VOLUME - 10, ISSUE - 02, FEBRUARY - 2021 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Pathology

MALIGNANT GIST OF SMALL INTESTINE WITH INNUMERABLE OMENTAL TUMORLETS : A CASE REPORT

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ABSTRACT Malignant GIST is a rare type of sarcoma that is found in the digestive system , most often in the wall of stomach. Multiple GISTs are extremely rare and usually associated with type 1 NF and familial GIST. We herein report a rare case of multiple sporadic GIST in the small intestine.

KEYWORDS : GIST, Multiple, Intestine

INTRODUCTION:

GIST are derived from the interstitial cells of Cajal or their stem cell like precursors[1]. Though GISTs are the most common mesenchymal tumors of the GIT[2], multiple GISTS are extremely rare, are generally associated with carney's syndrome[3] , Pediatric GIST[4], type 1 NF[5,6] and familial GIST[7,8].

CASE REPORT:

A 60 year old who reported pain in the abdomen and weight loss for 6 month. There were no skin lesions or any familial history. USG revealed a small bowel mass along with multiple peritoneal deposits. A differential diagnosis of Adenocarcinoma with metastatic deposits was made. The findings of Barium enema were normal. The patient undergone exploratory laparotomy and the specimen of small intestine with involved segment send to the department of pathology for histopathological diagnosis. Histopathological features were consistent with multiple malignant gastrointestinal stromal tumour .

Pathological findings:

Gross findings-- Part of small gut measuring 15 cm in length with attached nodular mass measuring 12X10X8 cm and omentum measuring 28cm in length. Cut section of nodular mass shows solid cystic and hemorrhagic areas. Omentum contain innumerable nodules largest one measuring 2X1 cm in diameter. Nearest resection margin is 1.5 cm away from tumour. Another single tissue received measuring 1.5X1 cm.

Microscopical findings–

Sections from the small gut mass showed tumour composed of intersecting bundles of spindle cells with focal storiform pattern and moderate cellular and nuclear pleomorphism. The tumor had involved the mucosa, submucosa, muscularis propria and subserosa. Mitotic count > 5/20 hpf. Cystic change and heammorrhage present. Histopat hological features were consistent with malignant gastrointestinal stromal tumour of small intestine. Both resection margins were free.

All omental nodules showed similar features of GIST (32/32).

Sections from the peritoneal nodule show features of GIST. No lymphoid tissue seen.

Based on the findings of gross examination, histopathology and immunohistochemistry (CD117) the final diagnosis of multiple malignant gastrointestinal stromal tumor was made .

Fig1: Gross photograph of small intestinal mass (yellow arrow) and attached omental tissue with small multiple nodules (red arrow)

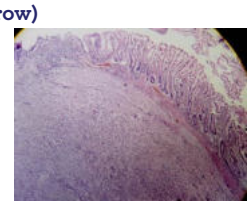

Fig.2a (4x H&E) : submucosal spindle cell tumor

Fig.2b(10x H&E): cellular tumor arranged in storiform pattern

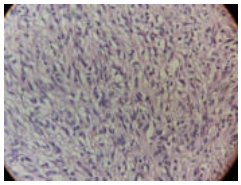

Fig. 2c(40x,H&E) spindle cell mass with moderate nuclear and cellular pleomorphism

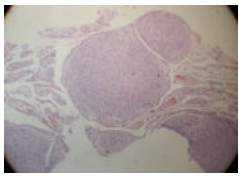

Fig 2d(4X,H&E): multiple tumourlets in omentum.

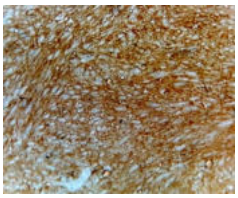

Fig 3(40x ,CD117 IHC) tumour cell showing positivity for CD 117

DISCUSSION :

Gastrointestinal stromal tumour are rare but the most common mesenchymal neoplasms of GIT and are thought to arise from intestinal pacemaker cell[9] . GIST most commonly arise from the stomach (60%- 70%)and small intestine (20%-30%)with ,10% arising from the rest of the GIT (esophagus,

mesentery, retroperitoneum)[10,11,12]. Intestinal GIST are more likely to be malignant, as seen in the present case.

Mesenteric GISTs are rare and may arise as primary lesions or as a result of metastatic spread . Usually metastatic, mesenteric GISTs are multiple and may simulate peritoneal carcinomatosis[13]. Tumours larger than 10 cm with more than 5 mitoses per 50 hpf are considered to be malignant.

GIST may present in the form of multiple tumors in the following situations ie Neurofibromatosis type 1, Carney's triad , associated with paraganglioma, or as a familial syndrome associated with germline mutation of KIT[14].

They can also appear as multiple tumors in the absence of any of these conditions. They are referred to as sporadic multiple GISTs. Those of a microscopic sizes (sometimes called tumorlets, mGISTs or seeding GISTs) are very common and predilect the upper stomach[15,16,17].

However in this case all 32 omental tumorlets were spindle cell mass, none of them showed any lymphoid tissue. Therefore it is considered as multiple tumourlets and not metastatic nodal deposits. Also it was not associated with any of the above mentioned syndromes and hence it is a rare presentation. The fact that they have different clonal molecular aberration is in keeping with an independent origin. Some have a very sclerotic backround ,suggesting spontaneous regression[18].
The IHC marker C-Kit (CD117) seems to be most specific marker for GIST.

The treatment of choice for malignant GISTS should be surgical resection, along with gene targeted therapy (inhibiting mutant

CONCLUSION-

The case has been reported because of its rarity and mimicry with primary small intestinal GIST with omental metastatic deposits.

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