

Glomus tumor of the stomach: A case report and review of the literature

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SUMMARY

Glomus tumor is a benign soft tissue neoplasm which commonly affects the subungual region of the fingers. But the tumors can also arise in the other sites such as the antrum of the stomach. We are reporting a case of a glomus tumor of the stomach in a 71-year-old female patient who presented with dyspepsia. The tumor was confined to the lamina muscularis propria, it consisted of round cells with small uniform nuclei, which surrounded thin walled blood vessels. Immunohistochemistry revealed the tumor to be positive for smooth muscle actin, vimentin, calponin, h-caldesmon and negative for c-KIT, S-100, CD34, CD99, synaptophysin, chromogranin, desmin and EMA. The proliferation marker Ki-67 was positive in less than 5 % of tumor cell nuclei. Glomus tumors are usually benign but malignant cases have been published. Criteria for the malignant potential of gastric glomus tumors remain poorly defined.

Keywords: glomus tumor – gastric tumors – glomus bodies

Glomus tumor žaludku – popis případu a přehled literatury

SOUHRN

Glomus tumor je málo častý nádor měkkých tkání vyskytující se převážně pod nehty na prstech rukou. Může se objevit i v jiných lokalitách, z nichž nejčastěji v antru žaludku. Presentujeme případ glomus tumoru žaludku u 71 leté ženy s dyspeptickými obtížemi. Tumor byl lokalizován v oblasti svaloviny žaludečního antra, byl tvořen kulatými uniformními buňkami, které byly nahromaděny v okolí tenkostěnných cévních formací. Immunohistochemicky exprimovaly nádorové buňky alfa aktin hladké svaloviny, vimentin, calponin a fokálně h-caldesmon. Negativní byl průkaz c-KIT, S-100, CD34, CD99, synaptofyzinu, chromograninu, desminu a EMA. Proliferační aktivita (Ki-67 index) byla nižší než 5 %. Většina glomus tumorů je benigních, ale byly popsány i maligní případy s metastázami. Kriteria malignity glomus tumorů v lokalitě žaludku nejsou dosud přesně stanovená.

Klíčová slova: glomus tumor – tumory žaludku – glomová tělíska

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Glomus tumor is a mesenchymal neoplasm which belongs to the group of tumors from perivascular cells of the glomus body. Most cases affect the subcutaneous tissue of the distal extremities, particularly the subungual region. The tumors can also arise in tissues containing few or no glomus cells (1).

The normal glomus body is a specialized form of an arteriovenous anastomosis associated with thermoregulation and regulation of arterial flow. It is located in the stratum reticularis of the dermis mainly in the tips of the fingers. The glomus body is made up of an afferent arteriole, efferent venule, smooth muscle cells, nerves and glomus cells. These rounded cells resemble smooth muscle cells. The term "glomus" comes from Latin word for „ball“. Three basic types of tumors, based on proportions of individual structures, can be distinguished. These are: glomus tumor, glomangioma and glomangiomyoma.

The second most common site of glomus tumors is the antrum of the stomach (2). Approximately 100 cases have been published in literature (3). The incidence of gastric glomus tumors is much less common than that of gastrointestinal stromal tumors (GIST) with only 1 in 100 GISTs being a gastric glomus tumor (3).

Gastric glomus tumors present with a variety of symptoms, including epigastric discomfort, nausea and vomiting, and hematemesis. Melena may also occur (4).

CASE REPORT

A 71-year-old woman was examined for epigastric discomfort. Gastroscopy, endoscopic ultrasonography and computerized tomography revealed a mural tumor in the antrum of the stomach. A laparotomy was done with a resection of the tumor. One perigastric lymph node was also resected, examined by frozen section, but it revealed no tumor involvement.

The tumor was 15 x 10 x 10 mm in size. Histologically, the tumor was confined to the lamina muscularis propria, without involving the mucosa, submucosa or serosal surface. The tumor consisted of round cells with small, uniform nuclei, which surrounded thin walled blood vessels (Fig. 1, 2). There was found no angioinvasion. Immunohistochemistry revealed the tumor to be positive

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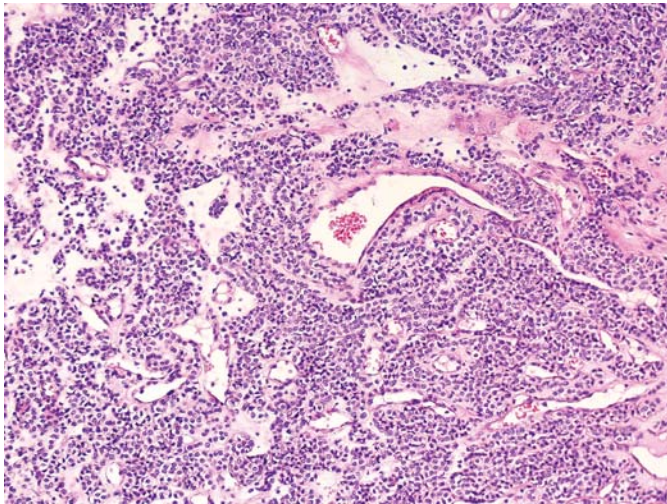


Fig. 1. Gastric glomus tumor – round cells around the dilated thin-walled blood vessels (hematoxylin and eosin, original magnification 200x).

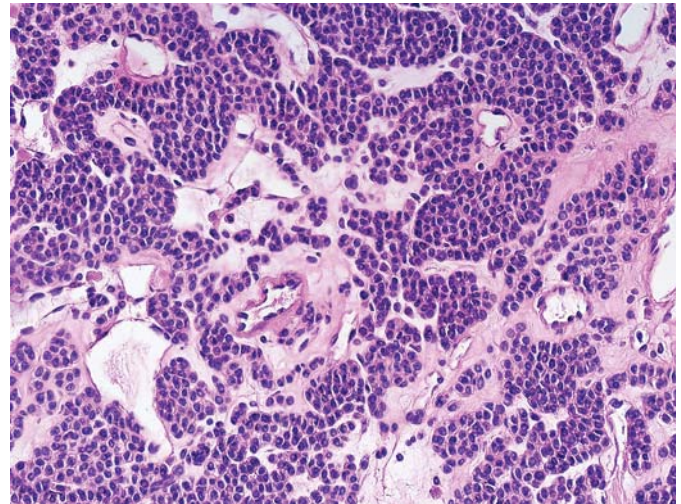


Fig. 2. Gastric glomus tumor – round cells with small uniform nuclei and sharp cell borders. (hematoxylin and eosin, original magnification 400x).

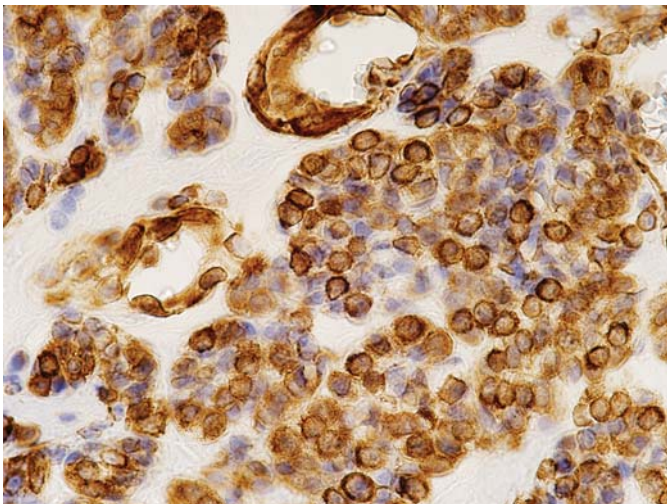


Fig. 3. Gastric glomus tumor – immunohistochemistry revealed the tumor to be positive for smooth muscle actin (hematoxylin and eosin, original magnification 400x).

for smooth muscle actin (Fig. 3), vimentin, calponin, h-caldesmon and negative for c-KIT, S-100, CD34, CD99, synaptophysin, chromogranin, desmin and EMA. The proliferation marker Ki-67 was positive in less than 5 % of tumor cell nuclei.

We established the diagnosis of a gastric glomus tumor. Now, nine months after resection, the patient still shows no evidence of relapse.

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DISCUSSION

Glomus tumors are usually benign, but malignant cases have been published. Folpe et al. (5) proposed the following classification criteria in 2001 for malignant glomus tumors: deep location, size more than 2 cm, mitotic activity (more than 5 cells undergoing mitosis per 50 high-power fields), and the presence of atypical mitotic figures. Classification criteria have been established for superficial and deep soft tissue tumors; however, it remains unclear whether these criteria are also suitable for gastric glomus tumors (3).

Vascular invasion is quite common in gastric glomus tumors, which probably represents only a pattern of local spread without association with malignant behavior (6). In a study of Miettinen et al. (6), only one of 11 gastric glomus tumors with vascular invasion metastasized.

Glomus tumors are immunopositive for smooth muscle actin, calponin, vimentin and h-caldesmon. Focal positivity for CD34 has been demonstrated in a minority of cases. Immunohistochemistry for c-KIT, desmin, cytokeratin, S-100, chromogranin, synaptophysin, CD20, CD45 and HMB-45 is negative.

The differential diagnosis includes GIST (gastrointestinal stromal tumor), which is c-KIT positive, paraganglioma (S-100, chromogranin and synaptophysin positive), carcinoid (cytokeratin, chromogranin and synaptophysin positive) and lymphomas (CD45 and/or CD20 positive).

Wedge resection with negative margins is the recommended therapy, enucleation alone has a high recurrence rate (4).