delayed phase (C). The tiny focus of internal calcification is illustrated in axial plain computed tomography scan (D).

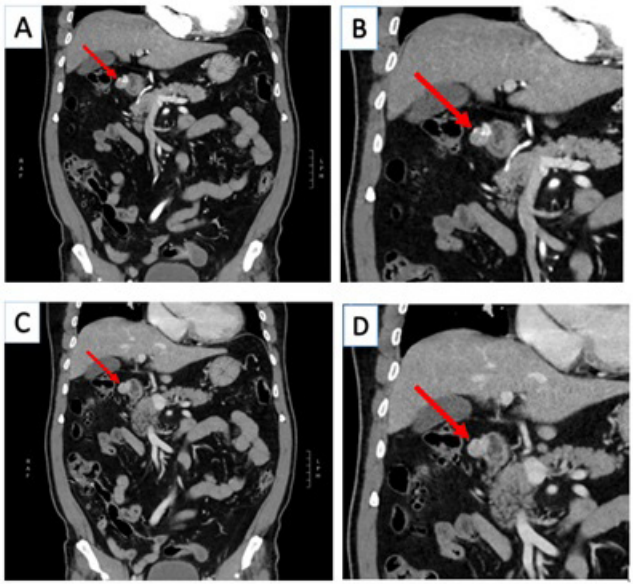



**Figure 2:** Coronal computed tomography (CT) scan demonstrated the lesion in arterial phase (A and B) and venous phase (C and D).

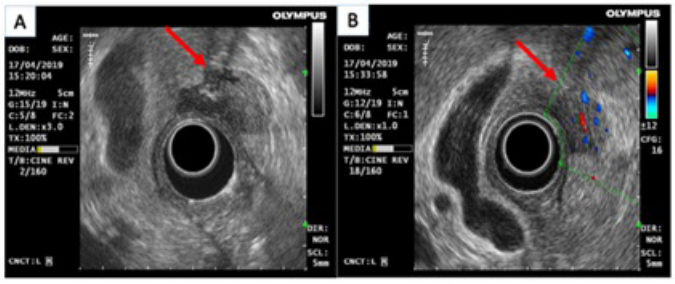



**Figure 3A and 3B:** Endoscopic ultrasound showed a 2 cm hypoechoic lesion appeared to be arising from muscularis propria layer of duodenal wall with increased doppler signal and internal calcification.

### 3.1. Gross Examination

Received was a piece of duodenal wall tissue measuring 2 x 2 x 1 cm in size, with the overlying mucosa measuring 1 x 1 cm in area. A tan colour submucosal nodule was identified on sectioning, measuring 1.7 x 1.5 x 1 cm.

### 3.2. Microscopic Examination

Sections showed duodenal tissue with a well-defined, non-encapsulated lesion arising from the muscular propria. It was composed of tumour cells arranged in nests, cords or isolation, and separated by delicated vascular channels. The tumour cells were roundish with fair amount of eosinophilic cytoplasm, mild hyperchromatic nuclei and inconspicuous nucleoli. Mitosis was not identified. A focus of calcification was present. The overlying duodenal mucosa was unremarkable. Excision appeared to be complete (Figures 4 - 6).

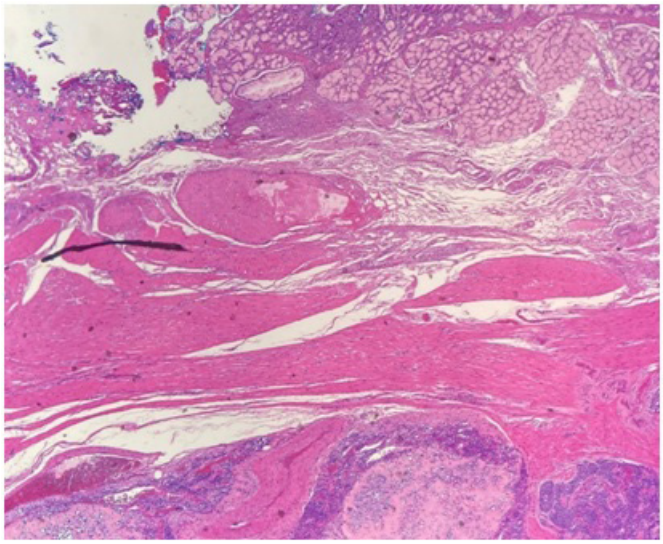


**Figure 4:** The overlying gastric mucosa and the underlying glomus tumour. The tumour showed a pushing boundary (40x magnification)

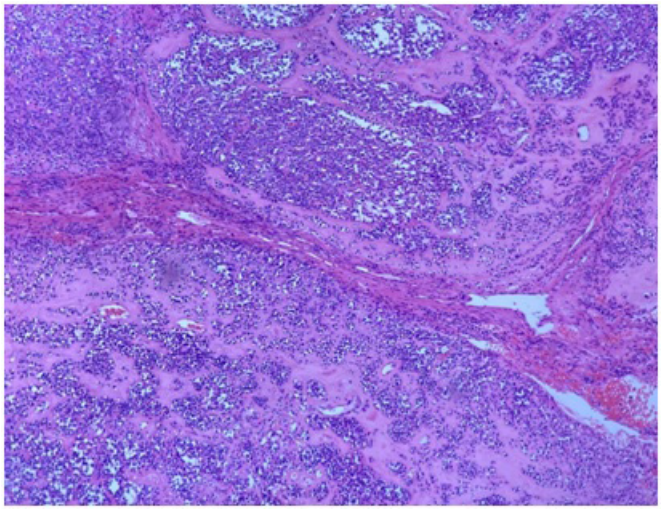


**Figure 5:** The glomus tumour, featuring clusters of bland looking cells. (100x magnificaition)

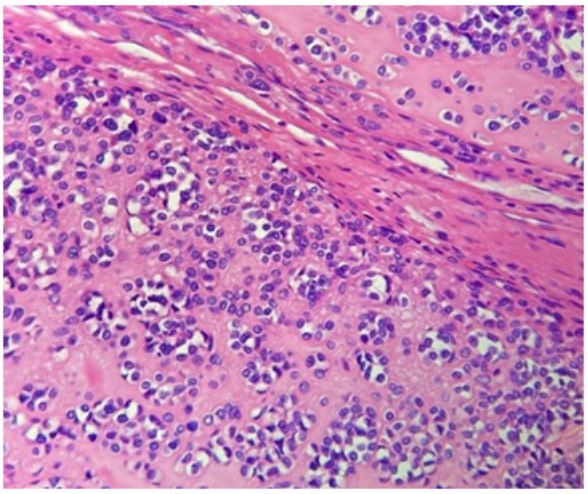
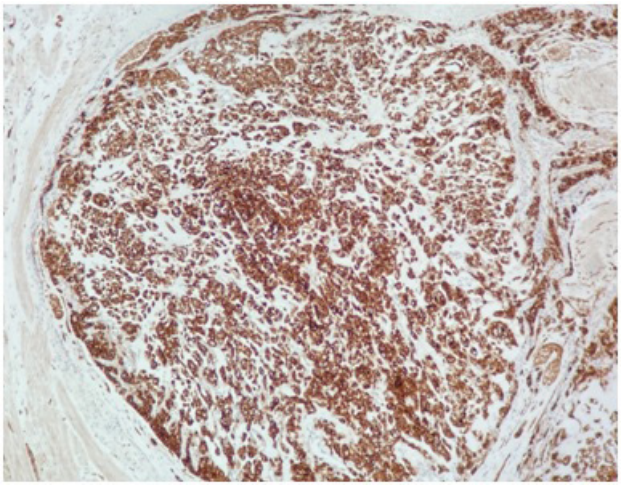
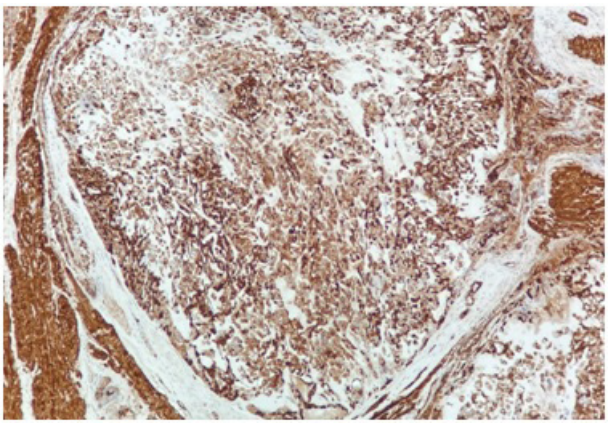


**Figure 6:** Bland looking cells with punch out nuclei. The cells are characteristic and essential for the diagnosis of glomus tumour. (400x magnificiation)

Immunohistochemical study showed that the tumour cells were diffusely positive for smooth muscle actin, caldesmon and synaptophysin. MNF116, Cam 5.2, AE1AE3, DOG1, S100 and c-kit are negative. The Ki-67 labeling index was about 1%.

The overall features were consistent with glomus tumour arising from duodenum.





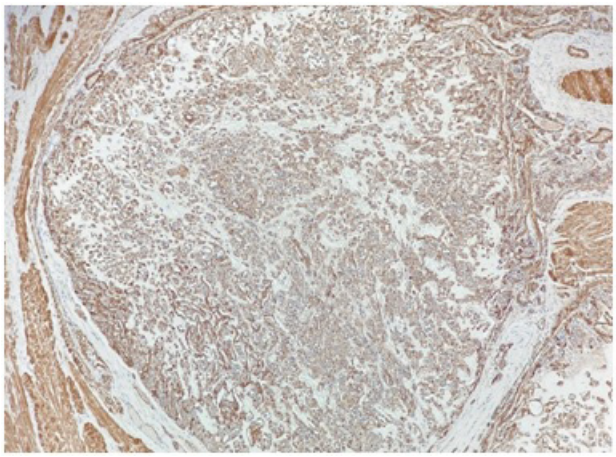


**Figure 7 to 9:** Immunohistochemical study for syanptophysin, h-Caldesmon, smooth muscle actin (100x magnificaition)

### 3.4. Progress

The surgery was uneventful and the patient remained well with no signs of recurrence after 18 months of surgery.

## 4. Discussion

Glomus tumour in the gastrointestinal tract is uncommon, in which the vast majority arise within the stomach. Glomus tumour arising from the intestine is exceedingly rare, making accurate preoperative diagnosis difficult. The differential diagnoses of a hypervascular tumour in contrast-enchanced Computed Tomography (CT) scan include gastrointestinal stromal tumour, neuroendocrine tumour, haemangioma and glomus tumour.

In microscopic examination, the diagnosis is relatively straight forward. Typical glomus tumor involves the muscularis propria, and is composed of uniform, small round cells with a discrete round nucleus and pale to eosinophilic cytoplasm. The cells are arranged in sheets and nests with a rich vascular network. The stroma is typically myxoid or hyalinized. Mitotic activity is usually low. Focal nuclear atypia and vascular invasion can be observed, and vascular invasion does not affect prognosis.

Immunohistochemically, glomus tumour are almost always diffusely and strongly positive for smooth muscle actin. Caldesmon is also positive in more than half of the cases. Focal synaptophysin expression has also been reported [2,11,12]. However, in our case, diffuse and strong positive for synaptophysin is observed. This maybe a potential diagnostic pitfall in the differential diagnosis with neuroendocrine tumour. The importance of the use of a panel of immunostains cannot be overemphasized.

The vast majority of gastrointestinal glomus tumour is benign. Malignant behaviour has been reported in two cases of gastric glomus tumours that metastasized to the liver. Both were larger than 5 cm, and one showed mild atypia, spindle cell foci (1 and 2). No malignant behaviour has been reported in duodenal glomus tumour.

Nevertheless, the criteria for malignancy in gastrointestinal tract are undefined due to insufficient data. The criteria for malignancy for

peripheral soft tissue glomus tumour are deep location and size > 2 cm, or atypical mitotic figures, or moderate to high nuclear grade and ≥ 5 mitoses/10 $mm^2$ [1].

## 5. Conclusion

Herein we describe a Chinese case of glomus tumour arising in the duodenum. An accurate pre-operative diagnosis is difficult. Contrast-enhanced Computed Tomography (CT) scan may show strong arterial enhancement and prolonged enhancement in portal venous phase with a hemangioma-like enhancement pattern [7]. The definitive diagnosis relies on histological assessment. Base on the typical morphology and the help of immunostains, glomus tumour can usually be confidently diagnosed. Glomus tumour should be included in the differential diagnosis list among the submucosal tumours.

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