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Case Report

Schwannoma stomach: a sheep in tiger's skin

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ABSTRACT

Carcinoma stomach- Captain of men of death, is the first diagnosis we have across any patient presenting with pallor, epigastric lump and loss of appetite. We present the case of a 47 year old lady with upper abdominal discomfort, anemia and a lump in the upper abdomen and family history of carcinoma stomach, suspected even intra operatively to have carcinoma or gastro intestinal stromal tumour(GIST) of stomach, to be surprised by the histopathology of a benign schwannoma. GIST, a more common pathology of stomach, has malignant behaviour upto 30%. On the other hand, schwannomas are more benign and have excellent prognosis. There lies the importance of differentiating the two.

Keywords: Carcinoma stomach, GIST, Schwannoma

INTRODUCTION

Gastrointestinal schwannomas are rare benign tumours with malignant potential.[1] The most common site of gastrointestinal schwannoma is stomach, which was first reported by Daimaru et al in 1988.[2]

They arise from the nerve plexus of the gut wall. Gastric schwannoma accounts for about 0.2% of all gastric tumours, constitutes about 4% of all benign gastric neoplasms and 0.4 to 1% of all submucosal tumours of gastrointestinal tract.[3,4]

The peak incidence is in the 4th and 5th decade of life, usually in females. They are benign, slow growing tumours, presenting with non-specific symptoms, or are asymptomatic and often detected incidentally.

Some may present with epigastric pain, bleeding or a palpable mass. Pre-operative investigation is not pathognomonic and many are therefore diagnosed as gastro intestinal stromal tumour (GIST).

Diagnosis is usually made only after histopathological examination.

CASE REPORT

47-year-old lady, coming from Northern Kerala (India) presented with dyspnoea on exertion and palpitation of 2 months duration. On evaluation from elsewhere, she was found to have anaemia (Hb=5g/dl) and was put on oral hematinics. One month later she developed upper abdominal discomfort which increased with food intake, with associated appetite loss and postprandial vomiting. Her father died of carcinoma stomach. On examination, she had pallor. A 6.5 x 5 cm, firm, non-tender, intra-abdominal mass with irregular borders and smooth surface was palpable in the left hypochondrium, extending to the epigastrium, with succusion splash. No hepatomegaly or ascites.

Ultrasound of abdomen and pelvis showed hypoechoic mass, 6.4 x 5 cm, arising from lesser curvature of stomach. CECT abdomen showed an exophytic contrast enhancing soft tissue density mass lesion, 5x6.7x6 cm

arising from body of stomach near lesser curvature with necrotic areas inside. Features suggestive of GIST (Figure 1A and 1B).

OGD scopy revealed ulcerated lesion along lesser curvature of stomach extending from antrum to body of stomach along the incisura (Figure 2).

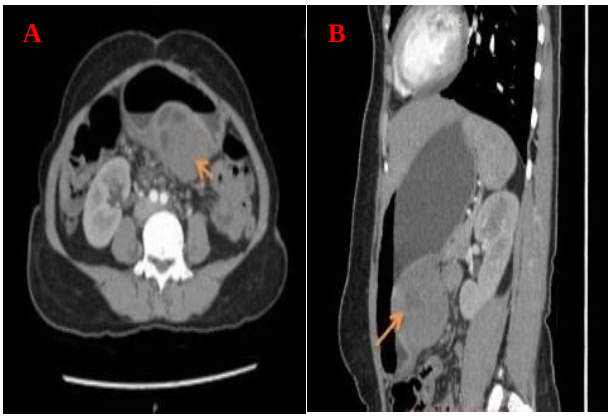

Figure 1: A) CECT abdomen - exophytic soft tissue density mass in the lesser curvature of stomach (arrow)- axial view. B) CECT abdomen - exophytic soft tissue density mass in the lesser curvature of stomach (arrow)- sagittal view.

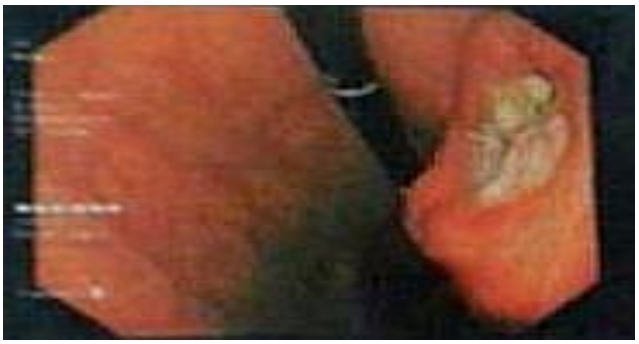

Figure 2: OGD scopy showing ulcerated lesion along lesser curvature of stomach (arrow).

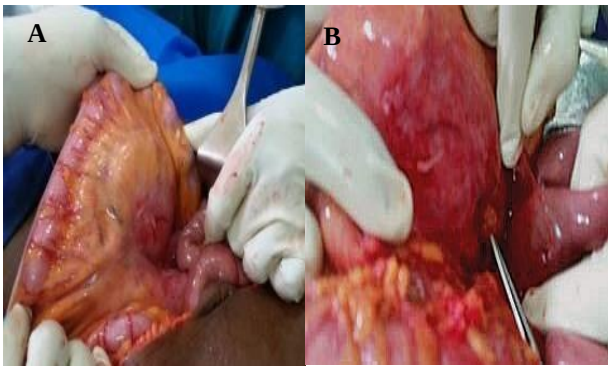

Figure 3: A) Operative finding: adherence of the tumour to the mesocolon. B) Operative finding- adherence of the tumour to the anterior aspect of pancreas.

Biopsy was not attempted due to fear of bleeding. Authors proceeded with exploratory laparotomy, during which a hard mass, 8x6x6 cm was seen arising from posterior wall of lesser curvature of stomach protruding into the lumen, adherent to transverse mesocolon and anterior surface of pancreas (Figure 3A and 3B) with minimal ascites.

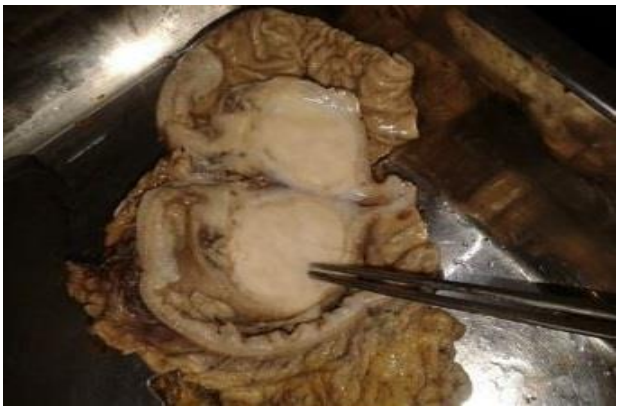

Figure 4: Gross specimen showing a polypoid submucosal growth arising from lesser curvature. Surface of mucosa showed punched out ulcer.

Liver, peritoneum and pelvis were free of metastatic deposits, without any palpable lymphnodes. A subtotal gastrectomy and transverse colectomy enbloc was done after shaving off the tumour from the anterior pancreatic capsule followed by gastrojejunostomy, jejunojejunostomy and colocolic anastomosis (Figure 4).

Microscopically, resected specimen revealed that tumour was in submucosa and muscle coat of stomach and was composed of sheets of spindle cells without nuclear atypia. Mitotic figures were scanty (<1/10 HPF). Ulceration of mucosa and necrotic area within the tumour were seen. Edges of resection, (perigastric) lymphnode and colon were free of tumour. Immunohistochemistry showed Vimentin, S-100 and GFAP (Glial Fibrillary Acidic Protein) positivity and CD117, SMA (Smooth Muscle Actin), CD34, Desmin and Cytokeratin negativity, which were consistent with gastric schwannoma. Postoperative period was uneventful. Patient was discharged on postoperative day 10.

Subsequent follow up was unremarkable. Patient had regained appetite and some weight, with a better performance score.

DISCUSSION

Schwannomas or neurinomas are spindle cell mesenchymal tumours which are generally benign and slow growing. They originate from any nerve that has a Schwann cell sheath. Schwannomas are most commonly seen in cranial vault, involving myelin forming cells of 8th cranial nerve, called as vestibular schwannoma.[5]

They occur rarely in GI tract, commonest site being stomach. Gastric schwannomas originate within the nerve sheath of Auerbach plexus or, less commonly, from Meissner plexus. They arise from the fundus, body or antrum of stomach, and are commonly intramural, however, they can be extraluminal or endoluminal. Isolated cases of malignant schwannomas have been reported.[6] It is important to differentiate between gastric schwannomas and other mesenchymal tumours, because the differential diagnosis of a submucosal gastric mass is broad, which includes GIST, leiomyoma, leiomyosarcoma and schwannoma. GIST has a higher chance of recurrence or being malignant. If GIST is diagnosed, surgical resection is required. Imatinib, a tyrosine kinase inhibitor, is used in adjuvant therapy when the GIST is completely excised and in palliative treatment for unresectable or metastatic disease.

Schwannomas are generally asymptomatic, but, in some cases, they can cause abdominal pain, discomfort or digestive symptoms. If the tumour is large and exophytic, it may present as palpable mass. In cases of deep ulceration, bleeding may be present.

Bruneton et al reviewed 112 cases of gastric schwannomas and found that majority (63%) reported gastrointestinal bleeding as the first symptom and 42% presented with abdominal pain.[7]

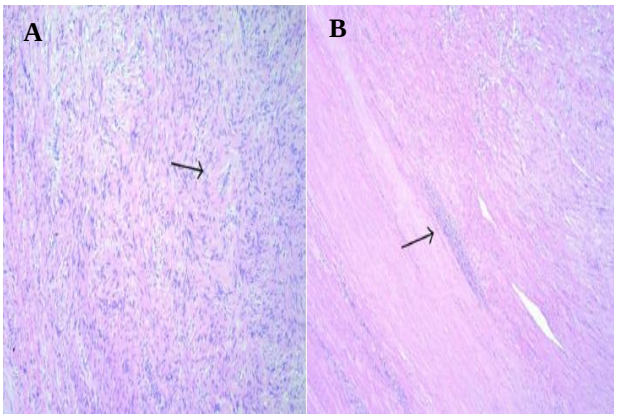

Figure 5: A) Spindle cell proliferation with relatively bland cytology and focal nuclear palisading (arrow); B) lymphocytic cuffing (arrow) at the peripheral part of the tumor is a common feature.

Definitive diagnosis of gastric schwannoma is determined by pathological examination. Preoperatively, it may be helpful to get limited information about the tumour through GI endoscopy and imaging studies. Endoscopy helps to define exact location of the tumour. Endoscopic needle biopsy is useful in diagnosing submucosal tumour, keeping in mind the risk of haemorrhage and rupture, which is associated with poor prognosis if the tumour is GIST. However, many a times, endoscopic tissue biopsies yield inconclusive results.[8]

On CT, schwannomas appear mostly as homogenous, strongly contrast enhanced tumours without signs of haemorrhage, necrosis, cystic changes or calcification, in contrast to GIST.

MRI provides further information about the exact location of tumour and its relation to surrounding structures. Schwannomas appear as strongly enhancing tumours, having low to medium signal intensity on T1 weighted images and high signal intensity on T2 weighted images. EUS is the best method to identify small lesions. Sonographically guided percutaneous core biopsy is one possible way of obtaining a precise preoperative diagnosis.[9] Microscopically schwannomas of gastrointestinal tract consist of spindle cells with a prominent lymphoid cuff (Figure 5A and 5B) and are characterized by the absence of Verocay bodies, Antoni A and Antoni B areas.

They are also GFAP positive in contrast to peripheral schwannomas. Immunohistochemical features of schwannomas are very important for differentiating them from various types of submucosal tumours like GISTs, leiomyomas and Gastrointestinal Autonomic Nerve Tumours (GANTs).

Gastrointestinal schwannomas are S-100 protein and vimentin positive, never express the CD117 antigen and are usually negative for CD34, in contrast to GISTs. Schwannomas are also negative for SMA, desmin and caldesmon in contrast to leiomyomas and leiomyosarcomas.[10] GANTs are usually negative for S-100 protein and GFAP and most are positive for CD117 and CD34.

Surgical excision is the treatment of choice. Size and location of the tumour and its relation to surrounding structure determines the type of surgery.

Local extirpation, wedge resection, partial, subtotal or even total gastrectomy are acceptable operations.[11] Laparoscopic techniques can also be used. Recurrence rates are very low. Postoperative prognosis is excellent.

The usefulness of molecular therapy or target therapy in gastric schwannoma is not clearly established. Xabier et al reported that there is high expression of platelet derived growth factor receptor(PDGFR) and c-kit in vestibular schwannoma.[12]

So, direct inhibition of these molecules using imatinib mesylate may have therapeutic applications. Hence, molecular therapy for gastric schwannoma may become an additional treatment modality in near future and requires further studies and research.

CONCLUSION

Gastric schwannomas are rare gastric neoplasms. It is one of the differential diagnoses in a submucosal gastric

lesion. Surgical resection is the treatment of choice. Prognosis is excellent after resection. No adjuvant therapies are available to date.

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