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Gastric Schwannoma: A Case Report

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Abstract

Schwannomas are mesenchymal tumors that are mostly benign and slow growing. They originate from nerve sheath cells showing Schwannian differentiation [1]. Schwannomas are usually seen in somatic soft tissues. Its incidence in Gastrointestinal tract is rare. Among gastrointestinal Schwannomas, stomach is the most common site. We are reporting a case of Gastric schwannoma.

Keywords: schwannoma, gastrointestinal schwannomas, gastrointestinal stromal tumor

Introduction

Mesenchymal tumors of gastrointestinal tract include GIST, leiomyoma, Schwannoma, inflammatory myofibroblastic tumor, fibromatosis, and inflammatory fibroid polyp. GIST is the most common among them [2]. Schwannomas of gastrointestinal tract are rare tumors that constitute only 0.2 % of all gastric tumors [3]. Male and female gender has equal sex predilection for gastrointestinal tract Schwannomas.The median age of incidence is 60 - 65 years [4].Surgical removal aids excellent prognosis and no lymph node metastasis is reported. Radiology is nonspecific and a definitive diagnosis requires immunohistochemical examination.

The clinical diagnosis was gastrointestinal stromal tumor. Here we present a rare case of gastric Schwannoma. This case also highlights the importance of immunohistochemistry in study of gastrointestinal spindle cell lesions to make a definitive diagnosis.

Case Report

A 73-year-old female was on follow up of an incidentally detected mass lesion in the posterior wall of stomach since 10 years. On routine follow up, it was found that the size has increased. The patient remained asymptomatic during the follow up period.

On Examination

Per abdomen examination shows a palpable mass in the epigastrium.

Investigations

1. CECT –Abdomen shows an exophytic well defined enhancing submucosal lesion from the posterior body of stomach closely abutting jejunal loops and body of pancreas without infiltration suggestive of GIST(FIG-1)
2. Laparoscopic Gastric wedge resection was performed, and the intraoperative findings include 6x6x4 cm Gastric GIST along the posterior stomach wall near lesser curvature of stomach. The specimen was sent for histopathological examination for confirmation of diagnosis .
3. Macroscopic examination shows an encapsulated, solid, well circumscribed grey white mass with firm in consistency (FIG-2

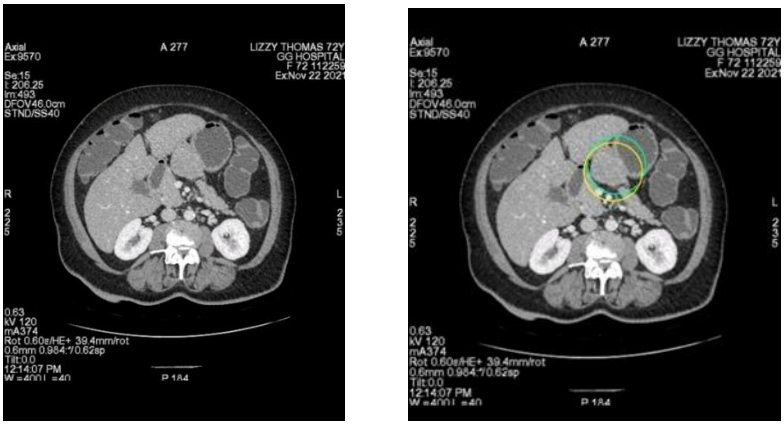

FIG.1: CECT Abdomen showing an exophytic well defined enhancing submucosal lesion

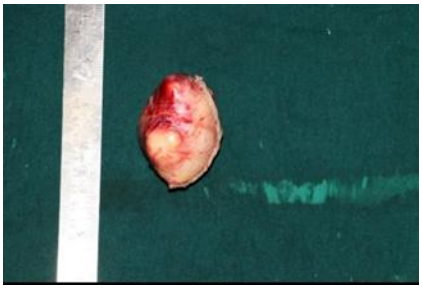

FIG. 2: Gastric wedge resection specimen

Microscopically the tumor is surrounded by gastric mucosal epithelium (FIG:3). Tumor cells are composed of spindly cells arranges as fascicles and bundles surrounded by a rim of lymphoid aggregates (FIG: 4) The individual cells are spindly with central oval to elongated nuclei. Few epithelioid cells are seen. Focal lymphoid aggregates are seen.

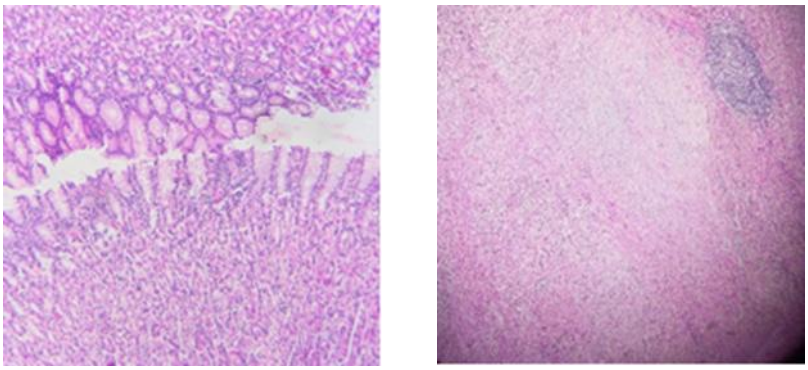

FIG. 3,4: Pathological findings

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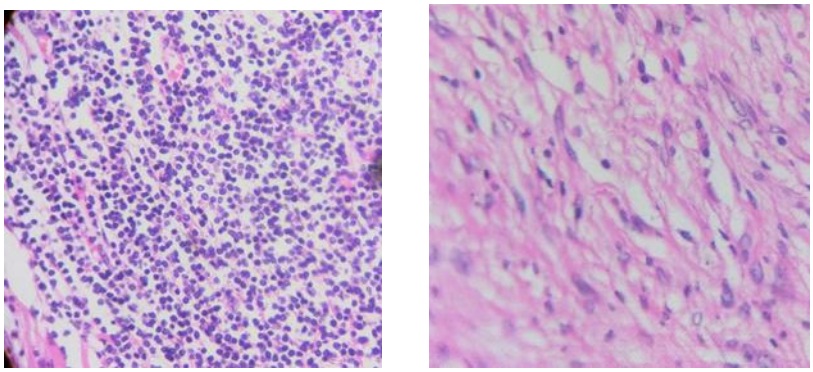

IG. 5,6: -High power photomicrograph showing lymphoid aggregates and spindle cells

Immunohistochemistry

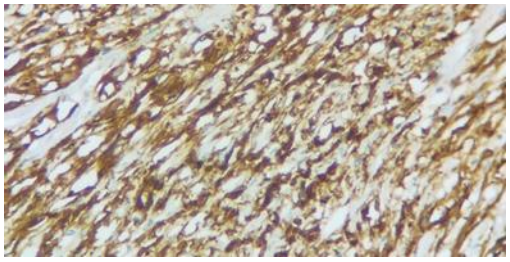

FIG. 7: Photomicrograph showing cells with diffuse and strong positivity for S100

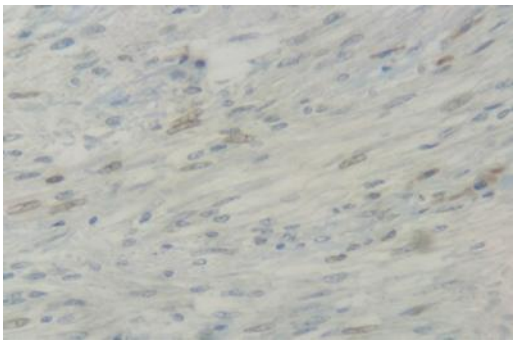

FIG.8: Photomicrograph showing negativity for DOG1

Discussion

In this case, it was an incidentally detected tumor in the posterior wall of stomach with well-defined margins and no adhesions. The tumor was clearly dissected out and histopathological examination shows features of schwannomas and was confirmed by Immunohistochemistry.

Mesenchymal tumors of gastrointestinal tract include GIST, leiomyoma, Schwannoma, inflammatory myofibroblastic tumor, fibromatosis and inflammatory fibroid polyp. GIST is the most common among them. Schwannomas of gastrointestinal tract are rare tumors that constitute only 0.2 % of all gastric tumors.

Most important differential diagnosis of a spindle cell mesenchymal tumors of gastrointestinal tract is GIST as treatment options are entirely different.

GIST arise from interstitial cells of Cajal (ICC) or ICC precursor cells [5]. The identification of *KIT* gene mutations in the majority of GIST has given GIST added importance because it has become a paradigm for targeted therapy of oncogenic proteins/oncogene addiction in solid tumors [6].

GISTs arise over a wide age range, from children to the elderly with a peak median age of 64 years at the time of diagnosis [7]. They are found along the entire length of the digestive tract but are most common in the stomach (60%), jejunum and ileum (30%),

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duodenum (5%), and colon and rectum (less than 5%) [8].

It is important to distinguish gastric Schwannoma from GIST, as the latter has malignant potential. Immunohistochemistry shows positivity for CD117 and DOG1 in GIST. CT images of Schwannomas are homogenous, well defined, intramural mass without necrosis, hemorrhage or cystic degeneration in contrast to GIST [9].

Schwannomas

A typical Schwannoma consists of spindly cells arranged in alternating hypercellular (Antony A) and hypocellular (Antony B) areas. Gastric Schwannomas are usually surrounded by intact gastric mucosa .A prominent lymphoid aggregates forming germinal centers with absence of Antony A and Antony B areas. This peritumoral lymphoid cuff is not seen in CNS or soft tissue Schwannomas. A variety of degenerative changes may be found in schwannomas including nuclear pleomorphism, xanthomatous change, vascular hyalinization, cystic change and necrosis. Some large mitotically active schwannomas lacking Antony B areas can mimic a sarcoma. Schwannomas has local recurrence if incompletely resected. Malignant transformation is extremely rare. Schwannomas can also have a predominant microcystic/ reticular appearance, these manifest a predilection for visceral location, particularly in the gastro intestinal tract [10].

Cellular Schwannoma is a term used for highly cellular Schwannomas that are exclusively composed of Antony A areas but lack Verocay bodies [11].

Psammomatous melanotic schwannoma is a distinctive type of peripheral nerve sheath tumor that occur as a component of the Carney complex. Most arise from spinal nerve roots.

The schwann cell origin of these tumors is borne out by their uniform immunoreactivity for S100. Immunohistochemistry shows S100 +,GFAP+ and negative for CD117,CD34,SMA.

Inflammatory myofibroblastic tumor

Inflammatory myofibroblastic tumor occurs primarily in the viscera and soft tissue of children and young adults. Inflammatory myofibroblastic tumors have been reported in virtually every anatomic site. The most common sites of extrapulmonary inflammatory myofibroblastic tumor are the mesentery and omentum [12] [13]. Grossly, most lesions are lobular, multinodular, or bosselated with a hard or rubbery cut surface that appears white, gray, tan-yellow, or red. Some cut with a gritty sensation because of the presence of calcifications. The tumor shows admixture of spindle cells and inflammatory cells with plasma cells. Immunohistochemistry shows negative for CD117,DOG1,CD34

Leiomyoma

Leiomyomas are benign tumors of smooth muscles. Extrauterine Leiomyomas are rarer and usually arise in the genitourinary tract (in the vulva, ovaries, urethra and urinary bladder), but may arise in nearly any anatomic site. Pilar leiomyomas arise from cutaneous erector pili muscles and rarely develop in deep soft tissue or the gastrointestinal tract.

They show spindly cells arranged in fascicles and bundles. The individual cells are plump with cigar shaped nuclei, with minimal atypia and few mitotic figures. Immunohistochemistry shows SMA+, desmin+, caldesmon H+

Fibromatosis

Although abdominal fibromatosis is indistinguishable grossly and microscopically from extra-abdominal fibromatosis, it deserves separate consideration because of its characteristic location and its tendency to occur in women of childbearing age during or following pregnancy. The tumor originate in the musculoaponeurotic structures of the abdominal wall, especially the internal oblique muscles, rectus and their fascial coverings. Fibromatosis exhibits classical histology of interlacing spindle cells with areas of hyalinization and blood vessels. It is beta catenin positive.

Another investigation that helps in the differential diagnosis of GIST and Schwannoma is mutational studies. Schwannoma has no mutational association while GIST shows c-KIT and PDGFRA mutation .

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Immunohistochemistry in the differential diagnosis of GIST

Diagnosis	KIT	DOG1	DESMIN	S-100
GIST	+++(99%)	+++(99%)	-(2%)focal	-(5%)focal
Leiomyoma	-	-	+++(100%)uniform	-
Schwannomas	-	-	-	+++(100%)uniform
Desmoid fibromatosis	-	-	-	-

Conclusion

We are reporting this case as it is a rare tumor in stomach and has to be differentiated from GIST because the treatment options are different. Gastric Schwannomas are rare spindle cell tumors with no malignant potential and good prognosis. Histopathological examination along with immunohistochemistry helps in the confirmation of diagnosis.

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