IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861. Volume 8, Issue 3 (Jul.- Aug. 2013), PP 67-69
www.iosrjournals.org

Multiple and Recurrent Extra Intestinal Gastrointestinal Stromal Tumor (EGIST) in omentum and peritoneum- a case report

[1]Samir Ranjan Nayak, [2]M G Nagakishore, [3]S Satyaprakash, [4]Sarma Y Y, [5]Ram J K

Department of Surgical Oncology. GSL medical college and hospital NH-5,Lakshmipuram,Rajahmundry EG dist.AP.533296

Abstract: *Multiple and Recurrent Extra intestinal gastrointestinal stromal tumor (EGIST) in omentum and peritoneum - a case report*

Introduction: *Gastrointestinal tumors are the intraabdominal tumors arising from muscular propia of gastrointestinal tract. Similar morphology, histopathological tumors rarely may arise from peritoneum, mesentry, or omentum and these tumors are called extra intestinal gastrointestinal stromal tumors.*

Case Presentation: *45-year male presented with lump over left flank and right hypochondrium region without bowel, bladder and biliary symptoms. Abdominal sonogram and computerized tomography of abdomen suggested two large tumors located separately at right sub-diaphragmatic and left lumbar regions. The pre-operative needle aspiration cytology was inconclusive.*

DISCUSSION*: Diagnostic laparoscopy showed two tumors situated at different locations. Tumor at the right sub-diaphragmatic region was arising from peritoneum and another large tumor at the lumbar region from omentum. Excision biopsy and further immunohistochemistry revealed extra intestinal GIST*

Conclusion: *Extra intestinal gastrointestinal stromal tumors arising from peritoneum and omentum in the same patient is rare occurrence.*

Key Words: *Extra intestinal gastrointestinal stromal tumors, omentum, peritoneum*

Submitted Date 05 July 2013 **Accepted Date: 10 July 2013**

I. Introduction

Gastrointestinal tumors (GIST) are the mesenchymal tumors primarily originating in the muscularis propria of stomach, small bowel and rectum. Similar pathological tumors arising from the omentum, mesentry and peritoneum are seen in less than 5% cases and called as extra intestinal GIST (EGIST). As the tumors are not connected with the gastrointestinal wall –the term EGIST is used. The true incidence and clinical behavior of EGISTare not yet completely defined.

Here we are presenting a case of recurrent multiple extra intestinal GIST arising simultaneously from omentum and peritoneum

II. Case Presentation

45-year-old male noticed a lump over left upper abdomen in December 2012. He noticed another lump over right upper abdomen in January 2013. Besides abdominal discomfort, the patient gave no history of anorexia, fever, vomiting, jaundice, pain abdomen, hematemesis or malena. Bladder habit was normal. He had undergone abdominal surgery 5 years back for similar complaint of lump abdomen but did not receive any postoperative adjuvant therapy. At admission, his vitals were normal. Inspection of abdomen revealed a right paramedian scar. A mobile non-tender lump with well-defined borders of size 10 cm x 10 cm was palpable over left loin. The lump was moving in all directions. The second mass was palpable at right hypochondrium region with variegated consistency. The mass was indistinguishable from liver enlargement. Abdominal sonogram revealed large mass lesions in the right sub-diaphragmatic region and left lumbar region. CECT abdomen suggested enhancing hypo dense lesion of size 15 x 15 cm displacing liver infero-medially with maintenance of fat planes. The second lesion was mixed dense lesion with cystic areas in left lumbar and Para-umbilical areas with loss of fat planes with adjacent bowel loops and sigmoid colon and of size 13.9 x10.3 cm. There was no abdominal lymphadenopathy or ascitis. Ultrasound guided cytology aspiration report was inconclusive. Diagnostic laparoscopy and then laparotomy was done. The findings were (1) a tumor from the peritoneum at right sub-diaphragmatic region extending upwards. [figure1], (2) large mobile encapsulated tumor embedded in omentum [figure 2]. The tumor embedded in the omentum was completely removed by laparotomy [figure 3]. Biopsy from the tumor situated at the sub- diaphragmatic region was done as for complete tumor removal thoracotomy approach was required and consent was not given by the relatives. Post-op histopathological

examination reveled gastrointestinal stromal tumor with high mitotic index. The immunohistochemistry was positive for CD117.

The diagnosis of Extraintesinal GIST was made with stage as r Δ (omentum) ◊ (peritoneum) -T4 N0 M0 G1/2 – III A/B and R 2 resection. The patient was started with tab IMATINIB 400mg orally once daily and the response assed the after 3 months. The CECT chest and abdomen showed no residual lesion. The patient is kept on observation with IMATINIB therapy.

III. Discussion

Gastrointestinal tumors originate from interstitial cells of Cazal situated in muscular propria of intestine or their stem cell like precursors. The varied morphology of GIST is spindle cells (70%), epitheloid cell (20%), mixed spindle, and epitheloid cell (10 %) [1, 2]

The usual locations of GIST are stomach, small intestine and rectum. The tumor may arise in gall bladder, pancreas, retro peritoneum, and paravaginal or periprostatic tissues. In less than 5 % cases, the tumor arises in omentum, mesentry or peritoneum and called as extra intestinal GIST. 95% cases of GIST are positive for CD117 antigen, an epitope of the KIT receptor tyrosine kinase. 85% tumors contain oncogenic mutation in one of two tyrosine kinase receptors: KIT or PDGFR [1, 2]

GIST occurring in the stomach or small bowel usually has symptoms of pain, abdominal discomfort or upper GI bleed. Palpable mobile lump without any GI symptoms usually gives a differential diagnosis of mesenchymal tumor, omental cyst, leiomyoma / leiomyosarcoma, low grade abdominal lymphoma. [3]

Contrast tomography (CECT) is the preliminary investigation for such abdominal lump located at multiple sites. For mesenchymal tumors CECT helps in exclusion of differential diagnosis, surgical planning, staging and postoperative follow-up. Tumor size more than 5 cm, irregular surface, loss of fat planes, hepatic metastasis in CECT points to malignant mesenchymal tumor. In the presented case, the tumor size was more than 10 cm in size at two locations with irregular surface and increased vascularity suggesting malignant abdominal mass. [4]

Tissue diagnosis for an intraabdominal lump needs FNAC/BIOSPY. A pre-operative diagnosis of EGIST makes the surgery more careful with less risk.[5] In our case, the preoperative FNAC was inconclusive. Diagnostic lap was performed for tissue diagnosis and feasibility of surgery. Surgical resection with negative margins is the mainstay of treatment for GIST/EGIST [9] but if the residual disease is there, then IMATINIB should start immediately. We did R2 resection only, hence Imatinib therapy was started from the 7^{th} postoperative day.

Tumor size and mitotic index are used as the guide to predict malignant potential of stromal tumors. [6, 7, 8] Tumor size more than 10 cm with high mitotic index is categorized under high-risk stratification. The previous history of surgery for abdominal lump, CECT abdomen and post operative biopsy categorized our case as recurrent, multiple extra intestinal GIST.

IV. Conclusion

The mesenchymal tumor may grow to a large size without many symptoms. Presentation of mobile abdominal lump with prior history of surgery for abdominal mass, the mesenchymal tumor is the first provisional diagnosis. Preoperative diagnosis was difficult and sometimes surgeons are forced to do diagnostic lap/ exploratory laparotomy. Complete enbloc resection of the tumor is the treatment of choice followed by adjuvant Imatinib if required.

References

[1]. Nancy S Behazin MD,Julian Katz ,MD .Gastrointestinal stromal tumors.available at http://emedicine.medscape.com/article/179669-overview.acessed march11 2013

[2]. Zhao X, Yue C. Gastrointestinal stromal tumor. *J Gastrointest Oncol*. Sep 2012;3(3):189-

[3]. Pidhorecky I, Cheney RT, Kraybill WG, Gibbs JF. Gastrointestinal stromal tumors: current diagnosis, biologic behavior, and management. *Annals of Surgical Oncology*. 2000;7(October (9)):705–712208. [Medline].

[4]. G Jindal, R Rastogi, S Kachhawa, GL Meena. CT findings of primary extra-intestinal gastrointestinal stromal tumor of greater omentum with extensive peritoneal and bilateral ovarian metastases. Indian journal of cancer 2011;48(1): 135-137

[5]. Nakagawa M, Akasaka Y, Kanai T, et al. Extragastrointestinal stromal tumor of the greater omentum: case report and review of the literature. *Hepatogastroenterology*. May-Jun 2003;50(51):691- [Medline].

[6]. Ilona D. Goukassian,S.R. Kussman,Y. Toribio,Jennifer E. Rosen, Secondary recurrent multiple EGIST of the mesentary: A case report and review of the literature.International journal of surgery case report 2012 ;3 (9):463-66

[7]. 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. *Modern Pathology*. 2000;13(May (5)):577–585

[8]. Claudio Casella, Vincenzo Villanacci, Filippo D'Adda, Manuela Codazzi, Bruno Salerni . Primary Extra-gastrointestinal Stromal Tumor of Retroperitoneum Clinical Medicine Insights:oncology 2012:6 189–197

[9]. Winer JH, Raut CP. Management of recurrent gastrointestinal stromal tumors. *Journal of Surgical Oncology*.2011;104(December (8)):915–920

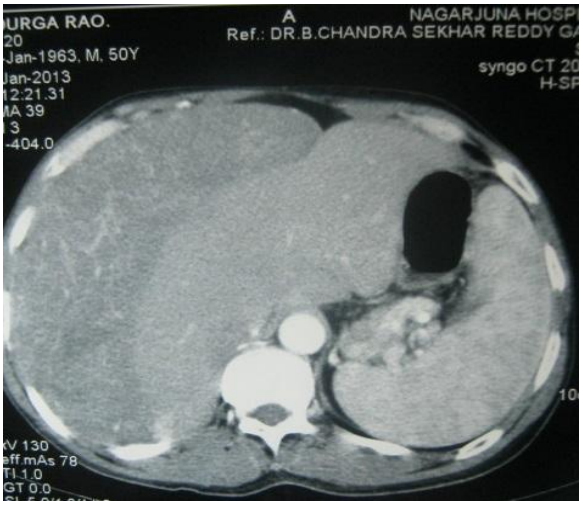

Figure1-CECT abdomen –large sub diaphragmatic tumor

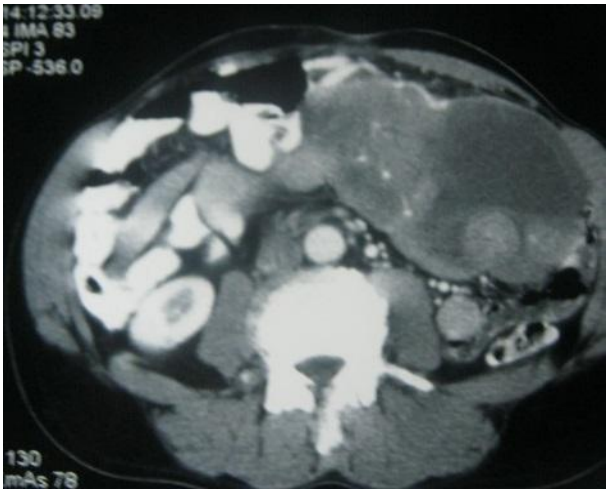

Figure 2-CECT abdomen-tumor embedded in omentum

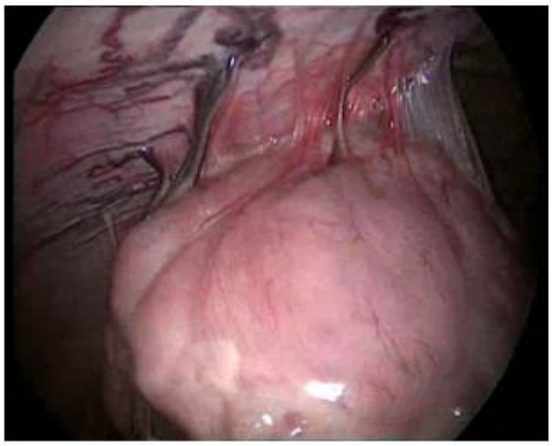

Figure-3-lap view of large sub diaphragmatic EGIST

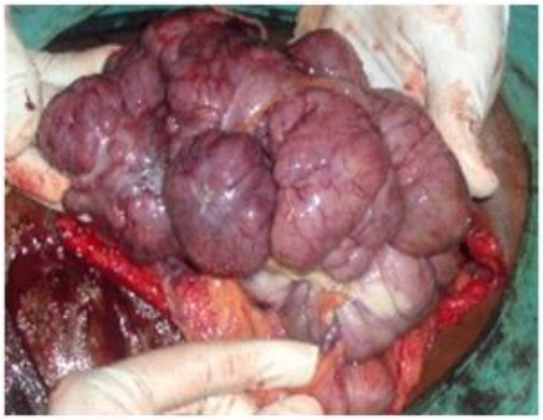

Figure-4- EGIST—embedded in omentum