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Original Research Paper

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EXTRA GASTROINTESTINAL STROMAL TUMOR- A RARE CASE REPORT

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ABSTRACT

Extra-gastrointestinal stromal tumors (EGISTs) are a recently described group of tumors. A handful of less than 70 cases have been reported in English literature, so far, to the best of our knowledge. Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal neoplasms of the alimentary canal. EGISTs are a unique entity, which require distinction from GISTs because, even though, they exhibit similar histology and immunohistochemistry to GISTs, they occur outside the gastrointestinal tract, i.e. in omentum, mesentery, retroperitoneum, etc. and have different behavior patterns as far as their prognosis and management are concerned. Retroperitoneal sub-group of EGISTs is extremely rare and we report such a case of primary malignant EGIST of the retroperitoneum which presented as a mass on radiological evaluation. The tumor turned out to be a histopathological surprise, and could be distinctively labeled as EGIST only after morphological and immunohistochemical studies. It is imperative for radiologists, pathologists and oncologists, among other clinicians, to be able to recognize and understand the presentation of this group of tumors due to their rapid progression and poor prognosis, so that an early diagnosis and management may be able to improve the final disease outcome.

KEYWORDS :

INTRODUCTION

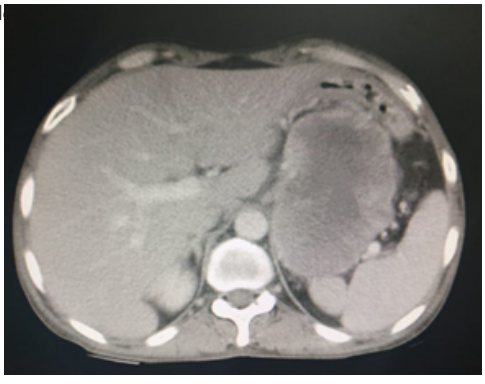
The most common mesenchymal neoplasm of gastrointestinal tract is Gastrointestinal Stromal Tumor(GISTs).[1-3] Rarely these neoplasms can occur outside the gastrointestinal tract and these are called Extra-Gastrointestinal Stromal Tumors (EGISTs).[4] A handful of less than 70 cases of retroperitoneal EGISTs have been reported in English literature, so far, to the best of our knowledge.[5,6] Many patients remain asymptomatic and their tumors are discovered incidentally at the time of other surgery or, increasingly, during imaging performed for other indications.

CASE REPORT

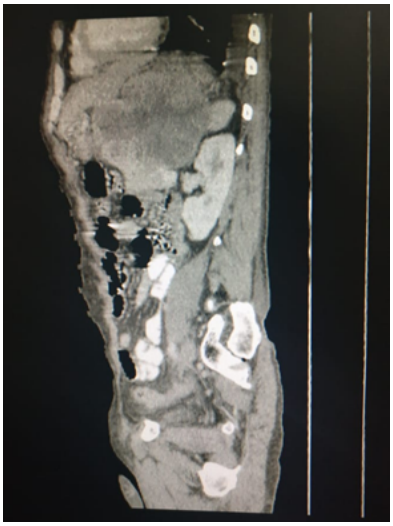
A 55 year old patient presented with gradual onset non colicky pain abdomen. On examination the patient looked healthy with no evidence of pallor or jaundice. Per abdomen examination findings revealed tenderness in the epigastrium and left hypochondrium with no evidence of palpable mass and, bowel sounds were normal. Patient had no episodes of vomiting, constipation. Per rectal examination was normal. Neck and inguinal region revealed no lymphadenopathy. Ultrasonogram of abdomen and pelvis revealed possible retroperitoneal neoplastic lesion. CECT- An ill-defined heterogeneously enhancing necrotic lesion in stomach bed region, measuring approximately 10.7 x 6.0 x 7.9 cms in size. Inferiorly it is seen extending up to body of pancreas with lost fat planes. Superiorly extending up to gastro-hepatic space. Body of stomach is seen displaced anteriorly. This suggests of neoplastic etiology, like GIST/ ?Pancreatic lesion.

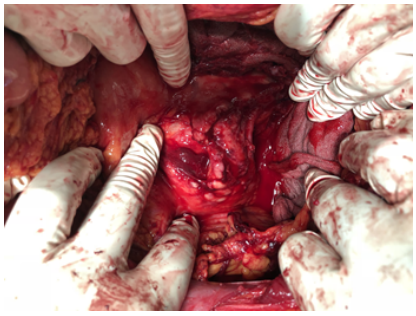
Few heterogeneously enhancing lesion scattered in both hepatic lobes- suggestive metastasis. Laboratory Investigations showed patient had microcytic hypochromic anaemia with Hb- 9gm/dl. All other routine laboratory investigations- Hemogram Liver function test and Renal function tests were normal including serum electrolytes.

The patient was electively posted for surgery with correction of anaemia. On laparotomy, a firm mass measuring 10 x 7 x 8 cm was found in the left hypochondrium in retro-peritonium with dense adhesions to posterior wall of stomach. A metastatic lesion was found in the left lobe of liver. The tumor tissue was removed in piece meal fashion and hemostasis was achieved. No perioperative complications were recorded and the patient was discharged five days after surgery. On gross examination of specimen, multiple white to grey brown soft tissue mass largest measuring 9 x 7 x 4cm noted. Multiple necrotic tissue with calcifications were noted. On microscopic examination grey brown mass shows tumor cells arranged in sheets. These tumor cells have high N:C ration with round to oval vesicular nuclei and few showing prominent nucleoli. Cytoplasm is scant to moderate and eosinophilic. Focal areas of necrosis seen. Tumor infiltrated with chronic inflammatory infiltrates. Good number of mitotic figures seen. These features suggestive of Malignant Epitheloid GIST. It was confirmed by IHC C-Kit and DOG-1. The patient was treated with Imatinib Mesylate for 3 months with regular follow-up at the hospital after which he is lost on foll

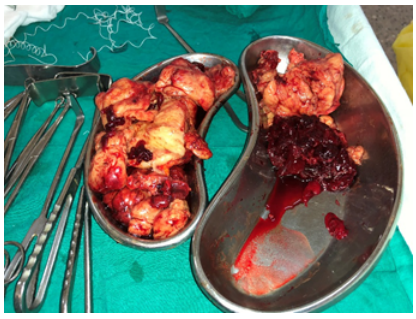


CECT Showing lesion

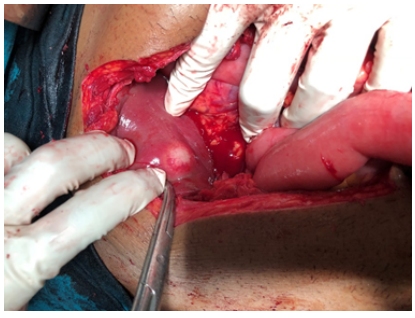




Intra-operative finding



Picture of the specimen



Metastatic lesion in the left lobe of liver

DISCUSSION

C-Kit (CD117) positive stromal tumors, although extremely rare, can involve retro-peritoneum.[1] This should be considered when imaging studies reveal an abdominal tumor involving the retro-peritoneum without a connection to the gastrointestinal tract. Secondary spread of the disease occurs in the form of hepatic and peritoneal metastases in high grade malignant tumors and carries poor prognosis.[3,7] On histopathological examination diagnosis of EGIST was confirmed by immunohistochemistry. In several cases reported in literature mass was misdiagnosed of arising from head of pancreas.[8]

As local and regional lymphnode involvement is rare in GIST, routine lymphnode dissection is not advocated and outcome depends upon pathological features of the tumor and completeness of surgical resection.[8]

CONCLUSION

Extra intestinal GIST is a very rare entity. As a result of the rarity of reports of primary

EGISTs of retroperitoneum we need to analyze the data of reported cases in the literature in order to gain a better understanding about the pathogenesis, clinicopathological features, prognosis and optimal treatment of this disease. The inclusion of this group of stromal tumors is a must on the list of differential diagnosis of solid tumors arising in the abdomen and retroperitoneum.

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