STROMAL (MESENCHYMAL) TUMORS OF THE GASTROINTESTINAL TRACT: A CASE SERIES

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ABSTRACT

This is a case series of four patients, two males and two females between 40-60 years of age. These patients presented to our unit with variable symptoms during the past 3 years. Postoperative histopathology reports suggested these masses to be mesenchymal tumors of gut origin.

Key Words: *Stromal Tumors, Plueripotential Cells, Sarcoma, Haematochezia, Haematemesis.*

INTRODUCTION

Gastrointestinal stromal tumors are relatively rare neoplasms among the gut origin tumors. They are the primary non-epithelial neoplasms of the gastrointestinal tract derived from plueripotential mesenchymal cells. These are capable of either partial or terminal differentiation along a variety of cell lines.[1,2] Stromal tumors are the most common form of sarcoma of gastrointestinal (GI) tract.[3,4] Their common presentation is bleeding due to their vascular nature, as an abdominal mass or intestinal obstruction.

CASE SERIES

In October 2002, we received the first patient from a medical unit. A 43 years old male presented with history of recurrent haematochezia and severe anaemia with no other complaints. On clinical examination, he was severely anaemic and had a non-tender mass in the hypogastric region. Investigations showed a hemoglobin of 5 gm%, normal coagulation profile, normal blood picture except for anemia, normal upper and lower gastrointestinal endoscopy and normal barium studies. Ultrasound showed a mass of 8 x 10cm adherent to the pelvic wall and urinary bladder. C.T scan and M.R.I scan suggested a mass of 8x10cm possibly hemangioma or carcinoma arising from the pelvic wall. At laparotomy, we found a highly vascular, fleshy looking, pedunculated mass arising from antimesenteric border of the jejunum, loosely adherent to urinary bladder and pelvic wall. The tumor along with part of jejunum excised and reanastomosis was done. Patient had a smooth recovery. Histopathology revealed a stromal (mesenchyma) tumor, benign looking with 5 mitosis/50HPF,however follow up was suggested.

Second case came in March 2004. This time a 35 years old lady presented with a big mass in left lumbar region with no history of abdominal pain, urinary or bowel symptoms of any kind. Clinical examination revealed a big, non-tender, slightly mobile mass in left lumbar region. Investigations showed hemoglobin of 10 gm% with normal blood picture. Ultrasound suggested a mass of 10x12 cm in left flank, pushing the kidney up and displacing the bowel loops, probably a sarcoma. Barium enema showed an otherwise normal looking colon being displaced by the mass. Abdominal CT scan showed a huge soft tissue mass arising from the posterior abdominal wall and pushing the surrounding structures without invasion. Resection of the tumor along with part of jejunum was done. Patient recovered uneventfully. Histopathology report was a borderline stromal (mesenchymal) tumor.

Third case was a 50 years old female, shifted from medical unit in October 2004. She had a history of bilious vomiting, weight loss and a palpable mass in the right mid-abdomen. Investigations showed hemoglobin of 11gm% with normal blood picture. Ultrasound suggested a solid mass of 8 x 6 cm in the umbilical region, possibly a retroperitoneal sarcoma. Upper G.I. endoscopy showed dilated stomach and duodenum. Biopsy from the third part of duodenum revealed normal gut mucosa. Abdominal C.T scan showed a solid mass of 8x6.5 cm in right mid abdomen, with markedly dilated duodenum and stomach. Laparotomy was performed. Peroperative findings were a highly vascular solid mass arising from the proximal

jejunum, causing dilatation and obstruction of the duodenum and stomach. Resection of the tumor along with part of jejunum was done. Patient had a smooth recovery. Histopathological report was that of a borderline stromal tumor of the gut.

Fourth case came in March 2005. The patient was 58 years old male, with a history of haematemesis for two weeks. On examination, he was very pale looking. He had hemoglobin of 5 gm% with normal coagulation profile. Ultrasound showed a solid mass of 4 x 5 cm in the posterior wall of the stomach. Endoscopy revealed a lesion in the posterior wall of the stomach. Biopsy was taken which reported normal gastric mucosa. The patient was transfused 6 pints of blood and put on the list for surgery. Peroperatively a solid nodule of about 4 x 3 cm in the posterior wall of the body of the stomach was found. Partial gastrectomy including the lesion was performed. Patient recovered smoothly. Histopathology report was a gastrointestinal stromal tumor with mitosis rate of less than 4 mitosis per 50 HPF, a benign tumor. However, follow-up was suggested.

DISCUSSION

Gastrointestinal stromal tumors are the primary non-epithelial neoplasms of the gastrointestinal tract. They originate from the plueripotential mesenchymal cells (interstitial or stromal cells) of the gastrointestinal tract. These cells have the potential to differentiate along a variety of cell lines[1]. Stromal tumor is the new name for Leiomyoma and Leiomyosarcoma due to their unpredictable behavior.

These tumors generally arise in the upper gastrointestinal tract approximately 60% in the stomach, 30% in small intestine and 10% in other parts of gastrointestinal tract.[5] These are life-threatening tumors because of their highly vascular nature. They are considered the most common malignant form of sarcoma of gastrointestinal tract but overall they are still relatively rare.[3,4] The incidence of this tumor is highest in people aged 30-60 years; with male to female ratio of 2:1.

Although these tumors have an uncertain behavior but some predictions can be made according to Amin's classification. STUMP (Stromal Tumors of Uncertain Malignant Potential) is the term used by pathologists to describe a neoplasm when they are unable to distinguish a benign from a malignant tumor.[1]

Three of the four cases had tumor size of more than 5 cm and showed <5 mitosis*I*50 HPF on histopathology, and thus were placed in borderline category. Tumor size, in the fourth case was 5 cm and had less than 5 mitosis/50 HPF on histopathology and hence was put in low risk category. The behavior of these tumors can be confirmed by immunohistochemical staining for actin, desmin, vimontin, chromogranin A, C kit etc.[6-9] The clinical presentation of patients with stromal tumors is very variable. Common presenting features are:

PREDICTION OF TUMOUR DIFFERENTIATION ACCORDING TO AMIN'S CLASSIFICATION

	Size	No. of Mitosis
Benign	< 5cm	< 5 / 50HPF
Borderline	> 5cm	< 5 / 50H PF
Malignant	Any	> 5 / 50HPF

Table 1

Vague non-specific abdominal pain as occurred in two of our patients; both females.

GI tract bleeding, hematemesis and malena: The first case of a 43 years old male presented with repeated massive malena leading to hypovolemic shock on three occasions. The fourth case a 58 years old male with gastric mass had severe hematemesis, causing hypovolemia.

Intestinal obstruction and perforation of gut. The third case of a 50 years old female patient presented with duodenal obstruction.

Clinical examination may reveal:

Pallor due to blood loss.

Palpable mass - three of our patients had big palpable mass.

Investigations commonly required are:

blood complete

Upper G.I endoscopy and biopsy (which is most of the time inconclusive because the tumor is extra mural. Endoscopic biopsy in two of our patients reported normal mucosa).

Abdominal ultrasound for extent and spread of tumor.

Abdominal CT scan for extent of the tumor.

Preoperative confirmation of the nature of the tumor is usually difficult.

Principles of treatment are:

Surgery is the main stay of treatmentit involves resection of the tumor along with regional lymph nodes.

There is no role of extended lymphadenectomy.

There is no role of conventional radio-chemotherapy, except in metastatic disease.

A new hope in the management of these tumors is the recently developed drug (Gilvec), which is said to be effective in stromal tumors of gastrointestinal tract.[3]

Trans-catheter embolization is a useful in patients presenting with massive GI bleeding.[11-14]

CONCLUSION

In patients presenting with massive haematemesis or malena along with ultrasonographic evidence of a solid mass, the possibility of stromal tumor should be kept in mind. Postoperative regular follow-up of these patients is very important because of the uncertain behavior of these tumors. The facility for catheter embolization needs to be developed here as it will help in saving many lives.

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