
Extra gastrointestinal stromal tumour of omentum – A case report

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Abstract

Extra gastrointestinal stromal tumours (EGIST) are rare neoplasms of the abdomen which have similar morphological and immunohistological features of gastrointestinal stromal tumours. These tumours are not connected to either stomach or intestine and most commonly arise from mesentery, retroperitoneum, omentum etc. We describe a case of EGIST arising from omentum in a 40 year old male patient who presented with features of acute intestinal obstruction.

Keywords: EGIST, Omentum, CD117.

1. Introduction

GISTs are the most common mesenchymal tumours of the GIT arising from the interstitial cells of Cajal (ICC). Most of the tumours are located in stomach and small intestine. Some of the tumours that are histologically similar to GIST are found outside the GIT and are called as EGIST [1]. The majority of the tumours arise from mesentery, omentum and retroperitoneum. The other sites include ovary, pleura, uterus and abdominal wall. Positive immunohistochemical staining for CD117 is a characteristic feature for majority of GISTs.

2. Case report

A 40 year old male patient presented with clinical features of acute intestinal obstruction of three days duration. On examination he was found to have a 9x8 cm tender mass in left iliac fossa. There was no significant past history of abdominal illness. Routine laboratory investigations were within normal limits. Plain abdominal radiograph revealed signs of intestinal obstruction. Abdominal ultrasound showed 8.7x5.9 cm size hypoechoic mass in left side of abdomen anterior to tail of pancreas and adjacent to small bowel loops. The other findings on ultrasound

were cholelithiasis, left renal cortical cyst and mild ascites.

CECT abdomen showed 10x9x7cm intensely enhancing, well defined mass with central necrotic areas and air pockets adherent to jejunal loops with minimal adjacent fat stranding (Figure 1&2). Upper GI Endoscopy revealed grade B esophagitis with hiatus hernia. Exploratory laparotomy revealed a large friable necrotic mass in the omentum with multiple small intestinal adhesions. End to end anastomosis of jejunum with feeding jejunostomy was done. On histopathology gross specimen revealed multiple grey brown soft tissue bits altogether measuring 14x11x2cms. Multiple sections were studied which revealed a cellular spindle celled lesion arranged in fascicles, interlacing bundles interspersed by areas of spotty necrosis, haemorrhages and sparse lymphocytic infiltrate. Individual cells are plump to elongated spindle cells with scanty to moderate amounts of cytoplasm with a central oval to spindle vesicular nucleus along with occasional mitotic figures. Mild nuclear pleomorphism is noted. There was no evidence of any mucin/pigment production/identifiable native tissue of either the bowel or lymph

node. In between the tumour cells sparse collagen is laid down. Features were suggestive of a Low grade Mesenchymal, spindle cell sarcoma arising from the mesentery with the possibilities of 1) Low grade Leiomyosarcoma, 2) EGIST & 3) Spindle cell mesothelioma (Figure 3). A panel of IHC marker study done revealed CD117- Diffuse and intense cytoplasmic positivity (Figure 4) & Negative staining for h-Caldesmon & Calretinin (Figure 5). Thus a final diagnosis of Extra-intestinal GIST was made.

3. Discussion

Majority of EGIST tumours are metastatic lesions of a primary GIST. 95% of GISTs carry an activating somatic mutation of CD117 [2]. CD117 is the product of proto-onco-gene c-KIT, a tyrosine kinase transmembrane receptor located on chromosome 4 [3]. The KIT proteins are detected by immunohistochemical assay for the CD117 antigen. EGISTs are thought to arise from stomach and small intestine but get separated from GIT during their development [4] and others found that multipotential mesenchymal stem cells are the cell of origin of these tumours[5].

Miettinen *et al* compared the cases of omental and mesenteric EGISTs and found that omental EGISTs have a better prognosis than mesenteric EGISTs. Reith *et al* in the report on clinicopathological and immunohistochemical features of 48 EGISTs concluded that tumour size has no association with the final outcome of the patients. Yamamoto *et al* in the study of 39 cases of EGIST suggested the molecular characterization of GIST as a prognostic factor.

There are many reports of primary omental GISTs in the literature [6-9]. Though the age

incidence in all published series is above 30 years, Mario reported a large EGIST from omentum in a 27 years old male which was surgically removed. Christian Franzini reported a case of EGIST from greater omentum in a 74 years old man and concluded that EGIST of greater omentum are slow growing tumours without clinical appearance. Dedemadi *et al* also reported a case of large EGIST from lesser omentum which was asymptomatic. Loiy *et al* reported a case of EGIST in the abdominal wall and proposed that it should be considered in the differential diagnosis of solid mass of the abdominal wall.

Barros A retrospectively analyzed 142 patients of GIST of which 9 were cases of EGIST arising from visceral sites like pancreas, mesocolon, spleen, pelvis and retroperitoneum. Other rare sites described in the literature include mesocolon transversum [10], pelvis [11]. Multiple recurrent EGISTs were also reported [12].

In the present case the patient came with acute intestinal obstruction which was a rare unusual presentation. These tumours have to be differentiated from other tumours like melanoma, desmoid tumours in which CD117 expression is detected. As per the national institute of health (NIH) modified classification these tumours have to be considered as high risk tumours as the size of the lesion was >5cms.

4. Conclusion

GISTs are not uncommon tumours and EGIST has to be considered in the differential diagnosis of solid masses of omentum or mesentery in an adult patient and pertinent IHC marker has to be done for accurate diagnosis.

Figure 1 & 2: CECT Abdomen showing heterogenous enhancing mass in the mesentery in close proximity with small bowels.



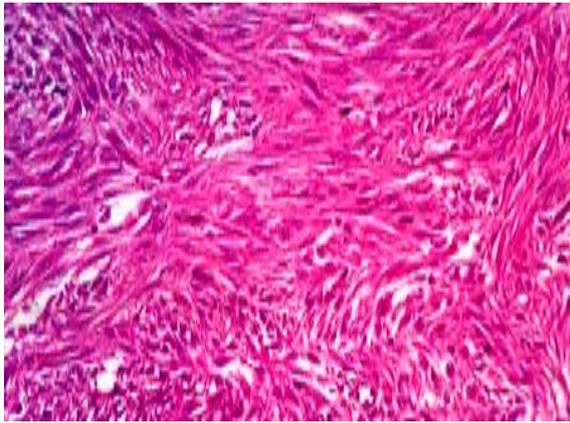
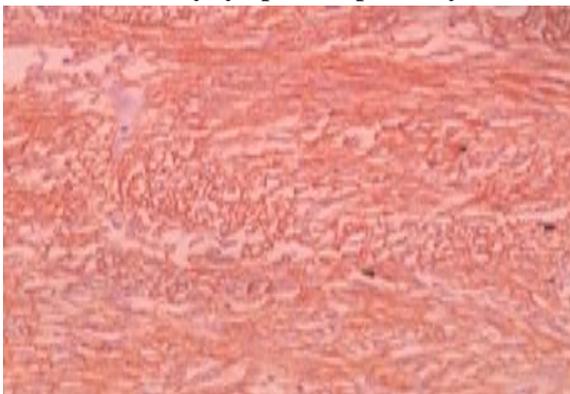
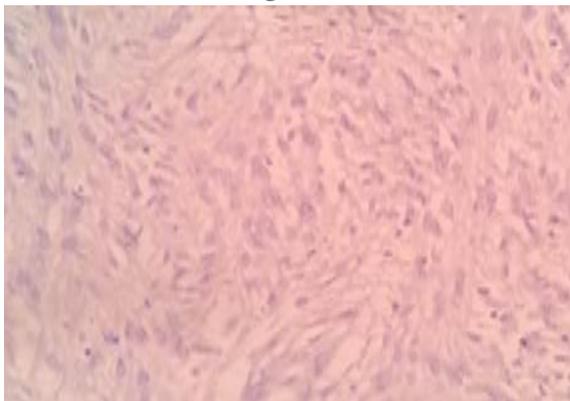
Figure 3: 40x View - H&E Stained section

Figure 3 showing Spindle celled tumour arranged in fascicles and interlacing bundles. Cells are having moderate amount of eosinophilic cytoplasm with elongated nucleus with blunt ends. There is mild nuclear pleomorphism and occasional mitosis.

Figure 4: 40x View – IHC: CD117 Staining: Diffusely cytoplasmic positivity.**Figure 5: 40x View – IHC: H-Caldesmon Staining: Negative.**

References

- [1] Reith JD, Goldblum JR, Lyles RH, Weiss SW: Extragastrointestinal (soft tissue) stromal tumors: an analysis of 48 cases with emphasis on histologic predictors of outcome. *Mod Pathol* 2000, 13:577-585.
- [2] Agaimy A., Wunsch P. Gastrointestinal stromal tumors: a regular origin in the muscularis propria, but an extremely diverse gross presentation. *Langenbecks Arch Surg.* 2006; 391(4):322–329.
- [3] Blackstein M.E., Rankin B., Fletcher C., Heinrich M., Benjamin R., Mehren M.V. Clinical benefit of Imatinib in patients with metastatic GIST negative for the expression of CD117 in the S0033 trial. ASCO. *J Clin Oncol.* 2005;9010
- [4] Miettinen M, Lasota J: Gastrointestinal stromal tumors – definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. *Virchows Arch* 2001, 438:1-12.
- [5] Li Zy, Huan XQ, Liang XJ, Li ZS, Tan AZ: Clinicopathological and immunohistochemical study of extra-gastrointestinal stromal tumours arising from the omentum and mesentery. *Zhonghua Bing Li Xue Za Zhi* 2005, 34:11-14.
- [6] Todoroki T, Sano T, Sakurai S, Segawa A, Saitoh T, Fujikawa K, Yamada S, Hirahara N, Tsushima Y, Motojima R, Motojima T: Primary omental gastrointestinal stromal tumor (GIST): Case report. *World J Surg Oncol* 2007; 5:66.
- [7] Castillo- Sang M, Mancho S, Tsang A W, Gociman B, Almaroof B, Ahmed M Y: A malignant omental extra gastro intestinal stromal tumor on a young man: a case report. *World J Surg Oncol* 2008; 6:50.
- [8] Franzini C, Alessandri L, Pisciole I, Donato S, Faraci R, Morelli L, Del Nonno F, Licci S: Extra gastrointestinal stromal tumor of the greater omentum: a case report: *World Journal of Surgical Oncology*, 2008; 6:25.
- [9] Basnet S: Extra-Intestinal Gastrointestinal Stromal tumor of Omentum: a case report: *Journal of Pathology of Nepal* (2014) vol.4, 682.
- [10] Kolarik J, Drapela J: Extragastrointestinal stromal tumor (EGIST) – a case review: *Rozhl Chir.* 2012 Apr; 91(4); 241-5.
- [11] Bugalia R P, Jenaw R K, Salvi A, Saxena J, Kankaria J: Extra Gastrointestinal tumor: A Case Review of Pelvic GIST, *Sch J Med case Rep* 2014; 2(9): 645-647.
- [12] Nayak S R, Nagakishore M G, Satyaprakash S, Sarma Y Y, Ram J K: Multiple and Recurrent Extra intestinal Gastrointestinal Stromal Tumor (EGIST) in omentum and peritoneum- a case report: *IOSR Journal of Dental and Medical Sciences* 2013; 8(3): 67-69.