

Case Report**An Unpropitious Surprise Post Vaginal Hysterectomy – A Case of Incidentally Discovered Extra-Gastrointestinal Stromal Tumour***Jessica Minal¹, Nikhil P. V¹, Bhargavi Kalburgi Nagabhushan², Nirupama Murali¹, Nidha Gaffoor¹***Abstract**

Extra-gastrointestinal stromal tumours (EGIST) are rare mesenchymal tumours that arise outside the gastrointestinal tract and mainly affect the omentum, mesentery, and retroperitoneum. We report a case of EGIST in an elderly female patient at an unusual location. The patient was referred to our centre with hemoperitoneum post vaginal hysterectomy for the treatment of third-degree utero-vaginal prolapse. Ultrasonography (USG) of abdomen and pelvis was done which revealed a heterogenous tumour in the upper abdomen. The tumour mass measured 23x18x8.5cms and was abutting the greater curvature of stomach without infiltration. A diagnosis of extra-gastrointestinal stromal tumour, spindle cell type, high grade, with pseudo signet ring appearance was rendered based on the morphology and immunohistochemistry with CD117(c-kit) and DOG1. Rupture of cancerous mass in the abdomen is one of the causes to be kept in mind in hemoperitoneum and EGIST should be considered in the differential diagnosis of large, multi-nodular, primary lesions of the abdomen. A high degree of suspicion is needed as these patients present late and are often diagnosed incidentally during investigations for unrelated medical conditions. 10% of EGISTs are primarily disseminated and the site of origin cannot be established with certainty, emphasizing the enigma of its origin as encountered in the present case.

Keywords: Abdominal neoplasms, CD117 antigen, Immunohistochemistry, Pathology, DOG1, Gynaecology

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Introduction

The most common primary mesenchymal tumour in the gastrointestinal tract (GIT) is the gastrointestinal stromal tumour (GIST) with an incidence of 11-15 cases per 100,000. These tumours arise from the interstitial cells of Cajal, which regulate the GIT peristalsis and demonstrate mutations in the platelet derived growth factor receptor (PDGFR- α) and c-KIT genes which lead to tyrosine kinase activation.^{1,2} Of much rarer occurrence, are extra-gastrointestinal stromal

tumours (EGIST), accounting for 10% of all GISTs, which originate from outside the GIT.³ These tumours are characterized by differentiation towards the interstitial cells of Cajal and have pathological, immunohistochemical and molecular biological characteristics like GIST.^{4,5} The diagnosis of EGIST is often unexpected.

Here, we report a case of EGIST in an elderly female patient, which was discovered incidentally after she underwent vaginal hysterectomy for third degree utero-vaginal prolapse and presented

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to our centre with hemoperitoneum post-surgery.

Case Summary

A 70-year-old female underwent vaginal hysterectomy at another hospital in a rural area, for the treatment of third-degree utero-vaginal prolapse. Soon after opening the pouch of Douglas, there was a gush of altered blood with subsequent blood loss of approximately 1.5 litres. The patient was referred to our centre immediately post vaginal hysterectomy for the management of hemoperitoneum. The previous scan reports were unavailable. Ultrasonography (USG) of abdomen and pelvis was done which showed a heterogenous tumour in the upper abdomen, adherent to the outer aspect of greater curvature of stomach, extending till the splenic hilum. The mass was not adherent to the bladder, bowel loops, liver, or spleen (Figure 1).



Figure 1: USG abdomen: A heterogenous tumour in upper abdomen measuring 23cms in maximum diameter.

The patient was taken up for emergency laparotomy. Gross hemoperitoneum was noted and a large mass was seen in the upper abdomen involving the greater curvature of stomach and extending till the splenic hilum. The mass was surgically resected and a sleeve gastrectomy with splenectomy was performed. The specimens were sent for histopathological examination. The tumour mass received in the histopathology section measured 23x18x8.5cms and was grey brown, irregular and nodular. Cut section showed grey white friable areas with haemorrhage (Figure 2).

The spleen measured 8 cm × 5.5 cm × 3.5cm and no gross tumour infiltration was seen. The fragment of gastric sleeve tissue measured 2x1.5cms and was unremarkable. The microscopy of the tumour mass showed spindled tumour cells arranged in short fascicles, nests, cords, and sheets. Individual cells were mild to moderately pleomorphic, with scant



Figure 2: Gross picture of the large, irregular, nodular tumour mass.

to moderate wispy eosinophilic cytoplasm (Figure 3A). Paranuclear vacuoles and foci of tumour cell palisades were seen. Occasional cells with pseudo-signet ring appearance were also noted (Figure 3B). The tumour showed extensive myxoid areas with thin walled ectatic blood vessels and areas of haemorrhage with mitotic count of 8-10 mitosis per 5mm². There was absence of necrosis. Spleen and stomach did not show tumour infiltration. Differential diagnosis of soft tissue sarcoma was ruled out as the tumour cells were negative for Masson- trichome stain. Immunohistochemistry showed positivity of tumour cells for CD117 and DOG1 and negative for CK5/6, smooth muscle actin (SMA) (Figure 3C and 3D).

A diagnosis of extra-gastrointestinal stromal tumour, spindle cell type, high grade, with pathologic stage of pT4 was rendered as per American Joint Committee on Cancer (AJCC), 8th edition.

Discussion

Gastrointestinal stromal tumour (GIST), was a hardly recognised entity until 2000, has now been better understood with discovery of an activating mutation of the c-kit tyrosine kinase and the ability to target those mutations with the specific tyrosine

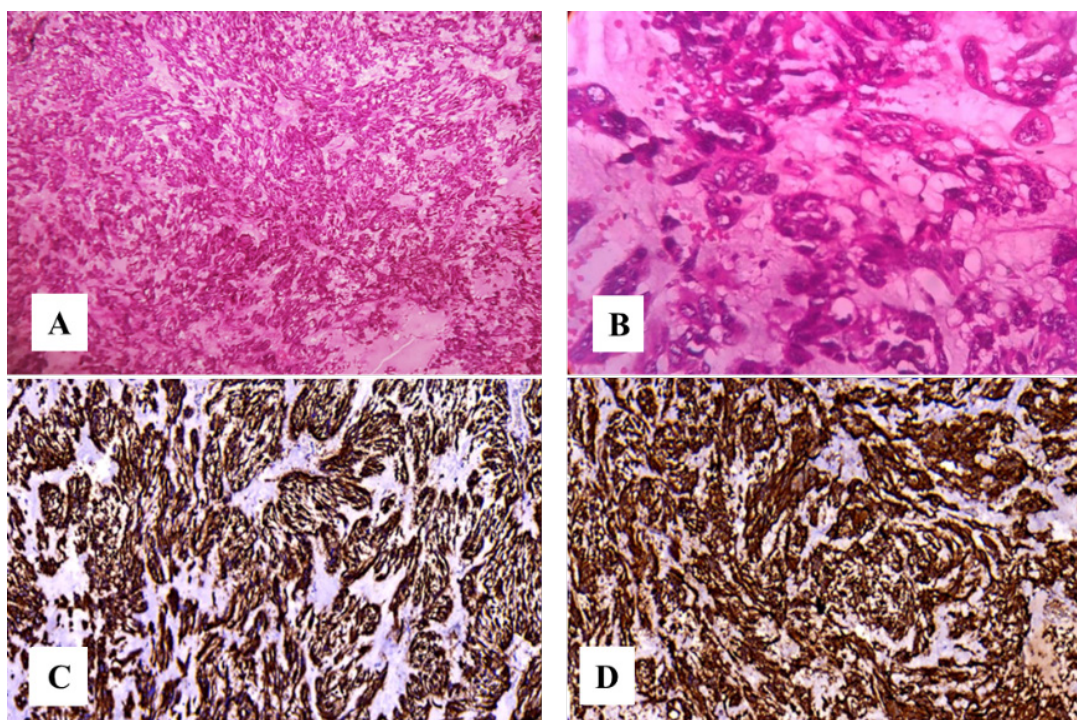


Figure 3: (A) Tumour cells arranged in predominantly short fascicles against a myxoid background (H&E, $\times 100$) (B) Pseudo-signet ring appearance of tumour cells (H&E, $\times 400$) (C) CD117 immunohistochemistry showed diffuse cytoplasmic positivity (IHC, $\times 400$) (D) DOG1 immunohistochemistry showed diffuse cytoplasmic and membranous positivity (IHC, $\times 400$).

kinase inhibitor (imatinib mesylate).²

EGISTs arise outside the GIT and mainly affect the omentum, mesentery, and retroperitoneum. Other uncommon sites include pancreas, gall bladder, pleura, paravaginal and periprostic tissue.⁴ It occurs in the elderly, most commonly in the 7th decade of life, as seen in the present case as well.⁶ In the present case, the tumour was abutting the greater curvature of the stomach. However, there was no infiltration into stomach on microscopic examination. The mass was not adherent to the bladder, bowel loops, liver, or spleen.

There are many hypotheses proposed regarding the origin of EGISTs:

- (1) these usually originate from mesenchymal undifferentiated stem cells
- (2) these arise from extra-intestinal undifferentiated mesenchymal cells capable of differentiating to interstitial cells of Cajal and
- (3) these lose the extramural contact with the intestinal wall in GIT.²

Many authors believe that mesenteric and omental GISTs originate from a primary intestinal or

gastric tumour and for unknown reasons, may become detached from the gastrointestinal wall during their development.⁷ It was difficult for us to determine whether the tumour had originated from the omentum or was it a detached mass from the GIT. Tumour haemorrhages may be the first presentation of an underlying mass.

10% of EGISTs are primarily disseminated and the site of origin cannot be established with certainty, emphasizing the enigma of its origin encountered in our case.⁴ A retrospective study by Yi et al stated the most common primary site was in the mesentery (n=15) followed by the retroperitoneum (n=13) and omentum (n=8).⁸ EGIST is an important differential diagnosis of masses in the abdomen such as leiomyoma and leiomyosarcoma, which show histologic similarities with GIST.⁶ They have no classical radiologic features, and has a wide range of differential diagnoses including leiomyosarcoma, liposarcoma, fibrosarcoma, paraganglioma, solitary fibrous tumour, schwannoma and lymphoma.² The presence of a bleeding mass of uncertain nature may result in a challenging situation for the surgeon, who is forced to perform a resection without knowing the

exact nature of the tumour and hence the extent of resection required.⁹

The histopathologic variants include epithelioid, spindled, vacuolated, nested and myxoid. In this case, we encountered the pseudo-signet ring appearance of tumour cells, which is rare and can pose a diagnostic challenge. Necrosis and brisk mitotic activity are adverse prognostic factors.⁴

EGISTs have a less favourable prognosis as compared to GISTs. Due to its extra- gastrointestinal location there is a delay in presentation of symptoms and subsequent impediment in its diagnosis.⁷ In contrast, early symptoms of intestinal obstruction in GISTs aid in its prompt diagnosis and treatment. Given the rarity of EGISTs, their management follows the guidelines of GISTs. Surgical resection with negative margins remains the standard treatment to be followed by targeted therapy by Imatinib mesylate if indicated.^{7,10,11}

Conclusion

Hemoperitoneum is rare as first presentation of EGIST and imaging plays a role in the diagnosis of hemoperitoneum, in elucidating a cause and detecting active haemorrhage. This case report attempts to create awareness to this unusual presentation of EGIST and stresses that rupture of a cancerous mass in abdomen should be considered as differentials in cases presenting with

hemoperitoneum. EGISTs are rare mesenchymal tumours and should be considered in the differential diagnosis of patients with large, multi-nodular, primary lesions of the abdomen. These patients present later in the course of disease with vague symptoms like tiredness and abdominal fullness and are often diagnosed incidentally during investigations for unrelated medical conditions. The case report also highlights the underreported pseudo signet ring appearance of tumour cells as it poses a diagnostic challenge. Complete surgical resection is the only effective modality despite significant advances in new chemotherapeutic drugs.

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