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Original Research Paper

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A RARE CASE OF LARGE GASTRIC SCHWANNOMA MASQUERADING AS GASTROINTESTINAL STROMAL TUMOR.

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ABSTRACT Gastric schwannomas are the rare mesenchymal neoplasm of the stomach accounting for only 0.2% of all gastric tumors. These tumors arise from the nerve plexus of the stomach and are generally located in the submucosal region. It occurs most often in the fifth and sixth decade of life with a female preponderance. Here we present a case of 55-year-old female with no other medical history presented with upper gastrointestinal symptoms for 3 months. Scan showed a large abdominal mass. The preoperative diagnosis was a gastrointestinal stromal tumor, but the postoperative pathologic and immunohistochemical examinations confirmed the diagnosis of gastric schwannoma. The patient underwent resection of the stomach mass without additional postoperative treatment, and his postoperative recovery was uneventful. Surgery is considered to be the treatment of choice and since it is a benign tumor, the prognosis is excellent.

KEYWORDS : Schwannoma, Gastric tumor, Submucosal mass, Excision.

INTRODUCTION:

Schwannoma is a benign nerve tumor originating from Schwann cells of nerve sheath (1). They are most commonly seen in extremities, head and neck location, however, their occurrence in gastrointestinal tract (GIT) is very rare. In GIT it is commonly seen in stomach (60 – 70%) followed by colon and rectum (3%) representing 2-6% of all the mesenchymal neoplasm (2). The incidence of schwannoma in stomach is 0.2% of all gastric neoplasms (3).

Daimaru et al. (4) in 1988 were first to describe the schwannoma in the GIT. In view of its rarity, they are usually diagnosed incidentally or diagnosed post operatively. They are classified under a heterogeneous group of mesenchymal or neuroectodermal neoplasms which arise from the wall of the gastrointestinal tract and include schwannomas, gastrointestinal stromal tumors (GIST), leiomyomas, leiomyosarcomas, neurofibromas, lipomas, ganglioneuromas, paragangliomas, granular cell tumors, and glomus tumors (5). Due to their benign nature, they have a good prognosis. We report of a case of 55-year-old female with gastric schwannoma, diagnosed post operatively.

Case Report:

A 55-year-old female presented to our institution with the history of dyspepsia for 3 months duration and associated black stools. There were no other comorbidities and there was no significant past history. CECT revealed an 8.6 X 6.5 X 6.4 cm mass involving the posterior wall of stomach body with an exophytic component interface between the mass, duodenum-jejunal junction and hepatic flexure of colon, suggestive of carcinoma stomach (Figure- 1). Followed by imaging, upper GI endoscopy was done, which showed a large submucosal mass with eccentric ulcer, elevated margins, slough on the base, present at the posterior wall of distal body and antrum of the stomach. Rest of the mucosa appeared to be normal. Endoscopic biopsy was reported as non-specific gastritis. Radiological and clinical diagnosis of GIST was made due to the submucosal location of the mass.

Ga68 FAPI PET/CECT done for the patient revealed increased tracer uptake in a soft tissue mass arising from the posterior wall of stomach with exophytic component with SUV max 11.10, suggestive of primary tumor (Figure- 1). No significant tracer avid abdominal nodes were noted. Patient's serum CEA level (1.99 ng/ml) was within normal limits. Later patient underwent wedge resection of the submucosal mass and the specimen was sent for frozen section.

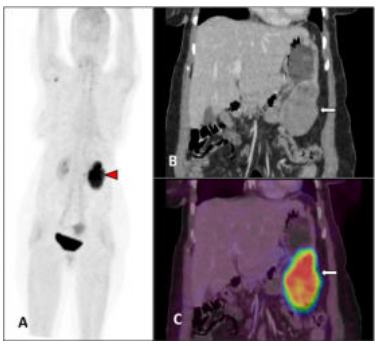

Figure1: (A) maximum intensity projection image showing intense uptake in a lesion in the left hypochondriac region (arrow head). **(B, C)** CT and fused Gallium 68 FAPi PET/CT images showing an enhancing exophytic soft tissue mass arising from greater curvature of stomach which shows intense FAP expression. (arrows)

We received a wide local excision specimen of gastric mass measuring 9 x 6.5 x 6 cm. The gastric mucosa was normal grossly. A greyish-white circumscribed solid tumor measuring 8.5x7x4.1 cm was identified in the submucosal region of the stomach (Figure- 2A). Serosal surface was unremarkable.

Frozen sections were cut, which showed unremarkable mucosal lining. Submucosa and muscularis showed a neoplasm composed of lobules of spindle cells without any significant nuclear pleomorphism. In the frozen section a diagnosis of spindle cell tumor with possibility of GIST was given (Figure- 2B).

Later specimen was grossed for further paraffin embedded sectioning and representative sections were given. Histopathology sections showed unremarkable gastric

mucosa with a well-circumscribed neoplasm in submucosa composed of spindle cells arranged predominantly in hypercellular (Antoni A) with intervening hypocellular areas (Antoni B). The cells had wavy buckled nuclei and moderate cytoplasm. Numerous Verocay bodies were noted. Mitosis was infrequent and no necrosis was identified. Since the initial diagnosis was GIST clinically, c- kit and DOG-1 immunohistochemistry was done and both the markers were negative. Next step was to rule out the smooth muscle neoplasms, for which markers like SMA, H-caldesmon was put and those markers also came negative. Finally, S-100 and SOX- 10 were put and the tumor cells were diffusely positive. In correlation with morphology and IHC final impression was given as gastric schwannoma (GS).

DISCUSSION:

Schwannoma commonly arises from the peripheral nerve of the skin but in GI tract it originates from the neural plexuses of the intestinal wall. GIT schwannomas are called gastrointestinal autonomic nerve tumors (GANTs) (6,7). Gastrointestinal schwannomas rarely occur in the esophagus and small intestine.

It involves the submucosal nerve plexuses, such as Auerbach's plexus or Meissner's plexus (8). In stomach they are frequently found in the body of stomach (50%), followed by the antrum (32%) and fundus (18%) (9). Gastric schwannoma occurs most often in the fifth and sixth decade of life with female preponderance and female to male ratio 4:1 (10). Gastric schwannomas in general present mostly with vague abdominal symptoms like abdominal pain, dyspepsia, bleeding, also palpable mass and less often as obstruction.

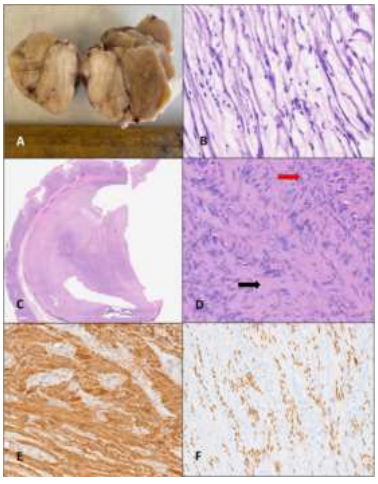

Figure 2: (A) Resected specimen with the nodular growth which on cut section shows a solid, homogenous and yellowish white surface. **(B)** Frozen section shows a mesenchymal tumor composed of lobules of benign looking spindle cells. **(C)** Microsection shows a circumscribed lesion in submucosa with unremarkable mucosa (H&E, 4X). **(D)** Lesion is composed of spindle cells arranged in hypercellular areas [Antoni A] (Red arrow) with intervening hypocellular areas [Antoni B] (Green arrow). The cells have wavy buckled nuclei, inconspicuous nucleoli and moderate cytoplasm (H&E, 40X). **(E)** The lesional cells are positive for S-100 (diffuse; strong). **(F)** Tumor are positive for SOX-10 (diffuse; strong, nuclear).

Endoscopy and radiological imaging cannot distinguish gastric mesenchymal tumors because they all present as isolated submucosal masses. Endoscopic biopsies are usually inconclusive, because of their submucosal location. In the present case the mass was seen in the submucosal site and radiologically carcinoma stomach was suspected.

On computed tomography (CT), they are well-defined lesions, rounded or lobulated, with low attenuation on unenhanced images due to their dense spindle cell composition. The closest radiological differential diagnosis for gastric schwannoma is GIST and Voltaggio et al. in their study estimated that the ratio of gastric GIST to gastric schwannoma is approximately 45 to 1 (10). The most notable difference between the two entities is the greater heterogeneity of GIST due presence of hemorrhage, necrosis, and cavitation as it is a rapidly growing tumor (5). GS is a slow growing tumor, hence there is increased vascularity. In a study done by Levy AD et al on gastric GISTs, 92% of cases had a peripheral enhancement pattern due to hemorrhage, necrosis, or cystic change within the tumor, whereas only 8% of cases showed homogeneous enhancement (5).

Surgical resection is considered as the treatment of choice in patients with gastric schwannoma. Depending upon the size, location and surrounding involvement of the tumor local excision, wedge resection, partial, subtotal or even total gastrectomy are acceptable surgical procedures with a low recurrence rate. Nowadays laparoscopic techniques are also used (11).

The characteristic features of fasciculating bundles of oval to elongated cells arranged in a parallel fashion having oval to elongated nuclei are seen in both touch cytology and frozen sections, which gives the close differentials to other benign spindle cell lesions like GIST and leiomyoma. Hence a diagnosis of benign spindled cell tumor with a suggestion of what are the possible differential diagnosis can be given as a reasonable diagnosis (17).

Schwannomas are usually diagnosed histopathologically, followed by IHC. Immunohistochemistry plays a pivotal role to differentiate among GISTs, leiomyomas, and gastrointestinal autonomic nerve tumors. Schwannomas are positive for S100 protein and vimentin but negative for DOG-1 and CD117 which are the markers positive in GISTs (12). Unlike leiomyomas, schwannomas are negative for smooth muscle actin (SMA) and H- caldesmon (13). Gastrointestinal autonomic nerve tumors are mostly positive for CD117 and CD34 and usually negative for S-100 protein and GFAP (14). They may also be positive for GFAP, which is in contrast to peripheral schwannomas (2). These markers are considered the most appropriate diagnostic indicators of gastric schwannomas. S-100 protein, which is derived from cells of the neural crest, is a unique indicator of schwannomas.

Molecular research has shown that gastric schwannomas typically exhibit a lack of c-Kit (CD117), mutation of PDGFRA (14), chromosome 22 monosomy, and polyploidy of chromosomes 2 and 18. Psammomatous melanotic schwannomas may occur in connection with Carney complex that also includes myxomas and Cushing syndrome (15). These schwannomas progressing to malignant tumor is very uncommon and the treatment included surgical excision. Post surgery majority patients have good prognosis and with complete excision, recurrence rate is reduced (16).

For the present case post operative period was uneventful and patient's symptoms were releived post-surgery. In follow up for 2 months period, patient is completely asymptomatic and follow-up scan is also unremarkable.

CONCLUSION:

Gastric schwannomas are rare spindle cell mesenchymal neoplasm which are in general not diagnosed through radiology. The clinical manifestations and radiological findings are nonspecific. Hence preoperative diagnosis of schwannoma is difficult and post operative histopathological examination is warranted. Moreover, it cannot be differentiated from the malignant neoplasms preoperatively.

Therefore, surgery is considered the most effective treatment and final HPE gives the confirmatory diagnosis of this entity.

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