CASE REPORTS

A rare case of an extra-intestinal gastrointestinal stromal tumour of the sigmoid mesocolon presenting as haemoperitoneum

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Introduction

Gastrointestinal stromal tumours (GIST) are rare and account for 0.1% - 3.0% of all gastrointestinal neoplasms [1]. However, extra-intestinal GIST constitutes up to 10% of cases [2]. Majority arise from the soft tissue of the abdominal cavity whilst the remainder arises from the retroperitoneum. Sites for extra-intestinal GIST includes the mesentery, omentum, peritoneum, gallbladder and liver [3-5]. Although stromal tumours in the gastrointestinal tract commonly metastasize to the omentum and mesentery, they may also occur as primary tumours outside the gastrointestinal tract, especially in the omentum and the mesentery [3]. Very rarely these tumours may present as a haemoperitoneum in an emergency setting. We report an unusual case of spontaneous rupture of a sigmoid mesocolonic GIST presenting as a haemoperitoneum in a 17 year old boy.

Case Report

A 17-year-old boy presented with acute onset abdominal pain and a drop in haemoglobin levels. Physical examination revealed tachycardia with a pulse rate of 124 beats per minute, blood pressure of 100/70 mmHg. His abdomen was mildly distended with mild tenderness and rebound tenderness in both iliac fossae. Peristaltic sounds were diminished. His blood count demonstrated a haemoglobin of 7.4 g/dl, haematocrit 31% and white blood count 18,000 cells/ml. CT scan revealed a 5 x 5 cm mass in the sigmoid mesocolon along with moderate haemoperitoneum [Figure 1 and 2]. Along with this, the patient had multiple congenital cardiac anomalies to the extent of being advised to

undergo a cardiac transplant.

Figure 1. Sagittal section of CT abdomen showing heterogeneously enhancing mass involving sigmoid mesocolon

Figure 2. Axial section of CT abdomen showing heterogeneously enhancing mass involving sigmoid mesocolon

Pre-operative transfusions were given and the patient was taken to the operating room for an emergency exploratory laparotomy. Intra-operatively about 500ml

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of blood with clots was removed and there was a solid tumour of 5 x 5 cm involving the sigmoid mesocolon. Excision of the tumour with a 2 cm margin was done, without any bowel compromise. Biopsy confirmed that the mass was a GIST. The tumour cells were immunoreactive for the KIT protein (CD117), CD34 and Discovered on GIST-1 (DOG-1). The patient was recovering well post-operatively for the first 24 hours and there was no further drop in haemoglobin. Drain output was non haemorrhagic.

Figure 3. Arrow showing mass involving sigmoid mesocolon

Unfortunately, the patient died on day two post-operatively due to sudden cardiac arrest because of his underlying cardiac disease.

Discussion

Gastro-intestinal stromal tumour (GIST) appears to arise from the interstitial cell of Cajal, but the exact cell of origin and precise steps in tumour genesis are not well established [2]. GISTs are commonly found in the stomach, followed by the small intestine, colon, rectum and oesophagus [6]. Extra-intestinal GIST express

CD117 (c-kit receptor)(100%), CD34 (50%), neuron specific enolase (44%), smooth muscle actin (26%), desmin (4%), and S-100 protein (4%). In our case tumour cells were immunoreactive for the KIT protein (CD117), CD34, Discovered on GIST-1 (DOG-1), and the mitotic rate was 10/50HPF. In relation to GIST, the symptoms of obstruction, bleeding, or rupture are less common. Our patient presented with haemoperitoneum, but the exact incidence of spontaneous rupture of a mesocolonic GIST has not yet been quoted conclusively.

References

1. DeMatteo R P, Lewis J J, Leung D. Two hundred gastrointestinal stromal tumors: recurrence patterns and prognostic factors for survival. *Ann Surg* 2000;231: 51-8.
2. Dematteo R P, Lewis J J, Leung D, Mudan S S, Woodruff J M, Brennan M F. Two hundred gastrointestinal tumors: recurrence patterns and prognostic factors for survival. *Ann Surg* 2000; 231: 51-8.
3. Miettinen M, Monihan J M, Sarlomo-Rikala M, Kovatich A J, Carr N J, Emory T S, Sobin L H. Gastrointestinal stromal tumors/smooth muscle tumors (GISTs) primary in the omentum and mesentery: clinicopathologic and immunohistochemical study of 26 cases. *Am J Surg Pathol* 1999; 23(9):1109-18.
4. Mendoza-Marin M, Hoang M P, Albores-Saavedra J. Malignant stromal tumor of the gallbladder with interstitial cells of Cajal phenotype. *Arch Pathol Lab Med* 2002; 126: 481-483
5. Hu X, Forster J, Damjanov I. Primary malignant gastrointestinal stromal tumor of the liver. *Arch Pathol Lab Med*. 2003; 127(12): 1606-8.
6. Schubert M L, Moghimi R. Gastrointestinal stromal tumor (GIST). *Curr Treat Option Gastroenterol* 2006; 9(2): 181-8.
7. Reith J D, Goldblum J R, Lyles R H, Weiss S W. Extra-gastrointestinal (soft tissue) stromal tumors: an analysis of 48 cases with emphasis on histologic predictors of outcome. *Mod Pathol* 2000; 13(5): 577-85.

Key Points:

- Extra gastrointestinal stromal tumours are neoplasms located outside the gastrointestinal tract in sites such as the omentum, mesentery and retroperitoneum.
- GISTs of the mesocolon are rare with only a few cases noted in the literature.
- It is a rare clinical entity that infrequently presents with spontaneous haemoperitoneum and emergency treatment should be sort for resolution of the haemoperitoneum.