# Glomus Tumor of Small Intestine: Case Report and Review of Literature

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**Case report**

**Keywords:** Glomus tumor, Small intestine, Case report

# Abstract

**Background:** Glomus tumors are exceedingly rare tumors arising from the normal glomus body. Only a very few cases of glomus tumors located in the small intestine were reported to date. Here, we present a case of glomus tumor of the jejunum in a 30-year-old female patient, and discuss its clinical, imaging, and pathologic features.

**Case presentation:** A 30-year-old female referred to our hospital with the chief complain of melena and fatigue for 3 weeks. Oral single-balloon enteroscopy discovered a 2.0*2.0cm tumor without epithelial lining in the jejunum. The patient then underwent partial enterectomy. HE stain illustrated that the tumor was interspersed with congestive capillaries of various size, and admixed with smooth muscle bundles. Immunohistochemical study showed that the tumor cells were strongly positive for SMA and collagen type Ⅳ. The Ki-67 proliferation index was less than 1% and mitotic activity is very low (about 1/50 HPF). The case was finally diagnosed as benign glomus tumor.

**Conclusion:** Glomus tumors arising from the small intestine are extremely rare. The final diagnosis is made by histological and immunohistochemical examination. Although there is no standardized management pathway for these patients, early diagnosis and treatment are important for a good prognosis.

## Background

Glomus tumors (GTs) account for approximately 2% of all soft tissue neoplasms [1]. However, rare visceral GTs do occur. GTs within the gastrointestinal tract have been reported to usually occur at the gastric, especially gastric antrum. The vast majority of gastric GTs follow a benign clinical course, without histologic or clinical evidence of malignancy [2]. GTs rarely involve small intestine, with only 8 cases reported in the English literature, among which 4 cases present malignant features [3-5]. Here, we report a patient with glomus tumor of the jejunum and review the literature on the clinicopathologic features, diagnosis and treatment.

## Case Presentation

A 30-year-old female referred to our hospital with the chief complain of melena and fatigue for 3 weeks. There were no significant positive signs other than anemic appearance. Routine blood examination showed that her hemoglobin level was 6.7g/dL. Contrast-enhanced computed tomography (CT) revealed an obvious arterially enhancing mass in the small intestine and continuous enhancement in the delayed phase of enhancement (Fig. 1). Capsule endoscopy indicated a possible mucosal eminence in the jejunum (Fig. 2a). Because the photographs captured by capsule endoscopy were limited, oral single-balloon enteroscopy was further applied to determine the mass. After insertion of about 250cm past pylorus, a 2.0*2.0cm tumor without epithelial lining was discovered (Fig. 2b). The patient was then transferred to the Department of Gastrointestinal Surgery and underwent a partial enterectomy.

At surgery, a 3.5cm long segment of jejunum was removed. Gross pathology showed a 2.0*1.3*1.3cm gray-red polypoid protuberance in the small intestinal mucosal surface (Fig. 3a). HE stain illustrated that the tumor was well limited by a thin fibrous capsule, interspersed with congestive capillaries of various size, and admixed with smooth muscle bundles (Fig. 3b and 3c). Muscularis propria and serosa didn't show any damage. Immunohistochemical study showed that the tumor cells were strongly positive for SMA and collagen type Ⅳ (Fig. 3d and 3e), and negative for CD31, CD34, CD117, Desmin, DOG-1, CgA, Syn, S100, EMA, STAT6, β-catenin, Caldesmon and Calponin. Ki-67 proliferation index was less than 1% (Fig. 3f) and mitotic activity is about 1/50 HPF.

According to the CT results, endoscopic images and pathologic findings, the case was finally diagnosed as benign GT. Postoperatively, the gastrointestinal hemorrhage was completely resolved. And no recurrence or metastasis was observed during a follow-up period of 6 months.

## Discussion

GTs arising from the small intestine are extremely rare. So far, only 8 cases have been described in the English literatures [3-5]. Among them, two cases located at the duodenum, one case at jejunum, and five cases at the ileum. The clinicopathologic features of small intestinal GTs are summarized in Table 1. The case reported in our study is a 30-year-old female. Together with this case, the sex distribution is basically balanced (5 males and 4 females). Small intestinal GTs usually present non-specific symptoms such as abdominal pain and intestinal bleeding (melena or hematochezia). Larger tumors can cause intestinal stenosis and obstruction.

Due to the deep location and non-specific symptoms, small intestinal tumors including GTs are usually hard to differentiate and diagnose. Abdominal contrast-enhanced CT may be useful to distinguish GTs from other small intestinal tumors. According to previous description (mainly on gastric GTs), hemangioma-like globular enhancement with central fill-in and persistent homogeneous enhancement were both visualized in GTs [11]. As for the reported case, the tumor on contrast-enhanced CT showed homogeneous high attenuation in the arterial, venous, and delayed phases. GTs in gastric usually display a submucosal pattern of growth. In the case reported by Knackstedt C et al., the GT located in duodenal bulb was polyp-like [7]. Shelton JH et al. reported a GT of the ampulla, in which a protruding mass was seen [8]. However, until now, there is no enteroscopic image presented in the jejunum or ileum GTs. In this paper, for the first time, we acquired the endoscopic imaging of small intestinal GT by balloon-assisted enteroscopy. Intriguingly, the enteroscopic imaging showed a fibrous capsule-covered mass, without any epithelial coverage, and on the surface of the tumor, no

definite hemorrhage, erosions or ulcers were observed. Only some minor erosions on the area around the tumor edge were seen, which may cause the persistent melena.

Most GTs depend on postoperative pathological diagnosis. Under the microscope, the tumor consists of a large number of smooth muscle bundles and dilated capillaries. Immunohistochemically, tumor cells were stained positive for SMA and collagen type Ⅳ. Negative CD117 and DOG-1 expressions help in excluding the diagnosis of gastrointestinal stromal tumors, while negative CgA and Syn expressions help in excluding neuroendocrine neoplasms [12]. Criteria for malignant GTs proposed by Folpe et al. include tumors with a deep location and a size of more than 2 cm, or atypical mitotic figures, or moderate to high nuclear grade and ≥5 mitotic figures/50 HPF [13]. According to the criteria, the present case was considered as low possibility of malignancy. And 6 months follow-up showed no indication of recurrence or metastasis after a partial enterectomy, which supporting the benign diagnosis.

All the small intestinal GTs recorded in the literature have received surgical resection, but no patients received regional lymphadenectomy. After surgery, also no patients received adjuvant radiotherapy or chemotherapy. However, 2 patients developed metastases in the 8 reported cases. Therefore, standardized systemic treatment needs to be further studied. For gastric GTs, endoscopic or laparoscopic resection is recommended. Also due to the fact of extremely rare cases of malignant GTs in gastric, no standardized neoadjuvant or adjuvant treatment was suggested. Radiotherapy or chemotherapy has been utilized for the treatment of malignant GTs of head and neck. However, in the reported cases, tumor progression was not reliably altered [14].

In conclusion, we have reported a case of GT of small intestine in a 30-year-old female patient. The final diagnosis is made by histological and immunohistochemical examination. Due to the exceedingly rare occurrence, there is no standardized management pathway for these patients. As there is a potential for malignancy, early diagnosis and treatment are important for a good prognosis.

## Abbreviations

GTs: Glomus tumors; CT: Computed tomography

## Declarations

### Acknowledgements

The authors would like to thank the patient for agreeing to have her information published.

### Authors' contributions

YT, XW and YZ reviewed the literature and contributed to the manuscript drafting. GY performed the histological analysis. JF, YC and QZ were physicians in charge of the patient and contributed to the manuscript drafting. All authors read and approved the final manuscript.

### Funding

None

### Availability of data and materials

Not applicable

### Ethics approval and consent to participate

Not applicable

### Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

### Competing interests

The authors declare that they have no competing interests.

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## Tables

**Table 1** Clinicopathological characteristics of documented small intestinal GTs

| Reference | Age/Sex | Location | Size in cm | Invasion | Treatment | Immunohistochemistry | Nature | Outcome |
|---|---|---|---|---|---|---|---|---|
| Chen et al. 2020 [4] | 73/F | Ileum | 2.0*2.8*1.2 | Muscularis propria | Laparoscopic resection | SMA(+), vimentin(+) caldesmon(+), CD34(+), Ki-67 (80%+), CD117(-), desmin(-), Dog-1(-), S100(-), leukocyte common antigen (-), cytokeratin(-) | Malignant | Metastasis,Died |
| Ma et al. 2020 [4] | 58/M | Ileum | 6.0*5.0 | Serosa | Laparoscopic resection | SMA(+), Ki-67(70%), CD34(+), Nestin(+), EMA(-), CD117(-), Dog-1(-), S100(-), desmin(-) | Malignant | Liver metastases |
| Campana et al. 2014 [5] | 51/M | Ileum | 3.7 | Serosa | Laparoscopic resection | Ki-67 (<5%+) | Benign | Follow-up |
| Abu-Zaid et al. 2013 [6] | 29/F | Ileum | 12.8*10.2*13.1 | Whole layer | Surgical resection | SMA(+), collagen type IV(+), caldesmon(+), calponin(+), CD117(-), CD34(-), S100(-), cytokeratin(-), desmin(-), HMB-45(-), chromogranin(-), synaptophysin(-) | Malignant | Follow-up |
| Knackstedt et al. 2007 [7] | 65/M | Duodenum | Not available | Submucosa | Endoscopic mucosal resection | SMA(-), CD117(-), CD56(-) | Benign | Follow-up |
| Shelton et al. 2007 [8] | 48/F | Ampulla | 3.0 | Minimal invasion of common bile duct and pancreatic duct | Whipple | Not available | Malignant | Unknown |
| Geraghty et al. 1991 [9] | 60/M | Ileum | 0.6 | Serosa | Surgical resection | SMA(+), desmin(-), chromogranin(-), euron-spectificendase(-) | Unknown | Die of unrelated causes |
| Hamilton et al. 1982 [10] | 82/M | Jejunum | 1.0*1.5 | Not available | Surgical resection | Not available | Unknown | Unknown |

# Figures
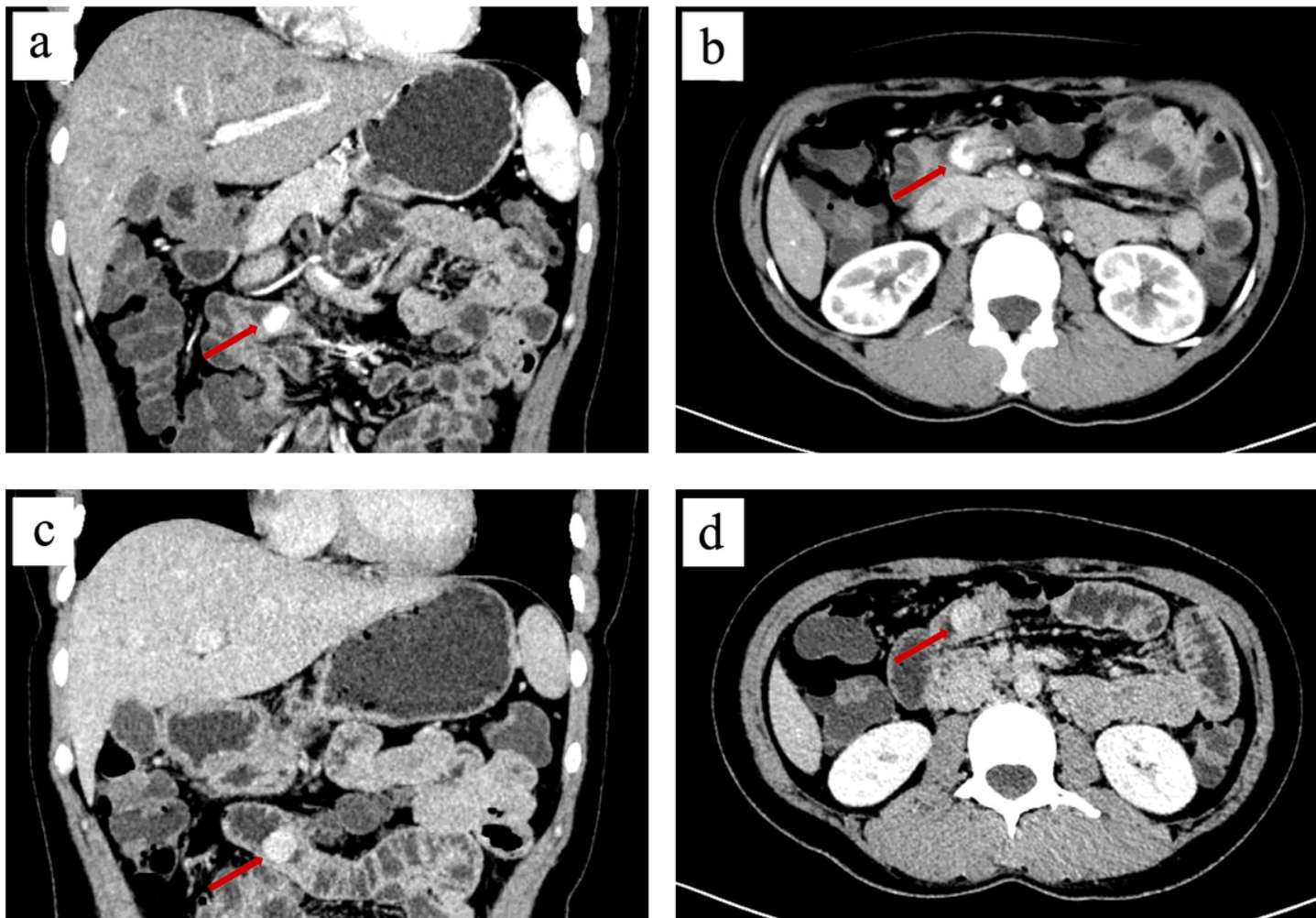
**Figure 1**

Features of the GT on contrast-enhanced CT. a, b CT images during arterial phase (coronal and horizontal planes) showed an obvious enhanced nodule in the small intestine (arrow head); c, d CT images during delayed phase (coronal and horizontal planes)
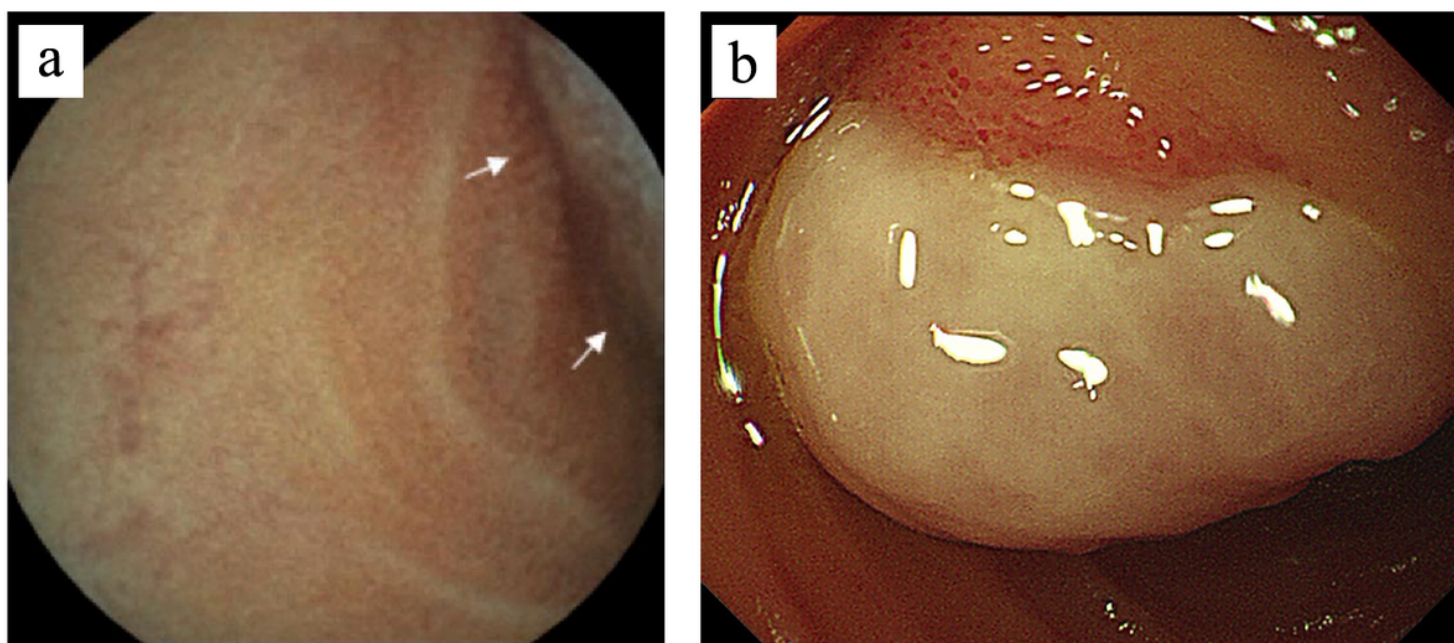
**Figure 2**

Endoscopic features of the GT. a Image of capsule endoscopy; b Image of oral single-balloon enteroscopy
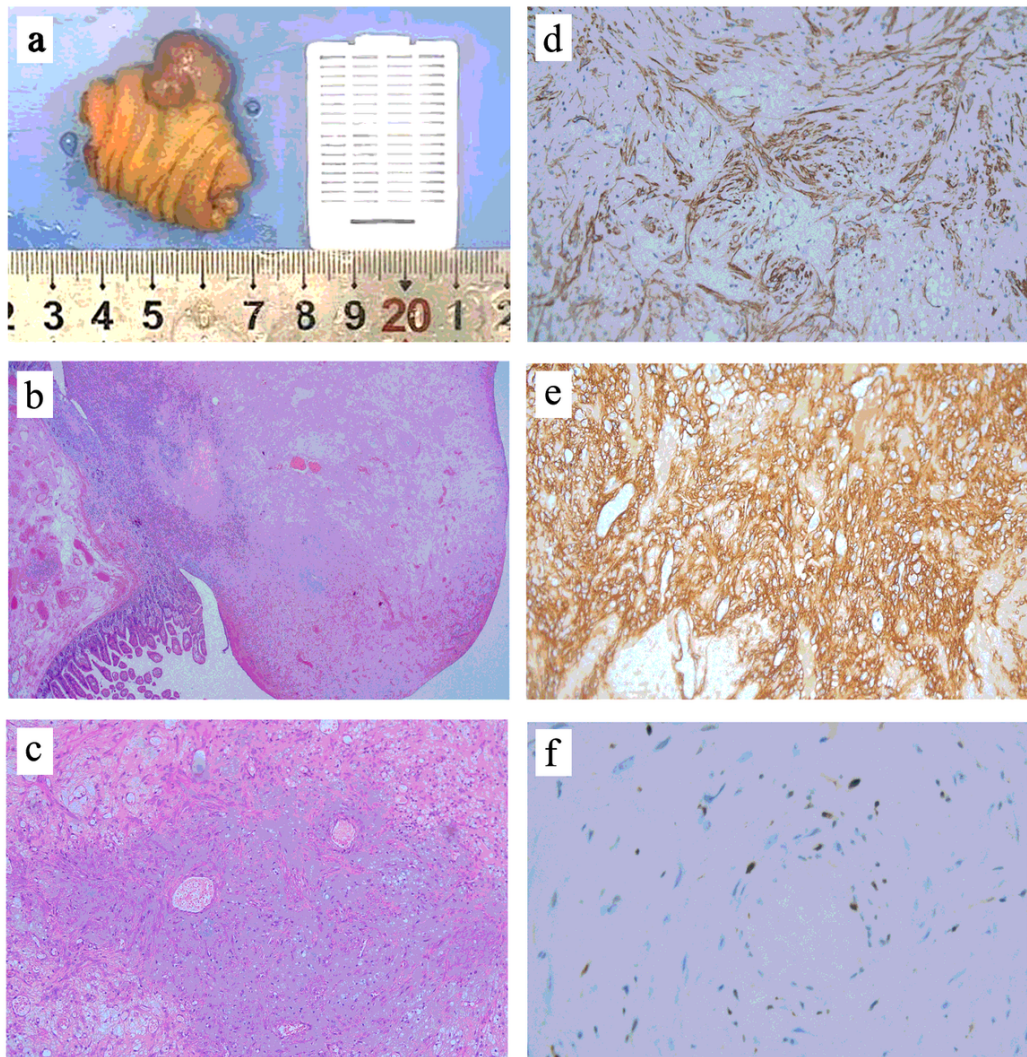
**Figure 3**

Gross findings, histological and immunohistochemical characteristics of the GT. a Gross image of resected mass; b H&E, ×40; c H&E, ×100; d SMA immunostaining, ×200; e Collagen type IV immunostaining, ×200; f Ki-67 immunostaining, ×400