Case Report

Gastrointestinal Stromal Tumour of Pelvic Wall: Report of a Case

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Abstract

Gastrointestinal stromal tumors (GIST) because of its rarity continues to be a curiosity for radiologists, surgeons and pathologist. These tumors are usually found in gastrointestinal tract (GIT) but account for less than 1% of GIT tumors. A 35-year-old male presented to us with complaints of painful micturation which on radiologic evaluation revealed a large mass in relation to urinary bladder. The tru-cut biopsy reported a low grade soft tissue tumour. The said mass was excised on laparotomy and sent for histopathological examination which reported as high grade GIST or a pleomorphic sarcoma as possibilities. Immuno-histochemistry finally confirmed the diagnosis of a GIST with CD 34 positive and C-Kit negative status. Approximately, 95% of GIST tumors are C-Kit positive and hardly 5% are C-Kit negative. C-Kit negative status makes this case report more worth reporting.

Keywords: Pelvic, GIST, tumor, mesenchymal, sarcoma

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Introduction

Gastrointestinal stromal tumors (GIST) account for less than 1% of gastrointestinal tumors (GIT). GIST are usually seen in the stomach or small bowel. Amongst the mesenchymal tumors GIST is most commonly found in GIT (1). These tumors at times may come out of the confines of GIT and involve omentum, mesentery or retroperitoneum (2). GIST can behave as a benign or as a malignant tumor depending on the size and mitotic activity (3). The diagnosis of these lesions many a times may be quite difficult and it will be possible only after knowing the immune-phenotypic features and histology of the lesion. These tumors are rarely seen outside the GIT.

Case Report

A 35-year-old male without any past medical history presented with painful micturation since 3 months. Physical examination showed firm mass in hypogastrium measuring about 10x8 cm. Ultrasonography showed a well circumscribed hypoechoic mass uniform in echogenicity about 763 x 95.6 x 80 mm with a volume of 324ml seen in relation to bladder and compressing right lateral wall of bladder. CECT abdomen and pelvis showed a heterogeneous mass seen in pelvis measuring 94x81 mm in size compressing urinary bladder. Exact origin of mass not clear on CT (Fig. 1). Impression was a GIST or a neural tumour or any other connective tissue tumour. Tru-cut biopsy of lesion was reported a low grade soft tissue tumour of spindle cell morphology, and could not be categorised further. Patient was taken for surgical exploration and operative findings were a large tumour of size 10x15 cm retro pubic in position adherent to posterior wall of pubic symphysis and infiltrating anterior wall of urinary bladder, tumour was firm to hard in consistency highly vascular, fleshy with areas of cavitations inside. Excision

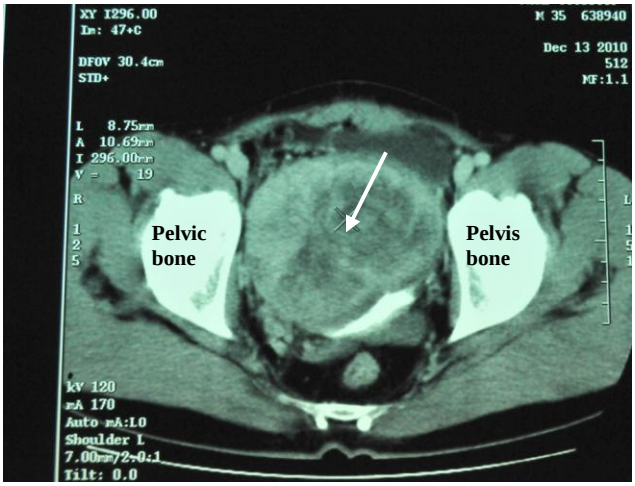

Figure 1: CECT picture showed a heterogeneous mass seen in pelvis measuring 94x81 mm in size compressing urinary bladder (arrow). Exact origin of mass not clear on CT but tumor showing a close proximity with left sided pelvic bone and bladder wall.

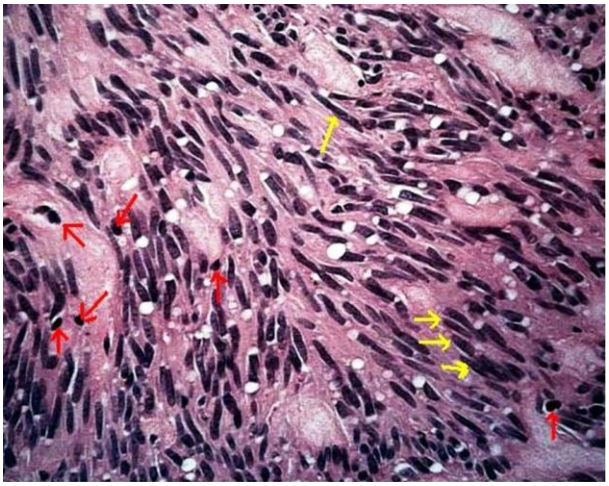

Figure 2: High Grade GIST pleomorphic spindle cells (yellow arrows) and mitotic figures (red arrows) in a neural background. H&E-(10X)

of pelvic mass along with anterior bladder wall with repair of urinary bladder was performed. Histopathology in multiple serial sections showed spindle cell neoplasm. The individual cells showed moderate pleomorphism, there are places of neural differentiation and diagnostic possibilities are GIST-high grade or pleomorphic sarcoma (Fig. 2). Paraffin block was revised which revealed GIST-high grade, and immune histochemistry of tumour was CD-34 positive and C-kit negative. On the basis of radiological, histopathology and immune-histochemistry GIST of pelvic wall was diagnosed. The post-operative period was smooth and patient is still on follow-up for more than 2 years with Department of Medical Oncology and General Surgery and has received imatanib mesylate one tablet daily as adjuvant therapy for two years. Follow-up investigations did not show any evidence of recurrence.

Discussion

GIST may present as a pelvic mass with extra intestinal origin; even though it is a rare occurrence but some cases with diagnostic dilemmas like our case have been reported in literature (4,5,6).

Gastrointestinal stromal tumors may occur anywhere in GI tract including the stomach, small intestine, esophagus, rectum and colon. In rare cases, the gastrointestinal stromal tumor can occur at other locations in abdomen (7). The actual origin of GIST is a pluripotential mesenchymal stem cell. This cell was programmed to differentiate into the interstitial cell of Cajal. These cells are believed to initiate and coordinate GI motility, hence the name GI pacemaker cells was coined by Kinblom and co workers (8). In subsequent years, research showed that Cajal cells are developmentally dependant on stem cell factor and express KIT which is regulated through KIT kinase. It was ultimately the work of Hirota et al. (9) in 1998 by discovering c-kit proto oncogene mutations in GIST and it was established as a separate entity. In present day time GIST is diagnosed on basis of histology, immunohistochemistry and oncogene c-kit mutation (10). Nowadays, based on histology, we can differentiate GIST from pleomorphic and spinde cell tumors. Approximately, 95% GIST tumors express C-Kit (CD117) and 70% express CD 34. In addition 80% express heavy caldesmon and 25% are positive for smooth muscle actin and 5% for desmin (10). Somatic mutation may occur independent of anatomic location of the tumor and this mutation of CD 117 is observed in majority of GIST tumors (11).

Conclusion

This case proves that GIST can have a varied clinical presentation and can even arise from the pelvic wall as a huge mass with a C-kit negative status. It would be always wise to send immunohistochemistry in such doubtful histological variants to establish the diagnosis. Even such huge tumors with unusual presentations if followed properly and put on adjuvant treatment can respond well.

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