# Extragastrointestinal Stromal Tumor: A Report Of A Rare Case

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**Citation**

S Sahu, P Srivastava, N Chauhan, S Kishore, D Bahl, P Sachan. *Extragastrointestinal Stromal Tumor: A Report Of A Rare Case*. The Internet Journal of Surgery. 2006 Volume 10 Number 1.

**Abstract**

Extragastrointestinal stromal tumors (EGIST) are mesenchymal tumors that occur outside the gastrointestinal tract and show histological appearance similar to the gastrointestinal stromal tumor and a positive c-kit expression. Only a few cases of EGISTs are reported. We report a rare case of extragastrointestinal stromal tumor of omentum, which was treated by surgical excision.

## INTRODUCTION

Mesenchymal tumors arising from the wall of the gastrointestinal tract have attracted a great deal of attention over the years. In the literature, different nomenclatures have been given to these tumors in the past. Leiomyoma, leiomyoblastoma and leiomyosarcoma were some of the terms used, as they were believed to originate from the smooth muscle layers of the gastrointestinal wall.[1] The term Gastrointestinal stromal tumor (GIST) was first proposed by Mazur and Clark in 1983 to describe those gastrointestinal non-epithelial neoplasms that lacked the immunohistochemical features of Schwann cells and did not have the ultra structural characteristics of smooth muscle cells.[2] Tumors previously described as gastrointestinal autonomic nerve tumor (GANT) are now regarded as a variant of GIST.[3]

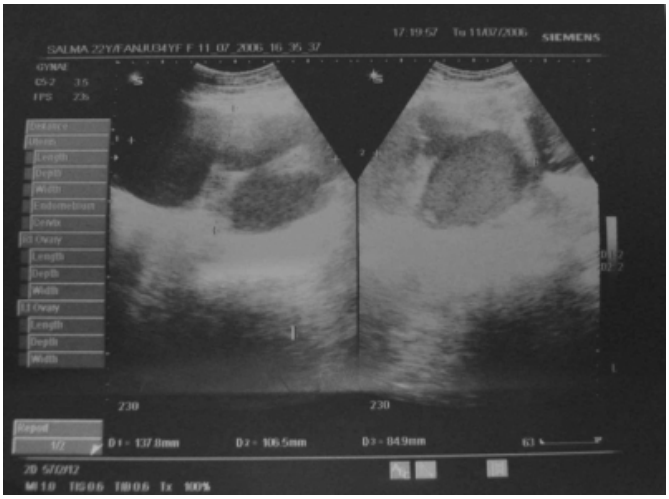
## CASE REPORT

A 22-year old female was admitted with history of severe pain in the lower abdomen since five days. At the time of admission her vital parameters were within normal limits. Examination of abdomen revealed diffuse tenderness with guarding in the lower half of the abdomen.

Investigations revealed: Hemoglobin 11.4gm/dl, Total leukocyte count 19390/cu.mm, Differential leukocyte count: neutrophils 82%, lymphocytes 12%, eosinophils 2% and lymphocytes 4%. Renal function and liver function tests were within normal limits. Imaging study in the form of ultrasonography of abdomen revealed a large right adnexal mass with predominantly multiple cystic areas of varying sizes, showing internal echoes with few solid areas [FIG-1].

**Figure 1**

Figure 1: Ultrasonography of the abdomen showing a large adnexal mass with predominance of multiple cystic areas of varying sizes with internal echoes and few solid areas.




Exploratory laparotomy was planned which revealed a large intraperitoneal mass composed of omentum and loops of ileum with few areas of cystic and hemorrhagic degenerations and a perforation in the involved loop of ileum. Resection of the involved omentum and ileum with primary end-to-end ileoileal anastomosis was done. Postoperatively the patient had an uneventful recovery.

Histopathology of the specimen showed a nodular and circumscribed smooth muscle tumor in the omentum showing whorling at few places with the smooth muscle fibres arranged in parallel bundles. [FIG-2] [FIG-3]

**Figure 2**

Figure 2: Histopathology of the omentum showing a nodular and circumscribed smooth muscle tumor with whorling and parallel bundle arrangement.

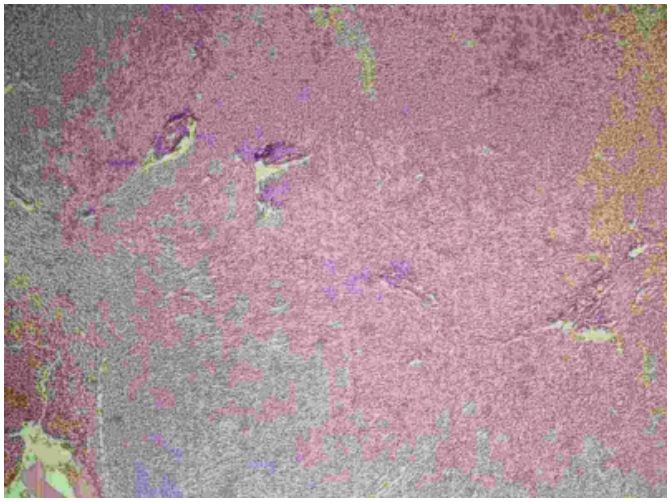
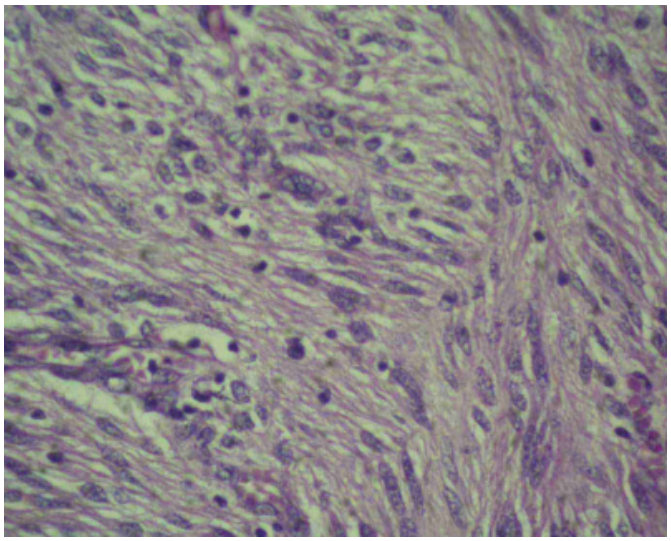


**Figure 3**

Figure 3: Histopathology of the tumor showing whorling and parallel bundle arrangement.



Immunohistochemistry analysis of the tumor tissues showed CD117 positivity, suggestive of extragastrointestinal stromal tumor of omentum. The wall of the resected ileum showed infiltration with lymphocytes, plasma cells and neutrophils with edema and fibrin deposition in the serosa.

Patient was discharged with the advice of strict regular follow-up.

Radiological imaging after six months shows no evidence of any recurrence.

## DISCUSSION

GISTs may arise from anywhere in the wall of the gastrointestinal tract, with about 60-70% occurring in the stomach.[4]

Tumors with similar features are also reported outside the wall of gastrointestinal tract primarily in the mesentery, omentum and retroperitoneum.[5] In 2000, Reith et al. termed these tumors as extragastrointestinal stromal tumors (EGISTs), which account for less than 10% of all stromal tumors of the abdomen.[6] EGISTs are also reported in liver, gallbladder, urinary bladder and vagina.[7,8,9,10] They may also present as vulvovaginal or rectovaginal septal mass.[11]

EGISTs usually affect females whose ages range from 31 to 82 years (mean, 58 years). The majority of the EGISTs arise from soft tissues of the abdominal cavity (approximately 80%) and the rest arise from the retroperitoneum.[6] Extragastrointestinal stromal tumor accounts for about 21% of all the primary omental tumors.[12]

Tumors may present as circumscribed or lobulated firm mass with or without cystic degeneration. They range in size from 2.1 to 32 cm with most of the tumor more than 5cm in size as they have enough space to grow and are usually inseparable from the wall of the stomach or intestine. Small tumors are rarely encountered as they seldom produce symptoms that may lead to their detection.[6,13,14,17]

The tumors are composed of either purely rounded epitheloid cells (predominantly) or short fusiform cells in a fine fibrillary collagenous background. Rarely, mixed pattern is also encountered. Similar to GISTs, EGISTs also display varying amounts of stromal hyalinization, myxoid change and cyst formation. However skeinoid fibers, a common marker in GIST of the small bowel, are absent in this tumor.[6]

Immunohistochemical analysis of EGISTs reveals expression of CD117 (c-kit receptor) (100%), CD34 (50%), neuron-specific enolase (44%), smooth muscle actin (26%) and S-100 protein (4%).[6] On the basis of c-kit (tyrosine kinase growth factor) receptor expression, similar to GISTs; as the origin of EGISTs, the gastrointestinal pacemaker cells of Cajal, located between the myenteric plexus and smooth muscle cells of the gastrointestinal wall, were also proposed. The recent observation that interstitial cells of Cajal do not express CD34, whereas the fibroblasts of Auerbach's plexus do, has complicated the issue of the source of origin of these tumors.[6]

Patients usually present with an enlarging abdominal mass often accompanied by vague abdominal pain of variable duration. Rarely, they are also detected incidentally during a workup for unrelated conditions.[6]

Abdominal ultrasonography is the first-line screening imaging study to detect the presence of a mass and to differentiate cystic from solid tumors. However it usually cannot identify the primary site of the tumor and its characteristics. CT scan can identify the primary tumor site and any displacement or compression of adjacent organs. On contrast-enhanced CT, EGISTs are usually well-defined, huge masses that contain large areas of low-attenuation necrosis and hemorrhage and lack central gas. MRI is also another modality that can aid in differentiating cystic from solid tumors.[12, 14]

Imaging with 18F- fluorodeoxyglucose (18-FDG) positron emission tomography (FDG-PET) can complement contrast enhanced CT in helping to differentiate benign tissue from malignant tissue and necrotic scar from active tumor. Baseline PET scan is recommended prior to initiation of chemotherapy in EGISTs to monitor the response.[12]Preoperative angiography is also an adjuvant diagnostic tool to determine the feeding artery as well as the vascularity of the tumor.[15] Fine needle aspiration cytology has also been reported as a useful diagnostic tool.[16]

Complete surgical excision is the treatment of choice for the localized disease. Imatinib mesylate, a synthetic tyrosine kinase inhibitor is now considered as a useful adjuvant agent.[12]

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