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"Gastrointestinal Stromal Tumour : Management in a rural Medical college in India "

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Abstract: Gastrointestinal stromal tumour (GIST)) is one of the most common mesenchymal tumors of the gastrointestinal tract that accounts for 1-3% of all gastrointestinal malignancies. For many years these tumors have been the subject of controversy and discussion in the literature regarding their histogenesis, diagnostic criteria, prognostic features, and nomenclature. It is now proved to originate from an intestinal pacemaker cell called the interstitial cell of Cajal. GIST may develop anywhere along the gastrointestinal tract, but most often it arises in the stomach and, less commonly in the intestine. It is a tumour that originates from spindle cells arising from muscle layer with mixed differentiation. The management depends on the histology and size of the tumour. With limited resources for advanced investigations this present case having size of 15 cms and high malignant potential was treated with resection with adequate margins.

Keywords :GIST , CD 117 ,mitotic index, jejunum.

Introduction : Stromal tumors of the gastrointestinal tract represent relatively rare lesions that are thought to arise from connective tissue elements located along the entire length of the gut. They constitute the most common mesenchymal neoplasm of the gastrointestinal tract and are highly resistant to conventional chemotherapy and radiotherapy. Not all GISTs ultimately behave as malignant but as it is difficult to predict which ones will do so it is proper to designate these tumours as low risk to high risk of being malignant tumours. Larger the size of tumour more are the chances of it being highly malignant. Studies have also demonstrated the diagnostic role of CD117 expression which has been proposed as the most sensitive and specific phenotypic marker of GISTs. Most (50-80%) GISTs arise because of a mutation in a gene called c-kit. In this case we have used simple criteria in treating the tumour as resources for detailed investigations were not available. It was a large tumour (Photograph 1) arising from jejunal wall with no lymph nodes. Adjuvant therapy has been used , but majority of study series deal with targeted therapy with Imitanib to make inoperable tumors operable however no consensus has been reached to use this technique as adjuvant therapy.

Case history : A 70 year old female presented with a painless lump in umbilical region since one year. This was not associated with vomiting, altered bowel habits ,malena. On examination a lump was palpable in umbilical quadrant which was not painful, variegated consistency with mobility more in horizontal than vertical direction. It was a well delineated mass .The lump was intraabdominal, intraperitoneal. Ultrasonography demonstrated a solid mass probably arising from the bowel with no evidence of bowel obstruction. As the patient could not afford a CT scan it was decided to explore the patient. On exploration the mass was arising from anti-mesenteric border of jejunum . There were no palpable mesenteric lymph nodes, liver was normal and the mass was not adherent to adjacent structures. Resection anastomosis was done. Histology revealed a high grade GIST tumour , showing spindle cells with short storiform pattern (Photograph 2).

There was increased mitotic activity with small areas of necrosis and hemorrhages. The mucosa was free and nearest and farthest margin free of tumour. There were no mesenteric lymph nodes in the specimen. CD 117 was done which was positive. The post operative period was uneventful. Patient was discharged on 10th postoperative day and asked to follow up every two months.

Discussion:

GISTs arise from the bowel layer called muscularis propria. Considerable studies have been done worldwide to investigate these unpredictable tumours. Various institutes have had different opinions regarding management and prognosticating the tumour. The problem is that surgeons do not have a reliable test to determine which tumors will recur and which ones probably won't. Therefore GISTs are labeled not as benign or malignant, but on a risk scale of low risk to high risk. Even small GISTs that looked "benign" have been known to act aggressively later.

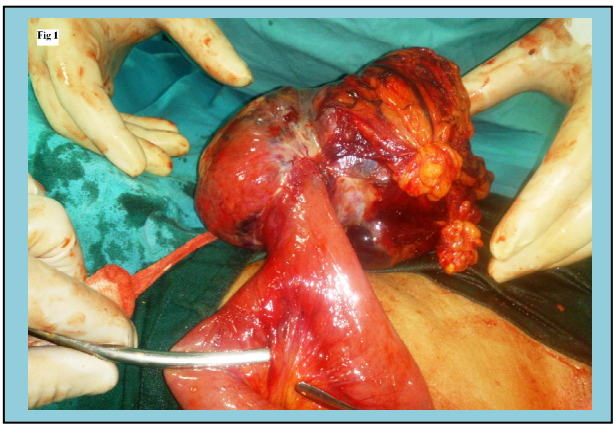
We have relied on two important criteria to treat this case as resources were limited. Mitotic index and size of tumour are two criteria [1] which in our opinion are sufficient to avoid confusion on advanced investigations. This case was 15 cms size and having >5/50 hpf mitotic index thus categorizing this tumour into high risk malignant tumour. Presence of c-KIT proto-oncogene (CD117) represents a histological feature of these tumors [2]. CD 117 was positive.

Apart from consistent positivity for CD117, about 60% to 70% of GISTs show immunopositivity for CD34, 40% show immunopositivity for smooth muscle actin (SMA), and around 5% show immunopositivity for S-100 protein. None of the mentioned antigens are specific for GIST. Desmin positivity in true KIT-positive GIST is very uncommon (1% to2% of cases) and is invariably focal, showing positivity in only few tumor cells [3]. All these data suggests CD 117 as a reliable indicator of malignancy. Various major /minor criteria have been stated by Pascal Bucher et al [4] that elaborately defines the tumour status. Most GISTs fall into a "gray zone," and, hence the ultimate determination of malignancy depends on the development of tumor recurrence or metastasis. Taking that into account we preferred to do a resection anastomosis of the tumour with 10 cms margin.

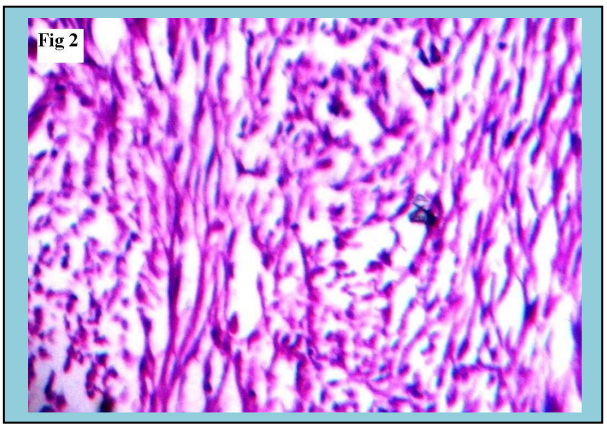
These tumors are known to recur locally within the peritoneal cavity or present with hepatic metastasis which makes the prognosis invariably poor. Chemotherapy in the form of imatinib mesylate has not been recommended as adjuvant therapy in all cases but should certainly be applied to recurrences [5]. In patients with resectable tumors use of Imitanib has the danger of not knowing the duration of treatment and not knowing which patients are likely to be benefitted[6]. Here as there was no evidence of spillage during surgery, absence of lymph nodes, no peritoneal seedlings, only resection was done and chemotherapy will be reserved for recurrence.

Conclusion :GISTs forms a very interesting tumour whose behavior cannot be predicted and judicial use of management criteria and application of proper immunohistochemistry panel minimizes use of advanced laboratory investigations in a rural set up with limited resources and low socioeconomic patient class.

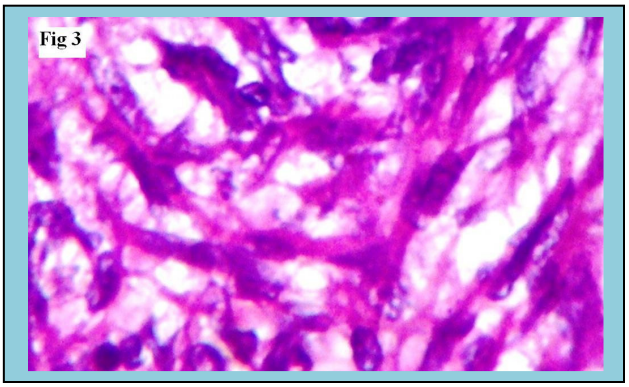
Legends:



Photograph 1 : Tumour arising from jejunal wall.



Photograph 2 : Tumor composed of spindle to epithelioid cells of varying sizes & spindles arranged in storiform pattern.



Photograph 3 : Spindle to epithelioid cells exhibiting pleomorphism of cells & nuclei.

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