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Case Report

Malignant glomus tumor of the gastric antrum with hepatic metastases: a case report and literature review

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ABSTRACT

Glomus tumor (glomangioma) is an uncommon pericytic neoplasm derived from glomus cells, which are modified smooth muscle cells of the glomus body. Malignant glomus tumors (glomangiosarcomas) are extremely rare mesenchymal neoplasms. There are limited reports of metastatic malignant glomus tumors within the gastrointestinal tract in the literature. We report a case of an 80-year-old woman who presented with vague epigastric discomfort. On computed tomography (CT) imaging, a 7.1 cm solid enhancing mass was seen originating from the gastric antrum and was clinically suspected to be a gastrointestinal stromal tumor (GIST). Needle core biopsies were obtained from the antral mass and demonstrated an epithelioid neoplasm with a brisk mitotic activity. The morphology and imunohistochemical profile (CKIT/DOG1 negative and strong immunoreactivity with smooth muscle actin), was consistent with a malignant glomus tumor, at least FNCLCC grade 2. At the time of the planned gastric resection, multiple hepatic metastases were identified and confirmed on frozen section to be consistent with metastatic malignant glomus tumor.

1. Introduction

Glomus tumor (glomangioma) is a rare type of pericytic (perivascular) neoplasm that is clinically indolent. It arises from the glomus bodies, arteriovenous anastomoses involved in thermoregulation, which are most numerous in the dermis or subcutis of the digits, especially subungally. Glomus cells (specialized pericytes) are derived from modified smooth muscle cells of the glomus body and show immunohistochemical and ultrastructural features of smooth muscle differentiation [1]. Typically, glomus tumors present as a single, small (often < 1.0 cm in diameter), encapsulated, blue-red, painful nodule that affects younger adults.

Malignant glomus tumor (glomangiosarcoma) is an extremely rare mesenchymal neoplasm, accounting for < 1% of glomus tumors [3]. They are distinguished from the benign counterpart by their size larger than 2.0 cm, deep or visceral location, infiltrative growth pattern, necrosis, and additional histologic criteria such as nuclear pleomorphism and increased mitotic activity (≥5 per 50 high-power fields). Despite their malignant histological characteristics, it has been reported that malignant glomus tumors infrequently metastasize [3–5].

We present a case of a large gastric antral glomus tumor showing malignant histologic features in an 80-year-old woman with rapid growth and hepatic metastases. Metastatic malignant glomus tumors, especially of the gastrointestinal (GI) tract, are a rare entity and thus clinical behavior is uncertain. Therefore, we performed a literature search of over 100 articles to try and better classify this rare entity.

2. Case presentation

An 80-year-old female presented with complaints of vague upper abdominal discomfort that worsened when eating meals, of approximately six weeks' duration. The patient described abdominal discomfort upon overeating and endorsed a voluntary 3-pound weight loss over the past two months, attributed to eating slightly less than usual lately. She denied decreased appetite, early satiety, obstructive symptoms, vomiting, hematemesis, or changes in bowel function.

She has a past medical history of hypertension (on Irbesartan and Metoprolol), hypercholesterolemia (on Pravastatin), depression (on Sertraline), neuropathy of distal lower extremities (on Gabapentin), and osteoarthritis (on Acetaminophen). She has a past surgical history of an ovarian cyst removal, appendectomy, Cesarean section, and hysterectomy; all > 40 years ago. Her family history is notable for breast cancer in a sister who is alive and well and a brother who died from esophageal cancer in his 60s. She has a past history of smoking for approximately 20 years, but quitted approximately 30 years ago. She also quitted alcohol use at that time. There is no current alcohol or drug

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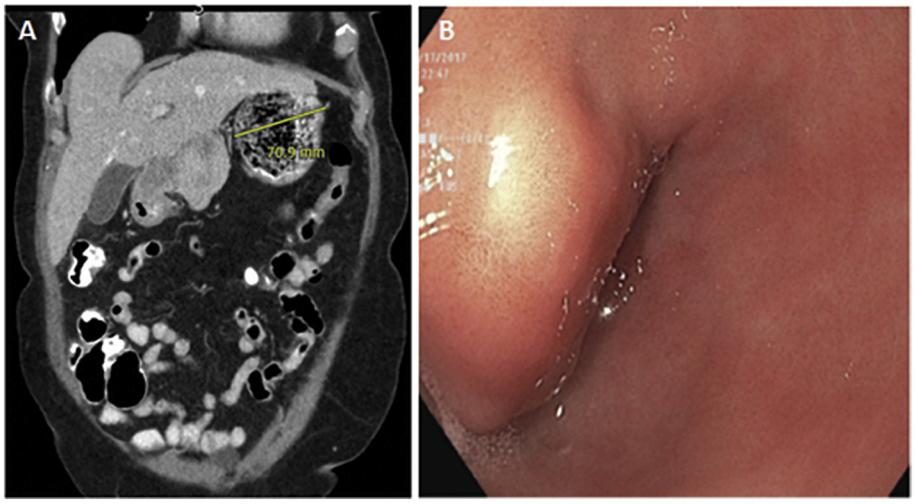

Fig. 1. Gastric submucosal malignant glomus tumor on CT scan (A) and endoscopy (B).

use.

On physical examination, fullness was appreciated in the epigastrium and right upper quadrant, just to the right of the xiphoid. In this area, slight abdominal tenderness was noted. There was no evidence of peritonitis during the examination. Her laboratory work-up, including complete blood count and complete metabolic panel, was unremarkable.

She was subsequently treated with a proton pump inhibitor. However, the discomfort persisted and this prompted further evaluation with an abdominal ultrasound. An abdominal ultrasound demonstrated a mass in the right upper quadrant measuring approximately 4.5 cm. A follow-up computed tomography (CT) scan with intravenous contrast confirmed the presence of a lobulated, heterogeneously enhancing solid mass with a central area of hypo-attenuation, measuring 7.1 cm (Fig. 1A). The mass had a broad-based area of contact with the gastric antrum. It was located near the duodenum, anterior to the pancreas, and abutted (but did not invade) the inferior aspect of the left hepatic lobe. The liver demonstrated no focal lesions and there was no evidence of metastatic disease on CT scan. Given the location and appearance of the mass on diagnostic imaging, the overall clinical impression was likely a GIST.

The patient subsequently underwent an endoscopic ultrasound-guided fine needle aspiration and biopsy. Endoscopically, the antral mass was submucosal, well-demarcated, and non-obstructing; the rest of the stomach appeared normal (Fig.1B). Cytologic evaluation of the mass revealed sheets of and clusters of crowded neoplastic cells with epithelioid to slightly spindled morphologies. Some cells had a conspicuous nucleolus. The differential diagnosis was broad and, unfortunately, a cell block was acellular and immunohistochemical staining, performed on the limited tissue material, was non-contributory. The concurrent needle core biopsy consisted of gastric mucosa with regenerative and reparative changes including foveolar hyperplasia. The submucosal mass was not identified, despite examination of multiple deeper sections.

With the aim of obtaining further tissue material for immunohistochemical stain evaluation and a definitive pathologic diagnosis, a subsequent CT-guided transgastric fine-needle aspiration and core needle biopsy were performed a month later. The repeat fine needle aspiration yielded cells with similar cytomorphologic features as previously described. Histologically, dilated and angulated blood vessels were surrounded by solid sheets of glomus cells (Fig. 2A). The neoplastic cells were mornomorphic and showed eosinophilic cytoplasm with sharply defined cell borders, central oval nuclei, and brisk mitotic activity (28 per 10 high-powered fields) (Fig. 2B), corresponding to a score of 3 (FNCLCC grading system) [6]. Necrosis was not identified in the biopsy specimen (score of 0) despite possible central necrosis noted on CT imaging. The biopsy specimen has a total score of 4, corresponding to a histologic grade 2 (FNCLCC grading system). Lympho-vascular invasion was not identified. Immunohistochemistry was performed and showed robust positivity for smooth muscle actin (Fig. 2C) within the tumor cells. Synaptophysin staining showed weak, patchy non-specific staining. Additional immunohistochemical stains, including Desmin, DOG-1, Ckit (CD117), Keratin AE1-AE3 (Fig. 2D), S-100, EMA, CD34, Stat-6, LCA, CD34, and chromogranin, were all negative (data not shown). The overall histomorphologic and immunohistochemical profile of the 7.1 cm tumor was most consistent with a pericytic (perivascular) tumor (WHO classification of soft tissue tumors), most specifically a malignant glomus tumor at least of histologic grade 2.

The patient was taken to the operating room for planned exploratory laparotomy and resection. Intraoperatively, the mass appeared to be arising from the antrum and involved the pylorus. However, during the exploratory laparotomy, multiple hepatic nodules were found and a liver wedge biopsy of a nodule was sent for intraoperative frozen section consultation. The liver wedge contained three metastatic foci, the largest measuring 0.4 cm (Fig. 2E, F). On frozen sections, these hepatic nodules showed identical histomorphology to the prior gastric malignant glomus tumor and the resection was aborted. Since there was mild narrowing of the distal antrum/pylorus, a palliative gastrojejunostomy was performed to prevent gastric outlet obstruction.

A post-operative CT scan with intravenous contrast was performed of the chest, abdomen, and pelvis for re-staging. The heterogeneous enhancing gastric antral mass now measured 11.0 × 8.6 × 7.5 cm, representing a significant interval growth. There was no definitive evidence of metastatic disease in the chest. The patient was discharged home on post-operative day 6. Palliative care and oncology consultations were obtained for this unresectable rare tumor with plans to start chemotherapy three weeks post-operatively.

3. Discussion

The majority of glomus tumors occur in the soft tissue of the distal extremities. However, rare deep or visceral glomus tumors do occur. Glomus tumors within the GI tract have been reported to occur at the gastric antrum [7–12], liver [13], duodenum [14], and cecum [7]. The vast majority of gastrointestinal glomus tumors follow a benign clinical course, without histologic or clinical evidence of malignancy. These reports support the notion that, overall, glomus tumors of the stomach have a good prognosis, with a low recurrence rate or risk of metastasis