Imaging findings of glomus tumor at duodenum: a case description

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Introduction

Glomus tumor is a neoplasm composed of cells that resemble the perivascular modified smooth muscle cells of the glomus body (1). Although glomus tumors usually arise in the subungual region, they may occur elsewhere in the body (2). Glomus tumors in the duodenum are rare, and only four case reports have been published in the English medical literature (3-6) (Table 1). Here, we report a surgically confirmed case of glomus tumor originating from the duodenum and describe the characteristic imaging findings.

Case presentation

A 42-year-old man was admitted to our medical institution with right upper quadrant abdominal pain for 1 month. The patient had an episode of melena 3 months before the admission. He had no history of surgeries. The physical examination was unremarkable. Laboratory hematologic and coagulation tests on admission revealed no abnormalities. Tumor markers were within the normal range: carcinoembryonic antigen, 1.97 ng/mL and carbohydrate antigen 19-9, 5.7 U/mL.

On ultrasonography (US), a well-circumscribed hypoechoic mass was seen in the duodenum, measuring 2.0 cm × 1.7 cm (Figure 1). Unenhanced computed tomography (CT) demonstrated hypoattenuating mass in the third portion of the duodenum, measuring 2.5 cm × 1.7 cm (Figure 2A). The mass showed strong enhancement with a well-defined margin in the arterial phase (Figure 2B) and persistent enhancement in the portal and delayed phases (Figure 2C,D). Lesion-to-aorta enhancement ratios were 0.62, 0.91, and 0.84 in the arterial, portal, and delayed phases, respectively. Coronal arterial phase CT demonstrated central ulceration (Figure 2E). No obvious metastases or intra-abdominal lymphadenopathies were identified.

The patient underwent a laparoscopic wedge resection of the tumor. Grossly, the tumor was a well-circumscribed intramural mass, measuring 2.5 cm × 1.5 cm. Microscopically, the mass involved the submucosa and muscularis propria (Figure 3A). On hematoxylin and eosin staining, the tumor was composed of uniform cells with round nuclei (Figure 3B). No nuclear atypia was seen, and the mitotic count was 0/50 high-power fields (HPF). No lymphovascular or perineural invasion was identified. Immunohistochemically, the tumor cells were positive for smooth muscle actin and collagen IV (Figure 3C,D) but negative for chromogranin A (Figure 3E). Synaptophysin was focally positive in the tumor cells (Figure 3F). Finally, the mass was diagnosed as a glomus tumor of the duodenum. The patient had an uneventful postoperative course and has remained free from the disease for a follow-up period of 18 months.

Discussion

Glomus tumor is a mesenchymal tumor arising from the perivascular modified smooth muscle cells of the glomus body (1). Gastrointestinal glomus tumors are rare but most frequently occur in the stomach as a subepithelial tumor (7). Epigastric discomfort is the most common symptom (8). Gastrointestinal glomus tumors often present with gastrointestinal bleeding because of ulcerations in the overlying mucosa (9).
Sonographic and CT findings of gastric glomus tumors have been described in the literature. On transabdominal or endoscopic ultrasound, they appear as well-demarcated hypoechoic masses originating from the third or fourth layer of the gastric wall (10-13). On CT, gastric glomus tumors show high enhancement in the arterial phase, which reflects their hypervascular nature. In the portal and delayed phases, the tumor shows persistent enhancement (9,14-16). Gastric glomus tumors are more strongly enhanced than other well-enhancing subepithelial lesions, such as neuroendocrine tumor, some gastrointestinal stromal tumor (GIST), and heterotopic pancreas (5). In a study by Hur et al., lesion-to-aorta enhancement ratios of glomus tumors were significantly higher than those of other subepithelial lesions in the arterial, portal, and delayed phases (5). In addition, the portal phase lesion-to-aorta ratio 0.86 or greater was significant variable for differentiating glomus tumors from other subepithelial lesions (5). In our case of the duodenal glomus tumor, imaging features were similar to those previous reported for glomus tumors, showing a lesion-to-aorta enhancement ratio of 0.91 in the portal phase.

Immunohistochemical findings are valuable for distinguishing glomus tumors from other subepithelial tumors (7). Gastrointestinal glomus tumors are positive for smooth muscle actin, vimentin, and collagen IV (7). Neuroendocrine tumors are positive for chromogranin A and synaptophysin (1). Glomus tumors are negative for chromogranin A, but there can be focal synaptophysin expression (1). GISTs are positive for KIT (CD117) and frequently positive for CD34 while glomus tumors are persistently negative for CD117 and occasionally positive for CD34 (7).

Although most cases of gastrointestinal glomus tumors are benign, cases of malignant behavior and metastases have been reported (7,17-19). Folpe et al. proposed the criteria for malignant glomus tumors, including tumors with a deep
location and a size greater than 2 cm, or atypical mitotic figures, or moderate-to-high nuclear grade, and ≥5 mitotic figures/50 HPF (18).

In conclusion, we reported a rare case of a glomus tumor arising in the duodenum. The characteristic strong and persistent enhancement on CT may be useful in differentiating glomus tumors from other subepithelial lesions.

Figure 2 Computed tomography of the duodenal glomus tumor. Axial unenhanced computed tomography (CT) scan (A) shows a well-circumscribed mass (arrow) in the third portion of the duodenum. Contrast-enhanced CT scans reveal strong enhancement in the arterial phase (B) and persistent enhancement in the portal (C) and delayed (D) phases (arrows). Coronal arterial phase CT scan (E) demonstrates central ulceration (arrow head).
Figure 3 Histopathology of the duodenal glomus tumor. (A) The tumor involved the submucosa and muscularis propria (×40); (B) hematoxylin and eosin (H&E) staining showed uniform small round cells with central nuclei and pale cytoplasm, no nuclear atypia, and no mitotic figures (×400); (C,D) immunohistochemical staining showed positive expression for smooth muscle actin and collagen IV (×40); (E,F) immunohistochemical staining showed negative expression of chromogranin A but focal positive expression of synaptophysin (×40).

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/qims.2020.03.22). The authors have no conflicts of interest to declare.

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