Glomus tumor of the stomach

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ABSTRACT

Glomus tumor is a rare benign mesenchymal neoplasm derived from the glomus body, an arteriovenous shunt mainly located in dermis and subcutis. The most common localization of this tumor is extremities, especially nailbed. Glomus tumor in the gastrointestinal system is a rare condition. Here we report a gastric glomus tumor to raise awareness of this tumor and show the difficulties in the diagnosis.

Keywords: Gastric, glomus, tumor, uncommon

INTRODUCTION

Glomus tumor is a rare benign mesenchymal neoplasm. As the name indicates, the tumor arises from the glomus body, which is an arteriovenous anastomosis, functioning without an intermediary capillary bed (1). It represents approximately 2% of all soft tissue tumors. The majority of glomus tumors occur in the distal extremities especially in the fingers, particularly in the subungual region and the skeletal muscle (2). The stomach is an exceptional site for glomus tumor. The first case of gastric glomus tumor was reported in 1951 by Kay et al. (3). Since then, few cases have been reported (1). Gastric glomus tumors, which are located in the antrum or pylor, are typically seen in the submucosa and muscularis propria. The tumors are generally benign. However, rarely they may show malignant behavior, according to localization, size, high nuclear grade, and atypia (4, 5).

CASE PRESENTATION

A 68-year-old man was admitted to our emergency service with an upper gastrointestinal bleeding episode. The patient was hemodynamically stable and laboratory results were in normal limits. An upper gastrointestinal endoscopy was planned (Figure 1). Endoscopy revealed a submucosal mass located in the antrum. Due to the submucosal mass existence, an endoultrasonographic evaluation was performed. A hypervascularized mass of 4 cm, with microcalcifications inside, was found in muscularis propria (Figure 2). With prediagnosis of gastrointestinal stromal tumor (GIST), endoultrasonographic fine-needle aspiration was performed. Hypocellular biopsy had few SMA positive smooth muscle cells, fibrin, and blood. Immunohistochemically, CD34, S100, DOG1, and CD117 were negative and Ki-67 index was 1%. With these findings, biopsy was not consistent with GIST. The patient was subjected to further investigation. Abdominal contrast-enhanced computed tomography (CT) was performed and revealed a well-circumscribed, homogeneously enhanced 25×23 mm solid submucosal tumor located in the gastric antrum in the greater curvature (Figure 3). The tumor was interpreted with a prediagnosis of neuroendocrine tumor (NET). Surgery was planned with prediagnosis of GIST and NET. Additional tests for CEA, CA 19-9, Chromogranin A, and gastrin levels were in normal limits. Wedge resection and a partial omentectomy were performed.

In macroscopic examination, 5.5×2.5×2.5 cm nodular mass, arising from the submucosa and extending through the muscularis of the stomach, was observed. The excised specimen had clear margins. The cut surface of the mass was white and hyperemic. In microscopic examination, it was a highly vascular tumor composed of thin-walled vessels. The vessels were surrounded by monomorphic, small, round-to-polygonal cells forming nests, sheets, and strands. The cells had centrally located nuclei, inconspicuous nucleoli, and clear-to-eosinophilic cytoplasm with sharply defined cell borders (Figure 4). There were neither necrosis nor mitosis in the tumor. Collagen type IV, an immunohistochemical marker in favor of glomus tumor, was focally and mildly positive. Immunohistochemically, SMA (Figure 5) and vimentin (Figure 6) were diffusely positive, and caldesmon was focally positive in tumor cells. CD 117, DOG 1, S100, CD34, chromogranin, synaptophysin, CD56, CD57, PGP9.5, and desmin were all negative. Ki-67 index was 1%, consistent with the former biopsy. Clinical, morphologic, and immunohistochemical findings were consistent with the diagnosis of glomus tumor.

After the diagnosis of glomus tumor, the patient underwent systemic examination, but no evidence of metastasis was found. The patient was discharged from the hospital six days later without any postoperative complications.
DISCUSSION

Glomus tumors arise from specialized cells of the glomus body. They are typically found in peripheral soft tissues, generally located in dermis and subcutis. Miettinen et al. (6) reported a series of 32 gastrointestinal glomus tumors, and 31 of them were gastric and one of them had cecal localization. According to them, gastric glomus tumors constitute 1% of the gastric stromal tumors. Gastric glomus tumors generally located in the submucosa or muscularis propria of the gastric wall. The symptoms are nonspecific, e.g., abdominal discomfort, epigastric pain, and/or upper gastrointestinal bleeding (5). Gastric glomus tumors are generally solitary and located in the greater curvature (7, 8). They are commonly seen in the sixth decade (9). These tumors are more frequent in women than in men (6). Although the gender was uncommon, our patient had the other common features of the tumor.
Most of the tumors have well-circumscribed borders and their size ranges from 1 to 22 cm. In microscopic examination, the tumors compose of vascular channels surrounded by monomorphic, small round cells with sharp cell borders, centrally located nuclei, and inconspicuous nucleoli. Pleomorphism, atypia, mitosis, and necrosis are uncommon findings. Immunohistochemically, the tumor is strongly positive for SMA, vimentin, actin, calponin, type IV collagen, and laminin. CD117, DOG1, S100, CD34, desmin, and neuroendocrine markers such as chromogranin, synaptophysin, neuron-specific enolase, CD56, CD57, and PGP 9.5 are negative (1, 5). Our patient’s morphologic and immunohistochemical features were consistent with the literature.

The diagnosis of gastric glomus tumor can be challenging with endoscopic and radiologic findings, which can also be seen in other gastric stromal and mesenchymal tumors (4). Gastrointestinal stromal tumor parangangioma, and NET should be considered for differential diagnosis. Gastrointestinal stromal tumor is the most common mesenchymal tumor of the stomach. Epithelioid type of GIST is one of the most important tumors in differential diagnosis of glomus tumor. Different from glomus tumor, GISTs are positive for CD117, DOG1, and CD34. Positivity for desmin, vimentin, and SMA are variable and S100 immunoreactivity is rare in glomus tumors (1, 9, 10). Different from GIST, our tumor had distinctive dilated vascular structures. CD117 and DOG1 were applied to three tumor blocks and all were negative.

Paragangliomas are generally located in the retroperitoneum. They have Zellballen and alveolar pattern with accompanying thin-walled vessels. They characteristically show immunoreactivity with chromogranin, synaptophysin, and S100 protein (1). All of these markers were negative in our submucosal localized gastric tumor.

Neuroendocrine tumors have nests and cords of oval and/or spindle large cells. The nuclei have salt and pepper chromatin features. Glomus tumor of uncertain malignant potential term is used for tumors having high mitotic activity and additional criteria such as superficial location, large size, or deep localization (13). Our patient had a 5.5-cm tumor without mitosis and necrosis. The tumor had small monotonous cells with a centrally located nucleus. Nucleolus was not prominent. According to these features, our case was considered as benign and follow-up was recommended. Our patient is under control and out of disease for 18 months.

CONCLUSION
Gastric glomus tumors are rare mesenchymal tumors that are difficult to diagnose before excision. Their clinical, radiologic, endoscopic, and even cytologic features can overlap with the common stromal and mesenchymal gastric tumors. After the removal, morphologic features and immunohistochemical examination play an important role for the diagnosis of gastric glomus tumor.

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