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Case Report

A Rare Case of Gastro Intestinal Stromal tumour presenting as Adnexal Mass

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Introduction

Gastrointestinal stromal tumors (GIST) are uncommon nonepithelial mesenchymal tumors originating usually from the gastrointestinal tract (GIT) and rarely within the abdomen[1].

Most commonly they occur in the stomach (60-70%), in the small intestine (25-35%), and in the esophagus, colon and rectum (10% or less)[2].

Tumors with similar features designated as extra-gastrointestinal stromal tumors are rarely seen.

Case report

A 45-year-old, unmarried, nullipara presented with complaints of pain in the abdomen and vomiting since 7 days. Her menstrual cycles were normal. Abdominal examination showed a nontender, mobile, hard mass of 20 weeks size, with a uniform surface. Its lower borders could not be made out. Vaginal examination showed a retroverted normal size uterus and the mass was felt separately from it with fullness in right fornix. Her CA-125 was normal. Ultrasound examination showed a 18x19 cm mass in the right iliac fossa extending to right hypochondrium. It was well defined with homogenous echotexture and a few areas of cystic degeneration of the right ovarian tumor. CT scan however reported a large left benign ovarian tumor with right sided simple ovarian cyst. The patient was taken up for laparotomy on 21/06/05. On laparotomy while the uterus, tubes and ovaries were normal, the right sided 10x8 cm cystic mass was attached to the ovary with a flimsy band and on the left side a solid tumor measuring 15x20 cm was adherent to the omentum and the left ovary. Total abdominal hysterectomy with bilateral salpingo-oophorectomy was done, both the tumors resected and sent for histopathology which revealed gastrointestinal stromal tumors. CD 117 tumor marker was positive thus confirming the diagnosis of gastrointestinal stromal tumor. The patient refused chemotherapy inspite of counseling and was discharged. However she was doing well on follow up.

Discussion

GISTs are mesenchymal tumors arising in the GIT and occasionally within the abdomen without gastrointestinal connection. They show differentiation towards interstitial cells of Cajal which are normally concerned with motility of the gut[2]. These tumors are

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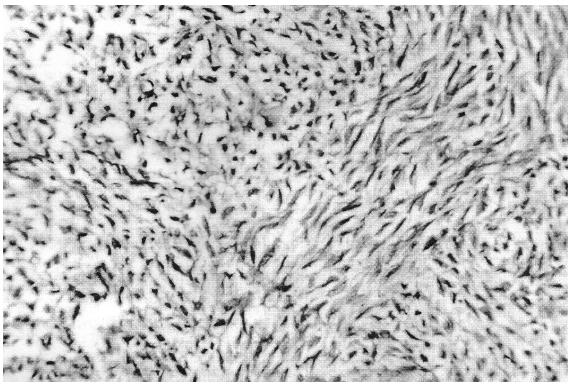

HPR of Gastrointestinal stromal tumour showing epitheloid pattern

rare with an incidence of 5-10% of all sarcomas. They are more common in the age group of 40- 80 years and are seen more commonly in men. In all GIST cases, there is abnormality in a gene called C-kit gene. This gene is found in all cells of the body. It leads to the formation of a protein called KIT and is usually inactive. In most GISTs the KIT is abnormal and active. This KIT (CD 117) acts as enzyme called tyrosine kinase which is important in the diagnosis and treatment of GIST[2]. Thus GIST can be diagnosed histologically and with immunohistochemistry.

Tumors can be submucosal, intramural or subserosal and grossly solid or cystic. Histologically they show a spindle cell or epitheloid pattern[3].

Immunohistochemistry CD34 and CD117 are reliable markers. CD117 is positive in 95% of the GISTs and CD34 in 60-80% of GISTs.

CT and MRI can also aid in diagnosis to help in determining extent and spread of the disease. Prognostic factors for GISTs are tumor size, mitotic count, and extragastrointestinal spread[3]. Accordingly they are graded as very low risk, low risk, intermediate risk, and high risk.

Surgery is the mainstay of the treatment. The 5 year survival rate after complete resection of GIST is approximately 50%[4]. Radiotherapy is not very effective. Previously GISTs were regarded as resistant to chemotherapy. Imatinib mesylate is a synthetic tyrosine kinase inhibitor which is shown to be effective in metastatic and inoperable GIST. Various larger trials have confirmed this finding and it is now considered to be the drug of choice for metastatic and inoperable GIST2.

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