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Case Report and Review of the Literature

Malignant Glomus Tumor of the Gastric Antrum with Liver Metastasis in a 35-year-old Female: A Case Report

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ABSTRACT

Glomus tumors (GTs) are mesenchymal tumors derived from modified smooth muscle cells of the glomus body. These tumors account for less than 2% of all soft tissue tumors, and of those less than 1% are malignant. A 35-year-old African American female presented with symptomatic iron deficiency anemia. On esophagogastroduodenoscopy, a 5 cm mass was seen partially obstructing the antrum and tissue biopsy was obtained. The biopsy showed low-grade epithelial mesenchymal neoplasm with features of a glomus tumor. A follow up computed tomography (CT) chest/abdomen/pelvis revealed a 5 cm relatively homogeneous solid mass in the gastric antrum, which appeared most similar to a gastrointestinal stromal tumor. During the planned distal gastrectomy with reconstruction, one hepatic metastasis was identified via intraoperative ultrasound and was confirmed by frozen section consultation to be the same histologic type as the antral mass. Pathologic and immunohistochemical findings were consistent with a malignant gastric glomus tumor with liver metastasis. Most GTs have a benign clinical course making the diagnosis and treatment of malignant GTs an ongoing challenge. The data on glomus tumors of the viscera is limited by the rarity of these tumors, with most of the known presentation and treatment gathered from case reports.

Introduction

Glomus tumors (GTs) are usually benign lesions, often found in the digits, presenting as painful subungual nodules [1]. The tumor itself originates from a glomus body, a component of the dermis involved in temperature regulation, proliferating from modified smooth muscle cells [2]. Although the majority of glomus tumors are benign and rarely associated with the gastrointestinal (GI) tract, rare cases of malignant gastric glomus tumors have been reported dating back to 1981 [3]. One study looking into the clinicopathologic and immunohistochemical features of gastric glomus tumors found the mean size of these tumors was 2.1 cm with a mean patient age of 49.3 years old, and the most common presenting symptom being epigastric discomfort [4]. Due to the poor utility of imaging in the diagnosis of GTs, they are often mistaken for GIST or other neuroendocrine tumors on imaging and need immunohistochemical analysis for a proper diagnosis. The information we have on these tumors in terms of their presentation, intraoperative challenges, and postoperative outcome is limited by their rarity especially when faced with a malignant glomus tumor with the potential for metastasis. We are presenting a case of a 35-year-old female with malignant glomus gastric tumor with liver metastases at an academic institution. This case report has been reported in line with the SCARE criteria [5].

Case Presentation

A 35-year-old female presented to an ambulatory care center after two months of worsening fatigue, lightheadedness, and shortness of breath. She reported some associated nausea and a decreased appetite. She denied vomiting, hematemesis, or changes in bowel function. Review of systems was negative for recent bleeding, easy bruising, melena or hematochezia. She was amenorrheic since an intrauterine device was

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placed three years prior. She had a past medical history significant for hypertension controlled on labetalol and mild intermittent asthma controlled with albuterol. She had a limited past surgical history of a breast lumpectomy 13 years prior for fibroadenoma excision, and cesarean section three years prior. Her family history was notable for lung cancer in her maternal grandfather with no other familial cancers noted. She denied any history of smoking and rarely consumed alcohol. There was no recent or past drug use. Physical exam was only significant for pale conjunctiva. Her laboratory data showed a haemoglobin of 4.6 g/dL and microcytosis with mean corpuscular volume of 65 fL. She was subsequently diagnosed with anemia and was referred to the emergency department for further evaluation.

On admission to the hospital, the patient was symptomatically anemic with haemoglobin and hematocrit 4.6 g/dL and 18.5% compared to her baseline 10.9 g/dL and 35.5%. She reported a history of iron deficiency after a cesarean section in 2019 although her iron level had since normalized, and she was not on any long-term iron therapy. Laboratory studies showed no evidence of hemolysis and hemoccult testing was negative. She was discharged after receiving 3 units of packed red blood cells and 2 doses of iron sucrose injections with an improved haemoglobin of 8.5 g/dL with daily ferrous sulfate supplementation. She reported overall improvement in energy level and was scheduled to follow up with gastroenterology (GI) for an esophagogastroduodenoscopy and colonoscopy. Her haemoglobin was followed up outpatient one month later with steady improvement reaching 11.4 g/dL.

The patient followed up with a gastroenterologist 3 months after hospital discharge, and on EGD a 5 cm malignant appearing mass was discovered partially obstructing the gastric antrum. The biopsy report was consistent with low-grade epithelial mesenchymal neoplasm with features of a glomus tumor, with a Ki-67 of 20%. A follow up computed tomography (CT) scan showed a 5 cm relatively homogeneous solid mass in the gastric antrum with concern for a GIST tumor, and an incidental right breast fibroadenoma (Figure 1). MRCP and MR abdomen were obtained to better characterize the lesion and its relationship to the porta hepatis, which demonstrated a 4.9 × 5.1 × 4.4 cm circumferential solid mass at the gastric antrum with enhancement and mass-effect on the pancreas, porta hepatis, first portion of the duodenum, and SMV. The liver demonstrated no focal lesions and there was no evidence of metastatic disease on CT or MR imaging. At a follow up surgical consultation, she endorsed early satiety, nausea, difficulty taking intake by mouth, and recent weight loss. On physical exam, a non-tender palpable mass was noted in the epigastrium. The patient was scheduled for a distal gastrectomy with reconstruction.

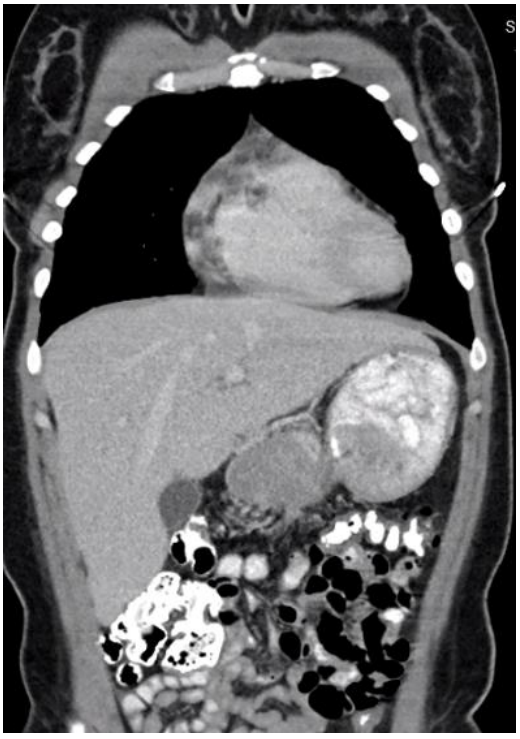

Figure 1: Computed tomography chest/abdomen/pelvis findings with IV and oral contrast showing a roughly 5cm solid mass in the gastric antrum. Radiographic differential favoured a GIST with gastric adenocarcinoma or lymphoma possible but less likely.

Intraoperatively, the gastric mass was found to invade the anterior portion of the antrum extending through the transverse mesocolon superior to the SMA, with a 1 cm mass in segment 3 of the liver. The mass was confirmed with intraoperative ultrasound and biopsy was sent for frozen section pathology. The pathology findings returned with features consistent with a glomus tumor. Due to this unexpected involvement of the mass, permission from next of kin was obtained intraoperatively to perform an en bloc colectomy and a wedge liver resection. Resection and reconstruction were performed as follows: distal gastrectomy, antrectomy, duodenectomy en bloc with transverse colon, partial omentectomy, and segment 3 liver wedge resection, with roux-en-y reconstruction and primary anastomosis of the transverse colon. Post-operatively the patient recovered well with no complications and was discharged on post operative day 6.

Pathology report confirmed a 5.7 × 5 × 4.3 cm malignant glomus tumor with 0/18 lymph nodes involved with pT2B pN0 pM1 with positive glomus liver metastasis; 4 mitosis per 10 hpf, grade 1. Immunohistochemistry markings were positive for smooth muscle actin and negative for synaptophysin, chromogranin, DOG1, CD 117, Pan-CK, CK7 and GATA 3 with Ki67: 5%. The College of American Pathologist's protocol for soft tissue tumor (abdominal organs) was used for staging.

Follow up for this patient included a postoperative repeat CT scan of the abdomen as well as a CT chest for completion re-staging. She also underwent Positron Emission Tomography/Computed Tomography from skull base to mid-thigh. Based on imaging findings there was no indication for adjuvant chemotherapy. She has remained disease free on follow up visits.

Discussion

While most glomus tumors are localized to the extremities and benign in nature, some present as deep or visceral tumors with malignant and metastatic potential. Gastrointestinal glomus tumors have been reported in the liver, colon, and peritoneum among other locations [6-8]. The most common location of GI glomus tumors is the antrum. A study by Miettinen, *et al.* reported that out of 32 cases of GI glomus tumors 31 were gastric and one was from the cecum. The most common presenting symptom in this study was severe upper GI bleeding causing anemia/weakness in 6 patients [9]. GI glomus tumors lack classic clinical and radiological characteristics and are often misidentified as GISTs preoperatively. Along with the case presented, there has been another successful preoperative diagnosis of a GGT via a pathologic examination of biopsies taken during an endoscopic ultrasound and upper gastrointestinal endoscopy, although the typical intramural location of GGTs can make this difficult with a conclusive diagnosis not made until pathologic examination of surgical specimens [10].

Due to the very few reported cases of malignant gastric glomus tumor, the criteria for malignancy of this tumor have been relatively undefined. Criteria for peripheral soft tissue malignancies are typically used in these cases. In an article published by The American Journal of Surgical Pathology, four classifications were proposed regarding atypical and malignant glomus tumors. The first being a malignant glomus tumor with criteria as follows: deep location and a size of more than 2 cm, atypical mitotic figures, or moderate to high nuclear grade and >/= 5 mitotic figures/50 HPF. The second classification is a symplastic glomus tumor, meaning a tumor with high nuclear grade in the absence of any other malignant features. The remaining two categories are glomus tumors of uncertain malignant potential and glomangiomatosis, or tumors with histologic features of diffuse angiomatosis and excess glomus cells. Metastatic disease was only seen in those tumors categorized as malignant according to this classification system [11]. In our case, while there is no high-grade atypia, atypical mitosis, or high mitotic activity, the tumor is more than 5 cm, meeting the criteria as described for malignancy. The tumor size, along with presence of lymphovascular invasion and a metastatic lesion in the liver, is consistent with a malignant gastric glomus tumor.

In a literature review published in 2018, there are only five documented cases of malignant gastric glomus tumors with metastasis [12]. It was noted that while most glomus tumors often have a benign clinical course, those with malignant features have a poorer prognosis and due to their rarity are limited in management guidelines and postoperative curative success. The surgical management is generally guided by preoperative imaging along with a heavy reliance on intraoperative findings. A 2002 study found that out of 32 cases of GGTs, most were excised by wedge or segmental resection followed by hemigastrectomy or antrectomy with subtotal gastrectomy being the least common surgical treatment, with the goal of any surgical intervention being wide local excision with negative margins [9, 13]. In terms of follow up for gastric glomus tumors there is little existing literature for recommendations for surveillance, with one case report recommending annual endoscopy based on the idea that the recurrence of GTs often occurs at the site of removal [1]. Due to the potential these tumors have for malignancy, long term follow up is generally recommended, although there is not enough information available on the postsurgical course of these patients for consensus guidelines to be determined. Most of the information available on the diagnosis, surgical intervention, and follow up of these tumors is obtained from case reports. Thus, it is important to share the details of how these patients present along with their disease course with the goal of having enough data to establish a consensus for management of these patients.

Conflicts of Interest

None.

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None.

Author Contributions

Contributions to this case report are as follows: EC collected data, wrote the manuscript, and generated figures. LK collected data, wrote and edited the manuscript. MH provided the case report topic, wrote and edited the manuscript, obtained consent, and supervised the report.

Consent

Written and signed consent obtained from the patient for the publication of this case report and any accompanying images.

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