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Research Paper

A Rare Case of Gastric Schwannoma – Case Report And Review Of Literature

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Abstract
Schwannoma is a benign and slow-glowing neoplasm that rarely occurs in the gastrointestinal tract. The diagnosis of gastric schwannoma can only be made by pathological examination with positive staining for S-100 protein. A complete resection with a negative surgical margin is considered the best treatment for gastric schwannoma, with an excellent prognosis. We present a case of 41-year-old female patient who underwent laparoscopic-assisted sleeve gastric resection for gastric GIST, histopathologically diagnosed as Gastric Schwannoma, with positive staining for S-100 protein and negative for c-kit and Desmin.
Keywords: *Gastric Schwannoma, S-100, c-kit, submucosal tumours, desmin.*

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I. Introduction

Schwannoma is a slow growing benign neurogenic tumour originating from Schwann cells, which can develop anywhere along the peripheral course of the nerve. It commonly occurs in the head or neck region. It is rare in the gastrointestinal tract, however in the GI tract the most common site is stomach where they usually arise from nerve sheath of meissner's plexus. They have a benign course and excellent prognosis. Gastric mesenchymal tumours do not have a high accuracy rate on imaging and endoscopy as the findings are overlapping and non-specific. A definite distinction between different types of mesenchymal tumours can only be made by histopathological examination followed by immunohistochemistry for S-100 protein, c-kit, CD34 and smooth muscle actin [1,2].

II. Case Report

A 41-year-old female presented to the hospital with complaints of pain in left hypochondrrium, off/on fever, recurrent episodes of vomiting and weight loss for last 2 months. On examination, the vitals were normal. On per abdomen examination, the patient had mild tenderness in left hypochondrium. The gastroenterologist advised CECT abdomen. The CT scan showed a heterogeneously enhancing growth along the cardia and lesser curvature of stomach with transmural involvement. An upper GI endoscopy was planned which revealed a 5 cm polypoidal growth just below GE junction along lateral wall of stomach between lesser and greater curvature. A differential diagnosis of gastric GIST was made and the patient was advised surgical removal of the tumour. Laproscopic– assisted sleeve gastric resection was done. A large tumour measuring 10 x 5.5 x 5.5 cm was resected out. On gross examination, the tumour was well differentiated with cut surface showing greyish-white whorly appearance. No necrosis was identified. Overlying gastric mucosa appeared to be uninvolved. 5 regional lymph nodes were also received, largest measuring 0.6 cm in diameter. Histopathological examination of the resected specimen revealed a well demarcated cellular tumour consisting of cytologically bland spindle cells arranged in short fascicles. The spindle cells showed minimal pleomorphism with low mitotic activity (<5 mitosis / 50 HPF). There were focal areas of haemorrhage and myxoid degeneration. (Fig.1) A presumptive diagnosis of gastrointestinal stromal tumour (GIST) was made on H & E staining and the sections were subjected to immunohistochemical(IHC) stains to confirm the diagnosis. However on IHC, the tumour cells were negative for CD117 and DOG 1, the two hallmark markers for GIST. They also showed negative staining for Desmin and CK. The tumour cells were diffusely positive for S -100 protein, focally positive for CD 34 and vimentin. In addition, the Ki67 index was 8%. (Fig 2a-d) On the basis of the findings of IHC, a diagnosis of Gastric Schwannoma was made.

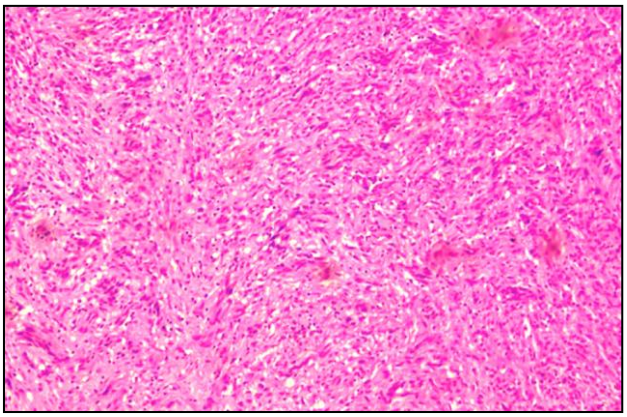

Fig 1 : H & E stained section shows whorls and fascicles of spindle cells having wavy nuclei and mild nuclear atypia. Mitosis is rare. (x 100)

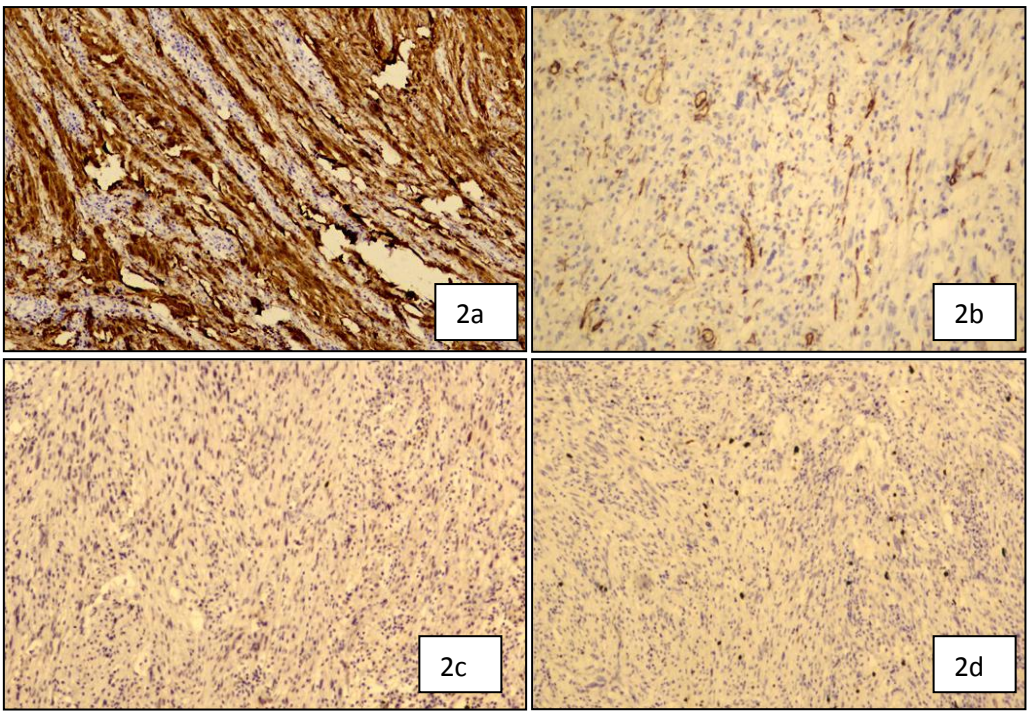

Fig 2: Immunohistochemical stains (a) S-100 – Diffuse positive; (b) CD 34 – Focal positive; (c) CD 117 – Negative (d) Ki67 – 8%

III. Discussion and conclusion

Schwannoma is a slow growing and benign neoplasm that rarely occurs in gastrointestinal tract. Gastrointestinal Schwannoma was first reported by Daimaru et al [1], who identified Schwannoma as a primary gastrointestinal tumor based on positive S -100 protein immunostaning. Gastric Schwannoma belongs to the categories of submucosal tumors (SMTs). The SMTs are divided in to three major categories – neurogenic tumours (Schwannoma, granular cell tumors and neurofibroma), myogenic tumors (leiomyomas or leiomyosarcomas) and Gastrointestinal stromal tumours (GISTs) [2].

The gastrointestinal neurogenic tumors derive from different components of nerve fibers, most commonly from Auerbach's plexus. They are mostly benign with 10% malignant transformation risk. They can be subdivided in to schwannoma, neurofibroma and granular cell tumour with schwannoma being the most common (91%) of the neurogenic tumour. Schwannomas arise from Schwann cells sheath. The most common site of GI schwannomas is stomach representing 0.2% of all gastric tumors with low malignant transformation risk [2].

Myogenic tumours include leiomyomas and leiomyosarcomas which are also rare but can be found anywhere along the GI tract. However, they are commoner in esophagus, stomach and colon and arise in muscularis mucosae or muscularis propria [2]. GIST arising from cajal cell (pacemaker cells) in GI tract are the

most common type of submucosal mesenchymal neoplasm of the GI tract. Stomach is most common location and they are commonly associated with c-kit gene mutation. GISTs are well responsive to Imatinib therapy which is a molecular inhibitor of c-kit [2,3].

For all SMTs, abdominal pain is the most common symptom [3]. GI bleeding, anemia, epigastric mass, nausea and vomiting can also occur. The upper GI bleeding is the most common symptom for symptomatic Schwannoma. Clinical picture is nonspecific and doesn't help much in distinction of gastric submucosal masses. The standard procedure for establishing diagnosis in patients with GI tumours is endoscopic biopsy. Gastric schwannoma and GISTs are grossly similar on endoscopy. In the diagnosis of SMTs, an endoscopic biopsy can lead to false negative results. Routine EUS –FNA (endoscopic ultrasound fine needle aspiration) is not recommended for primary resectable GIST by the National comprehensive cancer network (NCCN) guidelines, due to risk of tumour rupture and spread, which is associated with poorer prognosis [4].

In imaging techniques, computed tomography (CT) is a common diagnostic tool for these cases. The CT scan presentation of GISTs depends on its size, aggressiveness, and the course of the disease. Due to overlapping properties at imaging, it is impossible to distinguish GIST or schwannoma and other SMT from each other with imaging modalities. The precise preoperative diagnosis to distinguish between Gastric Schwannomma and another SMT remains difficult, even with modern imaging modalities such as CT or ^{18}F – fluorodeoxyglucose-positron emission tomography (FDG-PET). None of these modalities has shown the diagnostic features specific to Gastric Schwannoma. Histopathological analysis is must for definite diagnosis and is the gold standard [4]. GI schwannomas are capsulated tumors consisting of spindle cells with prominent lymphoid aggregations. They are characterized by Antoni A and Antoni B areas; however, there is absence of typical Verocay bodies [5]. Immunohistochemical analysis should be done to prove the probable diagnoses in all SMTs. GISTs are mostly considered CD117 – positive , generally CD34-positive, actin-positive , and S100 – negative [5,6]. In addition, Desmin staining can help in distinction between the GISTs the myogenic tumors [6,7]. Positive staining for S100 protein and vimentin and negative staining for smooth muscle actin, c-KIT, and CD34 supports the notion that the tumor is neurogenic. In our case, the tumor cells were negative for CD117, and desmin while were positive for S100.

The treatment of choice for all submucosal tumours is surgical resection [7]. However, in GISTs surgical excision is followed by imatinib therapy. The present case was operated to relieve the symptoms with a probable diagnosis of GIST. Laproscopic gastric sleeve resection was done. The definitive diagnosis of schwannoma was determined through postoperative pathological assessment and no follow up therapy with imatinib was given. Gastric Shwannoma is a benign neoplasm with no recurrence irrespective of its size but regular follow up is necessary because of very rare malignant case reports [8].

Hence, it is concluded that gastric schwannoma is a submucosal tumour which cannot be preoperatively correctly diagnosed based on imaging modalities and endoscopic biopsies. The definite diagnosis depends on specific immunohistochemical profile. The correct diagnosis of tumour is necessary as it saves the patient from Imatinib chemotherapy given for the commoner gastric submucosal lesion – GIST.

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Disclosure statement

The authors have no conflicts of interest to declare.

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