Case Report

EGIST of mesentry – A case report and review of literature

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Abstract

GISTs are rare intra abdominal tumors arising from mesenchymal stromal cells of gastro intestinal tract. EGIST's are mesenchymal tumors that originate outside the GI tract and tend to have similar characteristics. To the best of our knowledge, few cases of long standing EGIST have been reported. We present a case with rare histological features of EGIST in the mesentery of a 55 years old female patient.
Keywords: EGIST, Mesentry, CD 117.

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INTRODUCTION

GIST's arise within the wall of the gut and recapitulate the phenotype of Interstitial cells of Cajal, the GIT pacemaker cell of Auerbach plexus. Because of the important biologic and therapeutic implications for this group of lesions they should be clearly distinguished from intramural tumors showing the smooth muscle differentiation as well as the occasional tumor showing other forms of lineage specific differentiation. A small number of GIST arising in soft tissue of mesentery or peritoneum are referred to as EGIST's and they account for less than 1% of all gastro intestinal malignancies.[1]

CASE REPORT

A 55 years old female presented with mass per abdomen and easy fatigability. A clinical diagnosis of mesenteric mass was done. She was operated and the specimen sent to Department of Pathology for histopathological examination.

Gross examination

We received a grey brown fleshy soft tissue mass of size 22x20x10cms. Cut surface of the mass showed solid with cystic areas. (Figure 1)

Microscopic Examination

Multiple H and E sections studied from the mesenteric mass showed sheets of monotonous round vesicular cells with eosinophilic/clear cytoplasm (Figure 2, Figure 4) divided by fibrovascular septa of varying thickness and focal pericytoma pattern (Figure 3). The histological picture taken from multiple bits, at various sites, was uniform in all sections and no other differentiation was noted. No matrix production noted. Overall picture was suggestive of small round cell tumor with a pericytoma pattern. Keeping in mind the following differentials - smooth muscle tumors, metastatic carcinomas, melanomas and GIST's immunohistochemistry was advised.

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Figure 1

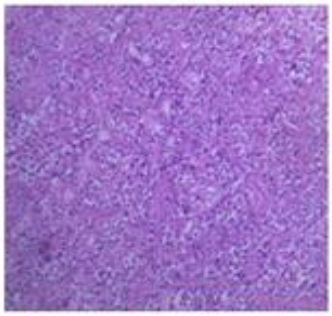


Figure 2

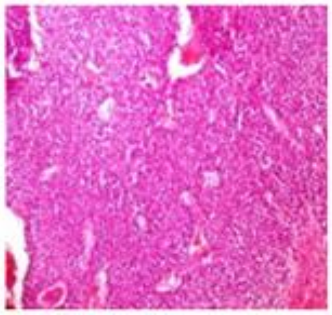
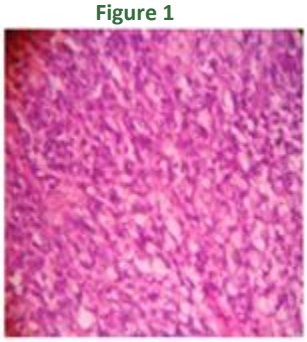


Figure 3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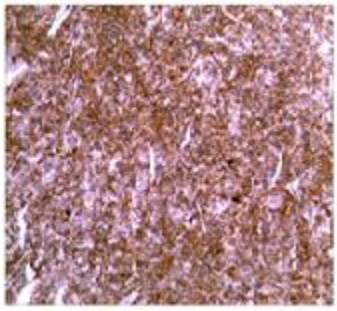
Figure 5

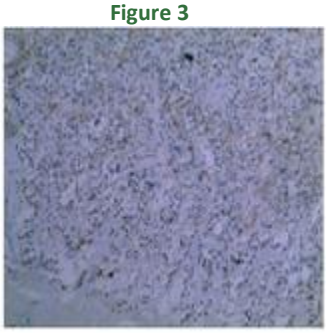
Figure 6

Legend

Figure 1: Gross showing soft tissue mass with solid and cystic areas
Figure 2: H and E 40X – Round cells with eosinophilic/clear cytoplasm divided by fibro vascular septa
Figure 3: H and E 10X - Sheets of monotonous round cells with a pericytoma pattern.
Figure 4: H and E 40 X – Uniform clear cells in sheets.
Figure 5: CD34 Diffusely Positive
Figure 6: CD117 Focally Positive

Immunohistochemistry

The immunohistochemical markers used were CD34, CD117, SMA, S100 and pancytokeratin. The tumor showed positivity for CD 34 (Figure 5) and focal positivity for CD117 (Figure 6) whereas all the other markers were negative. Hence, a diagnosis of extra gastrointestinal stromal tumor (EGIST) of mesentery was made.

DISCUSSION

GISTs are the most prevalent mesenchymal neoplasms of GIT. Annual incidence of GIST is reported to be 7-19 cases/million individuals[5] Neoplasms occurring outside the GIT particularly omentum, mesentry or retroperitoneum are defined as EGIST's, as they display no connection with gastric or intestinal wall, their incidence being < 10%.[5] EGIST in the mesentry is relatively rare with an incidence of 1%.[1] The current literature of EGIST is limited with some controversy on the origin of EGIST's. One hypothesis supports the idea that GIST and EGIST both arise from a common precursor of Intersitial cell of Cajal and smooth muscle cells due to expression of CD117 while another hypothesis asserts that EGIST's are simply mural GIST's with extensive extramural growth that eventually lost connection with gut wall.[3] But as a rule their histological appearance and immunophenotype are identical to classical GIST's[7] Most patients diagnosed with EGIST's present with abdominal pain, abdominal mass or GIT bleeding[1, 6] There is no predilection based on gender but there is for age. Most patients are diagnosed beyond middle age[6.] In our case the patient was beyond middle age, female and presented with mass per abdomen. EGIST's are classified into 3 cell types spindle cell, epitheloid round cell or mixed types, usually combination of all cell types may be seen[4] IHC findings in EGIST's is generally parallel to those of GIST. Nearly all are positive for CD 117. One half to two thirds are CD34 positive and one quarter SMA positive. Desmin and keratin are rarely present (<5%) In our case we had only epitheloid type of uniform rounded cells with eosinophilic /clear cytoplasm resembling a round cell sarcoma, epitheloid leiomyosarcoma or metastatic carcinoma. The vascular pattern in our case also mimicked a pericytoma. The pattern was uniform in multiple sections and in no place, we have noted other cell types, indicating a stromal tumor. Hence, Immunohistochemistry was suggested to know the histogenesis of the tumor which revealed CD34 positivity and CD117 focal positivity giving a final

diagnosis of EGIST. Negativity of SMA, pancytokeratin and S100 clearly rules out smooth muscle tumors, metastatic carcinomas and melanomas.

CONCLUSION

EGIST's are an aggressive group of stromal tumors with variable histologic patterns - spindle, epitheloid or mixed pattern. So whenever we come across a tumor with this pattern in single or in combination, in the mesentry and retroperitoneum, we should always keep in mind EGIST as one of our differential diagnosis, as it shows a good response to targeted adjuvant chemotherapy with tyrosine kinase inhibitors following surgical resection.

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