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Pathology

A RARE CASE REPORT OF GASTRIC SCHWANNOMA – A CLINICOPATHOLOGICAL PERSPECTIVE AND REVIEW OF LITERATURE

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ABSTRACT Schwannoma is a slow growing, benign tumor of neural origin. They are commonly found in the neck, trunk, and extremities. Gastrointestinal tract is a rare site for schwannomas. Gastric schwannomas contribute 0.2% of all gastric tumors. Despite their rarity at this site, these tumors often mimic more common conditions like gastrointestinal stromal tumors (GIST) and other spindle cell tumors, making the diagnosis challenging. Here we present an intriguing case of a 57-year-old woman whose incidental discovery of a submucosal gastric mass was initially suspected to be Gastrointestinal stromal tumor, ultimately revealed a Gastric schwannoma through histopathological and immunohistochemical analysis. This highlights the importance of accurate histopathological and immunohistochemical diagnosis in rare tumor presentations.

KEYWORDS : Gastric schwannoma, Schwannoma, Gastrointestinal stromal tumor

INTRODUCTION:

The gastrointestinal(GI) tract is an uncommon site for schwannoma and contribute 0.2% of all gastric tumors. [1,2] Females in the fifth to sixth decades of life are mostly affected. [1,3] Daimaru et al. first described this entity in 1988 based on the S-100 protein positive immunostaining that this tumor displayed .[1] The most common clinical and morphological differential for Gastric schwannoma(GS) is gastrointestinal stromal tumor (GIST). It must also be differentiated from other spindle cell tumors that may occur at this site .[2,4,5] Imaging modalities like CT, 18FDGPET and MRI are useful in detecting the exact location of the tumor. However it is the histopathological examination along with immunohistochemistry that helps to confirm the exact diagnosis .[2,3,6] The likelihood of malignant transformation in GS is low, and these tumors have excellent prognosis with complete surgical excision being curative of the tumor. [5,7] Hence it is of utmost importance to recognize these tumors and differentiate it from other tumors like GIST for which treatment options differ. In this case report, a 57-year-old female was incidentally found to have an enhancing submucosal, exophytic, extra-luminal mass lesion in the stomach raising a suspicion for GIST. Post surgery, histopathological examination with immunohistochemistry helped confirm the unexpected diagnosis of GS.

Case Presentation:

A 57-year-old female, known case of Gastro-esophageal reflux disease – Grade A, had come to the hospital with continuing complaints of reflux symptoms. The physical examination and laboratory values showed no abnormalities. On further investigating, Ultrasound and CECT abdomen incidentally showed a well-defined, heterogeneously enhancing, exophytic, extra-luminal mass lesion measuring 1.4 x 2.1 cm noted arising from the greater curvature of the stomach at a junction of distal body and antrum suggesting a possibility of GIST (Figure 1). Following which laparoscopic wedge resection of the mass was done and removed. Pathological gross examination revealed a well circumscribed firm lesion with yellowish and pale white areas. Lesion grossly measured 2 x 1.5 x 1.3cm (Figure2). Microscopically, a well circumscribed cellular neoplasm with prominent lymphoid cuffing was seen involving the muscularis propria, comprising of interlacing bundles of spindle cells exhibiting mild to moderate nuclear atypia having elongated nuclei with ill- defined cytoplasmic borders and occasional palisading nuclei. Areas with microtrabecular growth pattern is also noted.

There was infrequent mitotic activity (Figure 3). The neoplastic cells were strongly positive for S100 and was non – immunoreactive for CD117, DOG1 and Desmin (Figure 4). Ki67 showed a proliferation index of 2%. Hence based on the histomorphology and immunohistochemistry a diagnosis of gastric schwannoma was confirmed. Post-op period was uneventful and the six months follow-up showed no recurrence or metastasis.

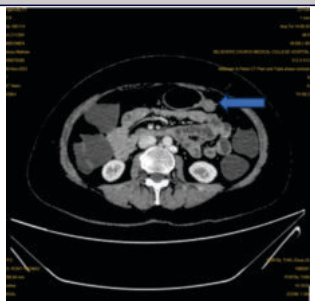
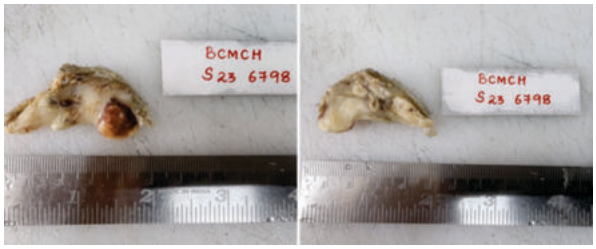
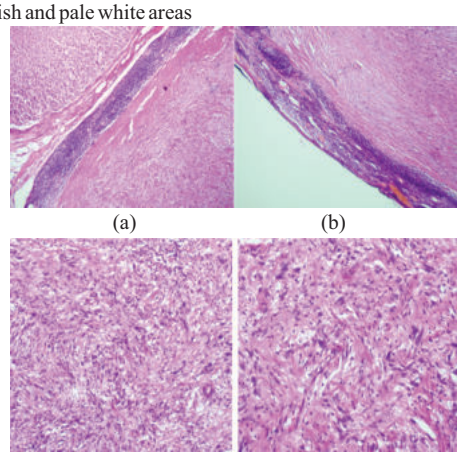


Figure 1: CECT Abdomen well-defined, heterogeneously enhancing, exophytic, extra-luminal mass lesion arising from the greater curvature of the stomach (Blue arrow indicates the mass)



(a) (b)

Figure 2: Gross image of the tumor (a) Well circumscribed lesion measuring 2 x 1.5 x 1.3cm. (b) Cut surface showing a firm lesion with yellowish and pale white areas



(a) (b)

(c) (d)

Figure 3: Histopathological examination (a) and (b) A well circumscribed lesion showing the characteristic lymphoid cuffing (hematoxylin and eosin, 4x); (c) Interlacing bundles of spindle cells with areas showing occational pallisading and microtrabecular pattern (hematoxylin and eosin, 20x); (d) Spindle cells exhibiting mild to moderate nuclear atypia with elongated nuclei with ill- defined cytoplasmic borders (hematoxylin and eosin, 4x).

Figure 4 : S100 protein showing diffuse staining of the tumor cells as a characteristic feature for schwannoma.

DISCUSSION:

Schwannoma is a neural tumor commonly seen in the head and neck region.[1,2,4] The gastrointestinal (GI) tract is a rare site for schwannoma. Gastric schwannomas (GS) contribute 0.2% of all gastric tumors, 4.0% of all benign gastric tumors, and 6.3% of all gastric mesenchymal tumors. Studies have shown that in the GI tract the most common site for GS is the stomach, predominantly involving the body (50%), antrum (32%) and fundus(18%).[4,6,8] These mesenchymal tumors arise from the submucosal nerve plexuses, predominantly involving the Auerbach's plexus more than the Meisner's plexus.[4,9] Daimaru et al. first described this entity in 1988 based on the S-100 protein positive immunostaining that this tumor displayed.[1,2] Females are mostly affected (female: male ratio of 4:1) commonly in the fifth to sixth decades of life. [4,9] However, any age group can be involved. [7] GS usually are asymptomatic, although some studies have shown that patients can present with abdominal discomfort or as a palpable mass or rarely gastric obstruction. Gastric bleed may also occur due to the ulceration caused by acid intolerance .[4–6,9] Therefore proper active intervention is required to relive the patient of these symptoms when present. [6] GS are often clinically and morphological mistaken for GIST. [2,10,11] The incidence ratio of GS versus GIST is 45:1 according to Voltaggio et al. whereas, other studies have shown ratios ranging from 8:1 to 14:1. [3,4] Imaging modalities help in providing diagnostic clues. Endoscopy and EUS FNA are helpful for the preliminary preoperative localization and diagnosis, to differentiate GS from other smooth muscle tumors. However, endoscopies may give false negative results and EUS FNA is not advised due to possibility of tumor rupture and spread which can lead to poorer prognosis [2,4,12,13] Studies have also shown that imaging techniques like CT/18FDGPET/MRI may be helpful in differentiating benign from malignant tumors but may be more useful to identifying the precise anatomic location of the tumor. On CT, GS appear more homogenous in comparison to GIST which appear heterogenous due to the presence of hemorrhage, cavitation and necrosis. Other smooth muscle tumors like leiomyoma, leiomyosarcoma also appear more heterogenous. [14] In our case, the mass had heterogenous enhancement. Few reported cases in literature have shown heterogenous enhancement on CT.[15] 18FDG-PET can show increased uptake in GS due to increased schwann cell glycometabolism, according to Beaulieu et al.[4,15] The tumor shows strong enhancement on MRI, showing low to medium signal intensities on T1-weighted scans and high signal intensities on T2-weighted images. [16,17] Imaging techniques provide the tip of the iceberg, however it is the pathological examination that helps to confirm the exact diagnosis. [6]

Grossly, these tumors range in size from 1 to 11cm.They may be located in the submucosa, muscular propria, serosa or in the subserosa. Most studies show that these tumors arise in the lesser curvature. [18,19] In our case, however, the tumor was located in the greater curvature. The submucosal location of the tumor, makes GIST a close differential for GS. However, GSs are more homogenous, firm and yellow to yellow white on sectioning whereas GISTs are predominantly heterogenous and tan-pink to hemorrhagic and may have cystic change. [3,10,20] In microscopy, GS contain spindle cells arranged in fascicular and trabecular pattern. Antoni A and Antoni B areas are seen without the presence of Verrucae bodies which is usually present in other peripheral schwannomas. There is usually no nuclear pleomorphism, increased mitosis or necrosis. Malignant tumors on the other hand, may show high mitosis (>10mitosis/hpf) and nuclear atypia. These tumors have a distinct lymphoid cuffing, that is unique to schwannomas at this site. [3,14,15,17] Confirmation and differentiation from other spindle cell tumors can be aided by immunohistochemistry. Typically, they are positive for S100, Vimentin and are negative for CD34, CD117(c-KIT), and DOG1, which are immunoreactive in GIST. In contrast to other peripheral schwannomas, GS could exhibit positive GFAP staining. Also, compared to leiomyoma, GS are negative for Smooth muscle actin (SMA) and Desmin. [3,4,21] In our case S100 highlighted the microtrabecular pattern of tumor cells by diffuse nuclear and cytoplasmic staining. CD117, DOG1 and Desmin were negative. Therefore, a diagnosis of GS was confirmed. There are fewer studies on the molecular genetics of GS. Some studies show that these tumors lack c-Kit (CD117) and PDGFRA mutations which are commonly seen in GIST. They also lack somatic NF2 gene mutations that are commonly seen in sporadic soft tissue schwannomas.[3,22] Malignant transformation oof GS is rare and accounts to about 2%. Malignant tumors may show metastasis to liver but not the lymph nodes. GS also has a low rate of recurrence that is only associated with incomplete surgical margins. Overall these tumors have an excellent prognosis with complete surgical excision being curative of the tumor. [5,7,9,10,16]

CONCLUSION:

Gastric Schwannomas are rare, benign, slow growing tumors that are commonly located in the submucosa of the stomach. Although uncommon, gastric schwannomas should be considered in the differential diagnosis, as their morphological resemblance to more common spindle cell tumors at this site can make identification challenging. While radiological modalities assist in tumor localization, definitive diagnosis is reliant on histopathological examination and immunohistochemistry. Malignant transformation is rare. Surgical excision is curative and prognosis is excellent if completely excised.

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