Case Report

Gastric Glomus Tumor

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Abstract

This case report describes glomus tumor of the stomach, a rare entity, which is a mesenchymal origin tumor. They are generally benign and account for nearly 1% of all gastrointestinal (GI) soft-tissue tumors. They are almost impossible to diagnose preoperatively because of the lack of specific characteristics and are often mistaken for GI stromal tumors (GISTs). In our hospital setup, a female aged 24 years, presented with right lumbar abdominal pain which after a thorough workup leads to the preoperative diagnosis of GIST, and distal gastrectomy with Roux-en-y anastomosis was done. However, pathological and immunohistochemical findings done after the surgery were found consistent with the diagnosis of gastric glomus tumor. Discovered on GIST-1 (DOG-1) nonreactivity is considered for the diagnosis of glomus tumor which helps in excluding the diagnosis of GIST, which is positive for DOG-1. Hence, it was concluded that differential diagnosis on the basis of immunohistochemical findings can lead to an accurate preoperative diagnosis and further targeted surgical intervention.

Keywords: *Gastrointestinal stromal tumor, glomus tumor, mesenchymal origin*

Submitted: 19-Feb-2019
Revised : 21-Oct-2019
Accepted : 16-Dec-2019
Published : 27-Jul-2020

Introduction

Glomus tumor is a mesenchymal origin tumor composed of modified smooth muscle cells. It arises from the glomus body, which is neuromyoarterial and composed of an arteriovenous shunt responsible for the body temperature regulation.[1] These tumors are most commonly found in trachea, tympanum, kidney, uterus, vagina, and stomach. These approximately constitute 2% of all soft-tissue tumors.[2]

In the gastrointestinal (GI) system, the antrum of the stomach is the most common site where glomus tumor is found although stomach is the rare site of occurrence for such tumors.[3,4] Gastric glomus tumor accounts for nearly 1% of all GI soft-tissue tumors. They present as submuscosal mass which either projects in the lumen or out of the serosa, and hence, they are frequently mistaken for GI stromal tumor (GIST) which is more common in occurrence.[4] In general, they are considered benign, but some of them can show a behavioral pattern that is more aggressive.[3]

Clinically, the patient presents with symptoms of GI bleeding, perforation, or abdominal pain, but many patients can be asymptomatic (as seen in our index case), in which tumor is detected incidentally.[4] They are difficult to diagnose preoperatively due to lack of specific clinical and endoscopic characteristics. Diagnosis is largely dependent on pathological and immunohistochemical findings.

Case Report

A 24-year-old young femalepresented with a complaint of mild-to-moderate pain abdomen in the right lumbar region. No complaints of any vomiting, abdominal distension, black-colored stools, loss of appetite, and loss of weight were observed. On ultrasonography, the right-sided renal lithiasis of approximate 8 mm size and an ill-defined nondiagnostic growth near the antrum of the stomach was reported.

Contrast-enhanced computed tomography of the abdomen showed a well-defined smoothly marginated [Figure 1] soft-tissue density submucosal lesion in

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Access this article online

Quick Response Code:

Website: www.nigerianjsurg.com

DOI: 10.4103/njs.NJS_8_19

How to cite this article: Singh S, Kumar A, Singh V. Gastric glomus tumor. Niger J Surg 2020;26:162-5.

relation to the antrum and pyloric canal of the stomach, bulging in its lumen, as well as causing bulge of its external contour laterally [Figure 2].

Endoscopic ultrasound showed 3.8 cm × 2.5 cm-sized heteroechoic submucosal [Figure 3] solid gastric mass in the antrum. Fine-needle aspiration cytology showed paucicellular smears, and only one smear showed clusters of epithelial cells with round to oval nuclei and mild anisocytosis. No definite malignant/diagnostic cells could be identified. Earlier, GIST was considered as a preoperative diagnosis, and the resection of the lesion was planned as findings raised concern for carcinoma. Distal gastrectomy with Roux-en-Y anastomosis was done.

Gross findings revealed a solid hemorrhagic mass measuring 3.1 cm × 3.3 cm × 3.5 cm, submucosal in the location at greater curvature near the pyloric antrum. Mucosa and serosa were found unremarkable.

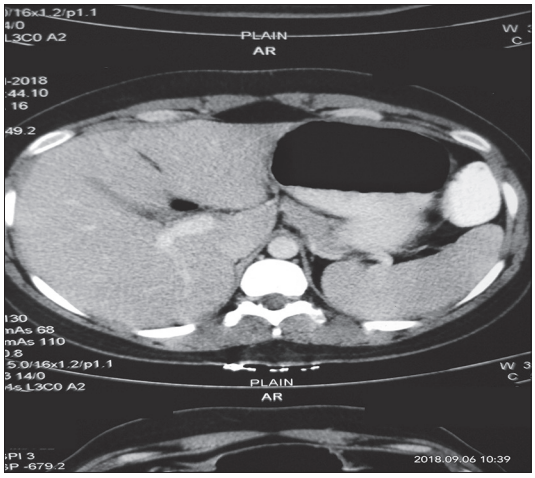

Figure 1: Contrast-enhanced computed tomography abdomen showing a well-defined smoothly marginated soft-tissue density submucosal lesion around the antrum and pylorus of the stomach

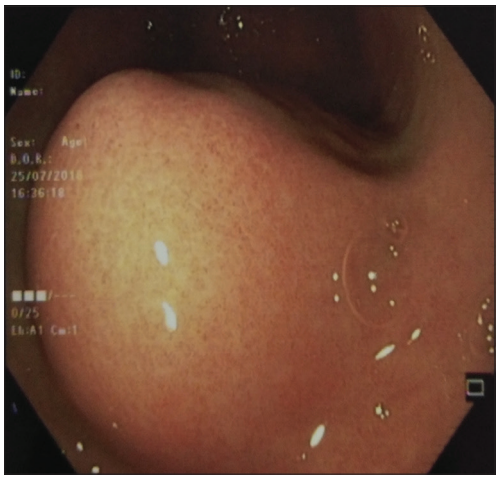

Figure 3: Endoscopic ultrasound showing heteroechoic submuscosal gastric mass in the antrum

Histopathological sections showed an intramuscular tumor composed of round to polygonal cells with moderate eosinophilic to clear cytoplasm with well-defined cytoplasmic borders, present in nests in close association with vascular channels and areas of hyalinization and calcification seen. Mucosa showed features of chronic gastritis. One reactive lymph node was identified. Considering the above features, the diagnosis of gastric glomus tumor was made.

Immunohistochemistry was performed to support the diagnosis [Figure 4]. The tumor cells showed high reactivity (4+) to smooth muscle actin (SMA), weak 1+ reactivity to Ki67, nonreactivity to CK, synaptophysin, DOG-1, and Desmin, confirming the diagnosis of glomus tumor.

The postoperative period was uneventful, and the patient was advised for regular follow-up.

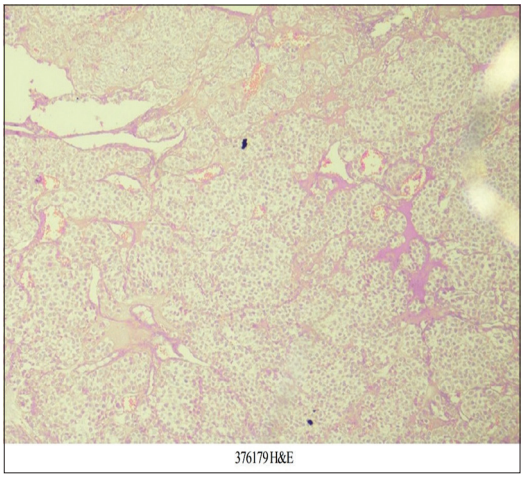

Figure 2: Histopathology section showing round to polygonal cells with moderate eosinophilic to clear cytoplasm and well-defined cytoplasmic borders, areas of hyalinization and calcification seen

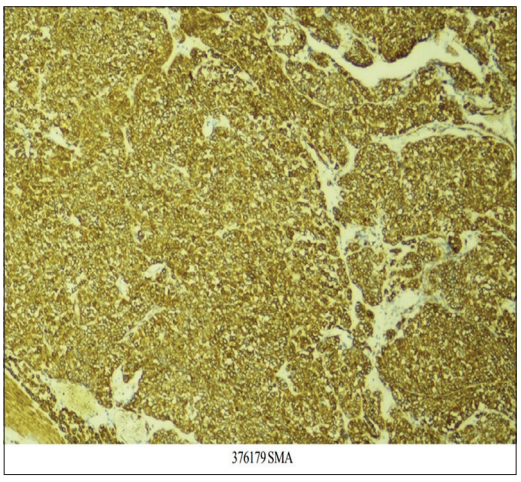

Figure 4: Immunohistochemistry panel showing high reactivity (4+) to smooth muscle actin

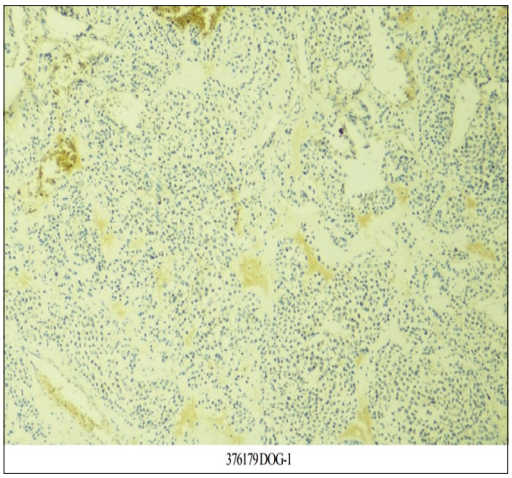

Figure 5: Immunohistochemistry panel showing nonreactivity to Discovered on GIST-1 (DOG-1)

Discussion

Intramural gastric tumors typically mesenchymal in origin include GISTs, non-GIST sarcomas, lipomas, leiomyomas, schwannomas, glomus tumors, hemangiomas, inflammatory fibroid polyps, inflammatory myofibroblastic tumors, and plexiform fibromyxomas. Glomus tumors of the GI tract (GIT) are rare, constitute around 1% of GIT soft-tissue tumors.[4] Soft-tissue tumors affect all ages and may arise at any location in the body. They account for <1% of all malignant tumors. Approximately 60% of soft-tissue sarcomas originate in an extremity (most commonly-thigh), 19% in the trunk wall, 15% in the retroperitoneum, and 9% in the head and neck.[5] GIST is the most common soft-tissue tumor of the stomach. The annual incidence of the GIST is 14.5/million. However, the prevalence of all GIST risk groups is 129/million.[6] Gastric glomus tumor occurrence is approximately 100 times less than that of GIST.[7]

Gastric glomus tumor is more commonly seen in females of the 5th–6th decades, but the presentation may vary.[8] Gastric glomus tumors can be described often as a solitary nodule, submucosal in location, and mostly found at the antrum.[8] Common presenting symptoms may vary from being completely asymptomatic to complaints of epigastric pain, upper GI bleeding, nausea, and vomiting.

The preoperative diagnosis of these tumors is often difficult due to deep-seated location along with overlapping features on imaging studies with GIST.[9] Computed tomography scan features of these tumors are nonspecific and can also be seen in other mesenchymal tumors such as schwannoma, GIST, neuroendocrine tumors, and hemangioma.[2] Endoscopy is useful in identifying origin layer of tumor, which shows a hypoechoic well-circumscribed mass in submucosa/muscularis propria.[10,11]

For the confirmation of diagnosis, surgical pathology and immunohistochemistry panel are important. Histologic features are central round to oval nuclei with inconspicuous nucleoli and clear to eosinophilic cytoplasm with distinct cell.[12] On immunohistochemistry panel, glomus tumors found strongly positive for SMA, Vimentin, calponin, type 4 collagen, and laminin.[2] DOG-1 nonreactivity is considered for the diagnosis of glomus tumor which helps in excluding the diagnosis of GIST, which is positive for DOG-1 [Figure 5]. According to recent literature, DOG-1 is considered more specific marker to diagnose GIST and it was negative in our study, so GIST was ruled out.

Criteria for malignant glomus tumors of soft tissue proposed by Folpe *et al*. include (a) deep location and size >2 cm, (b) atypical mitotic figure, and (c) moderate to high nuclear grade and mitotic activity (5 mitoses/50 HPF). For gastric glomus tumor size ≥5 cm suggests the possibility of malignancy.[11,13]

Treatment of choice for gastric glomus tumor is wedge resection with negative margins. Completely resected small tumors without necrosis and mitosis have a good prognosis. As there is a potential for malignancy, long-term follow-up and monitoring are recommended.

Conclusion

Gastric glomus tumors should be considered in the differential diagnosis of gastric submucosal tumors. It will help in diagnosis and targeted surgical intervention.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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