CASE REPORT

RARE OCCURRENCE OF MIX STROMAL TUMOURS: A CASE REPORT OF GASTROINTESTINAL AND EXTRA-INTESTINAL STROMAL TUMOUR ON THE JEJUNUM AND OMENTUM OF A NINE YEAR OLD GIRL FROM ADAMAWA STATE, NIGERIA

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ABSTRACT

BACKGROUND: Stromal tumors of the jejunum (GIST) and omentum (EGIST) are uncommon and dearth of information still exists on their occurrence concurrently.

CASE DETAILS: Here, we report a nine year old girl that presented with tender abdominal mass measuring 14 x 8 cm associated with features of gastric outlet obstruction and hyponatremia of 115 mmol/L. A diagnosis of hyponatremia in a child with gastric outlet obstruction secondary to intra-abdominal mass was made for which exploratory laparotomy was carried out. Histology of the tumour revealed stromal spindle epithelioid as well as myxoid cells. Complete resection of the tumour and correction for hyponatremia was done in addition to antibiotics therapy with remarkable improvement.

CONCLUSION: Index case is that of mixed stromal tumours that presented with features of gastric outlet obstruction, and the patient did well after surgery.

KEYWORDS: Gastrointestinal/extraintestinal stromal tumors, Jejunum, Omentum, Paediatrics, Adamawa state, Nigeria.

INTRODUCTION

Gastrointestinal stromal tumor (GIST) is a rare form of soft tissue sarcoma of the gastrointestinal tract. This has led to GIST being classified under smooth muscles neoplasm of the gastrointestinal tract by earlier workers (1). Walker and Dvorak (2), in 1986, however, revealed that smooth muscle differentiation is usually absent in a typical case of GIST. Mazur and Clark (3), in 1983 have identified that leiomyomas that lacked smooth muscle cells and immunohistochemical features of Schwann cells were also classified as GIST. More recently, Hirota and colleagues (4), found that GIST arises from interstitial cells of Cajal on the submucosal and myenteric plexus of the gastrointestinal tract. Many workers have used the term GIST to signify mesenchymal tumors arising from the submucosa of the gastrointestinal tract thereby indicating a shift from smooth muscles neoplasm nomenclature to a more appropriate term called GIST (1-5).

Gastrointestinal stromal tumor occurs in about 2/100,000 population (5). Of this 60% occurs in the stomach, 30% in the ileum, 5% in the duodenum, rarely from the colorectal, esophagus and appendix, and extremely rare from the jejunum (1, 5, 6). Reith et al in 2000 reported another rare GIST that originates outside the gastrointestinal tract; this was termed extraintestinal GIST (EGIST) (7). Stromal tumors in general could be seen as a wide spectrum of tumors with origin from the intestine or extra-intestinal or both as was the index case.

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Here, we report a rare mix of GIST/EGIST arising from the duodenum and omentum of nine nine years old girl at the Department of Paediatrics, Federal Medical Centre Yola, Adamawa state Nigeria.

**CASE Details**

A 9 year old girl presented with a huge painful abdominal swelling associated with projectile vomiting. A firm tender mass measuring 14 x 8 cm was palpated extending from the left hypochondriac, periumblical to the right hypogastrium. Abdominal ultrasonography showed hypochoeic mass occupying almost the entire quadrants of the abdomen. Abdominal ultrasonography showed hypochoeic mass occupying almost the entire quadrants of the abdomen. Abdominal X-ray (Fig 1) showed a mass with features of gastric outlet obstruction (massive gastric shadow ascending up to the level of the thoracic cavity obscuring the left cardiac border and meniscal sign, however, no areas of calcification were observed. There was hyponatremia of 115mmol/L, other laboratory profiles and haemogram were normal. Lack of facility hindered us from doing computed tomography (CT) of the abdomen. A diagnosis of hyponatremia in a child with gastric outlet obstruction secondary to intra-abdominal mass was entertained. At laparotomy a tumor (Fig 2a) was found on the jejunum and the omentum extending to the level of the bladder with adhesions, however, no evidence of metastasis in the abdominal cavity. Histology of the resected tumour specimen subsequently revealed proliferating stromal spindle, epithelioid and myxoid cells in line with the diagnosis of GIST (Fig 2b). Patient had complete resection of the tumor, correction for hyponatremia in addition to antibiotics with remarkable improvement. She was subsequently discharged on follow up visits.

**Fig 1:** Abdominal X-ray (yellow arrows shows huge gastric shadow with diaphragmatic elevation distorting the left cardiac silhouette. Black arrow indicates meniscal sign, while green arrow point to left hypochondriac part of the mass
DISCUSSION

Stromal tumors whether GIST or EGIST originates primarily from mesenchymal interstitial cells of Cajal (1, 5, 6). Our patient presented with two very rare types of stromal tumors that were found on the jejunum and the omentum. Literature search on GIST/EGIST occurring simultaneously on a patient such as ours revealed dearth of information, however, many authors have found GIST or EGIST to occur as an entity. Alexander et al (1), in 2012 in Germany and Dhull et al (5), in 2011 in India have identified EGIST and GIST independently occurring on patients. Patients with this condition may be asymptomatic, which was not the case in our patient (1-7). Patients can also present with painful abdominal mass, mechanical obstruction and bleeding (5). These features were evident in our present case except bleeding. Histology and Immunocytochemical studies are important among other investigative measures used in making the diagnosis of GIST, however, facility for the latter is lacking in our centre. Other investigative findings may not be specific for GIST. In fact, diagnosis of GIST on CT imaging becomes very difficult if the mass is small and lacks ulceration and necrosis (5). Bleeding was not manifest in our patient, she was not pale and her haemogram was normal, these exclude the possibility of ulceration and necrosis.

From the explanation proffered above, CT imaging may have been fruitless even if facility for it is available in our setting. In such cases histopathology is often required for confirming the diagnosis as was done in this case report. Histology in index case showed predominantly spindle cell tumors, which agreed to findings by other investigators (6). Other histologic types such as epithelioid and myxoid variants were also observed in recent case as documented in other studies (2-4, 7). Of note is that histology unlike in other tumours does not determine outcome of GIST (6). Miettinen and colleague (8), in 2006 have argued that tumor size, which indirectly reflects mitotic activity, gives better prognostic features. Another study further stated that tumor size above 10 cm has a poor outcome due to possible metastasis that is observed with huge tumors (9). On this basis, our patient may be having un-favorable prognostic feature because her tumor size was beyond 10cm. However, no visible features of metastases were observed during surgery. Mutation involving tyrosine kinase proteins leading to gain of function remains vital in the pathogenesis of GIST (1, 5, 6, 8). Thus, tyrosine kinase inhibitor Imatinib has been successfully used in the treatment of
metastatic GIST and EGIST in addition to preventing relapse for many years (8).

Imatinib can kill GIST cells that have metastasized to other parts of the body and cannot be surgically removed. Because cells of GIST depend on activation of growth pathways for their proliferation, withdrawal of this pathway may lead to their death through a mechanism called apoptosis (8, 9). Treatment of GIST with Imatinib has effectively resulted in the withdrawal of growth factor pathway leading to apoptosis. Theoretically, our patient could benefit from this drug because she had omental stromal tumour (EGIST) coupled with the fact that microscopic metastasis may have set in due to her massive tumour size. Practically, Imatinib is not readily available in our hospital and Nigeria at large. Reasons for this may be due to its rare indication in terms of usage, as such, it does not form part of our essential drugs list. Moreover, omental stromal tumours are rare making it difficult to study effectively their malignant tendencies and response to Imatinib (5). Despite this setback, most patients may not require the drug because cure can be achieved by surgery even though relapse after surgery has been published (1, 5, 6).

We presented a case of mixed stromal tumours involving the jejunum and omentum associated with features of gastric outlet obstruction, which responded to surgical treatment. Because omental EGIST are known for their relapse, drug treatment in addition to surgery is advised, but due to lack of this drug in our center, our patient did not benefit from it. However, we are recommending Imatinib therapy as an adjuvant to surgical intervention in order to prevent relapse and to prolong long-term survival of patients.

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