Glomus Tumor of the Stomach

Midenin Glomus Tümörü

Ufuk USTA¹, Filiz ÖZYILMAZ², Çiğdem ÖZDEMİR³, Güлara HÜSEYİNOVA¹, Atakan Yavuz SEZER²

Departments of ¹Pathology and ²General Surgery, Medical Faculty of Trakya University, Edirne

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Glomus tumors are uncommon tumors of mesenchymal origin which generally arise from peripheral soft tissues, particularly in the distal parts of the extremities.¹,² However, rare cases with visceral involvement are reported in the literature.³ In the gut they are most commonly

A sixty-four-year old male patient presented to the emergency room with massive upper gastrointestinal tract bleeding. During distal gastrectomy, a submucosal, well demarcated mass with a diameter of 2.4 cm was palpable in the antral region of the lesser curvature. Histopathological examination of the mass revealed uniform tumor cells with clear cytoplasmas and round nuclei, forming nests, strands and sheets between dense, irregularly branching, thin-walled vessels. Immunohistochimistry was positive for vimentin and SMA and negative for cytokeratin, CD34, synaptophysin, chromogranin A, NSE, CD117 (C-kit) and S-100 protein. Electron microscopy demonstrated tumor cells with clear cytoplasmas and round nuclei with coarsely dispersed chromatin, forming groups around vascular structures. Clinical findings, gross appearance, tumor pattern, immunohistochemical findings and electronmicroscopic findings were all consistent with a glomus tumor.

Key Words: Glomus tumor/pathology; immunohistochemistry; stomach.

Glomus tumors are uncommon tumors of mesenchymal origin which generally arise from peripheral soft tissues, particularly in the distal parts of the extremities.¹,² However, rare cases with visceral involvement are reported in the literature.³ In the gut they are most commonly
found in antrum of the stomach and typically they are single tumors.\[4,5\] We present a 64-year-old man with a glomus tumor in his stomach presenting with massive upper gastrointestinal tract bleeding, which led him to an urgent laparotomy and distal gastrectomy.

CASE REPORT

A 64-year-old man presented to the emergency department of our hospital, suffering from vomiting fresh blood for two hours. Because of massive hematemesis, the patient has undergone an urgent distal subtotal gastrectomy and Billroth II anastomosis after his initial examination and endoscopic evaluation. At exploration, a well demarcated mass was palpable in the wall of the antral region of the lesser curvature. The location of the tumor precluded a successful resection with safe surgical margins. The patient did not require any additional medical therapy except for a blood transfusion of 500 ml and intravenous fluid support for four days. The postoperative course of the patient was unremarkable and he was discharged from the hospital on postoperative day five tolerating a regular diet.

The macroscopic inspection of the dissected specimen revealed a 2.4 cm submucosal mass in the lesser curvature of the antrum. The hyperemic and hemorrhagic mucosa overlying the mass had an ulceration of 0.5 cm in diameter. The cut sections revealed a grossly well demarcated, solid, gray-colored, partly hemorrhagic mass located in the submucosa, extending to the subserosa of the stomach (Fig. 1).

The air dried and Giemsa-stained imprint cytology revealed individually scattered tumor cells throughout the slides. The tumor cells did not form any distinct, well formed groups. They had eccentrically located, uniform nuclei with an average diameter of two to three times of an ordinary mature lymphocyte. There was no nuclear atypia and mitotic figures were scanty. At first glance, the cytomorphologic features of the tumor suggested a carcinoid tumor (Fig. 2).

A sample of 0.5 cm in diameter was obtained from the fresh tumor tissue and fixed in gluteraldehyde for electronmicroscopic evaluation. The rest of the distal gastrectomy specimen was sampled after an overnight fixation in 10% formaldehyde at room temperature. The 5 mm-thick paraffin sections were stained with routine hematoxylin-eosin (H-E) stains. Sections from a selected tissue block were immunohistochemically stained for vimentin, cytokeratin, alpha smooth muscle Actin (SMA), CD34, synaptophysin, chromogranin A, NSE, CD117 (C-kit), S-100 protein and Ki-67. Reticulin silver stain was applied to a section of the tumor for histochemical evaluation.

The HE-stained slides revealed a tumorous mass beginning from the submucosa, extending to the subserosa of the stomach. The monotonous, uniform tumor cells with clear cytoplasms and round nuclei, formed nests, strands and sheets between dense, irregularly

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Fig. 1. Gross appearance and the cut section of the tumor.

Fig. 2. Tumor cells with broad cytoplasms and eccentrically located round nuclei. Some nuclei are bare and there are cytoplasmic fragments in the background (MGG x 400).
branching, thin-walled vessels (Fig. 3a). The tumor cells revealed 2 mitosis/50 high power fields (HPFs). Neither cytological atypia nor necrosis was present. There was a ruptured vessel causing hemorrhage through the partly ulcerated mucosa, into the lumen of the stomach (Fig. 3b). The tumor showed local invasion to the adjacent tissues with scattered tumor islands. Immunohistochemistry was positive for vimentin and SMA and negative for cytokeratin, CD34, synaptophysin, chromogranin A, NSE, CD117 (C-kit) and S-100 protein. The Ki-67 proliferative index of the tumor was 30%, which may be considered high for a benign tumor. The reticulin stain revealed fine reticulin fibers encircling nests of two to four tumor cells (Fig. 3c). Electron microscopy revealed tumor cells with clear cytoplasm and round nuclei with coarsely dispersed chromatin, forming groups around vascular structures. Well-formed intercytoplasmic desmosomes were established between the tumor cells. There were excess amounts of mitochondria and some neurosecretory granules in the cytoplasms of the tumor cells (Fig. 4a, b).

The clinical findings, gross appearance, tumor pattern, immunohistochemical findings and electron microscopy results were all consistent with a glomus tumor.

**DISCUSSION**

Vascular tumors of the gastrointestinal tract (GIT) are extremely rare, accounting for <2% of all benign tumors of the GIT. The largest portion of these vascular tumors are glomus tumors. Glomus body is a neuromyoarterial receptor that regulates blood flow within arteries and these tumors originate from the modified smooth muscle cells of the glomus body.

Although gastrointestinal glomus tumors may present with a variety of symptoms, they typically present with gastrointestinal bleeding because of the ulceration of the overlying mucosa. Other presenting symptoms include obstruction and non-specific ulcer symptoms, but still many may remain asymptomatic. Our patient was asymptomatic until the massive upper gastrointestinal bleeding led him to an urgent surgical operation.

**Fig. 3.** (a) The monotonous, uniform tumor cells with clear cytoplasm and finely dispersed chromatin, forming nests, strands and sheets between dense, irregularly branching, thin-walled vessels (H-E x 100). (b) Hemorrhage through the partly ulcerated mucosa, into the lumen of the stomach. (H-E x 50) (c) Fine reticulin fibers encircling nests of two to four glomus cells (Reticulin silver stain x 100). H: Hemorrhage, U: Ulcerated mucosa, M: Mucosa, T: Tumor.
The differential diagnosis of gastric glomus tumor includes carcinoid tumor, hemangiopericytoma and gastrointestinal stromal tumor (GIST). It has been postulated that the glomus tumor exists as an intermediate form between hemangiopericytomas and leiomyomomas in a broad spectrum of smooth muscle tumors and this may account for similarities between entities considered during differential diagnosis.

The examination of vascular spaces usually reveal ectatic thin-walled vessels in glomus tumors, whereas in carcinoid tumors vascularity and fibrosis is prominent. The glomus cell nuclei are round and frequently have a perinuclear pale area or clear cytoplasm. Both hemangiopericytomas and epithelioid GISTs lack these features. The cells of glomus tumors usually lack both immunohistochemical and electronmicroscopic evidence of neuroendocrine granules, though some may present these features. In our case there were a few neuroendocrine granules at ultrastructural level which were not supported by immunohistochemistry. Glomus tumors are almost always positive for SMA and vimentin and negative for cytokeratin and S100 protein, and are usually negative for CD34, CD117 and neuroendocrine markers such as chromogranin A, synaptophysin, NSE and neurofilament. In the presented case, results of immunohistochemistry were all consistent with the findings of a suspected glomus tumor.
The diagnosis of glomus tumor is almost never made preoperatively and even intraoperatively, as frozen section and imprint cytology diagnosis is difficult and a large number of cases are misinterpreted as carcinoid tumors.\(^7,9\) The imprint slides of the presented case also had the cytomorphologic features of a carcinoid tumor.

As mentioned above, glomus tumors are essentially benign in nature and local resection is curative.\(^7\) Our patient still survives without any significant morbidity at 12 months after resection of the tumor. Nonetheless, long-term follow-up of this patient will be necessary, because of the unpredictable potential for malignant behavior of these tumors.

REFERENCES


