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Case Report

GASTROINTESTINAL STROMAL TUMOR OF RECTUM

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Abstract

Although gastrointestinal stromal tumors (GISTs) frequently occur in the gastrointestinal tract, they are relatively rare in the rectum. Biopsy of the lesion and immunohistochemistry (IHC) confirm the diagnosis. Complete surgical resection is the principal curative procedure. In combination with surgery, immunotherapy with Imatinib shows cure in intermediate risk and improvement in high risk rectal GIST. We report a case of a 45-year-old female who presented with constipation and generalized weakness, ultimately diagnosed to have rectal GIST.

Key words: Gastrointestinal stromal tumor, Rectal GIST, Imatinib

Introduction:

Gastrointestinal stromal tumor is a name given to a group of gastrointestinal tumors which were otherwise unclassifiable as being of smooth muscle or neurogenic origin[1]. They are mesenchymal neoplasms expressing KIT(CD117) tyrosine kinase and showing presence of activating mutations in KIT or PDGFRá (platelet-derived growth factor alpha)[2]. It is the commonest gastrointestinal mesenchymal tumor[3] with the commonest site being stomach (50–60%), followed by small intestine (30–40%), colon (7%), and oesophagus (1%)[4]. GISTs of anal canal and rectum are often grouped together and account for nearly 5% of all GISTs[4,5]. We present a case of rectal GIST, treated by anterior resection and adjuvant immunotheraphy with Imatinib.

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Case Report:

A 45-year-old female presented to our hospital with history of constipation and generalized weakness for 3 months. Rectal examination showed a well defined mass located at the posterior aspect of the rectal wall free from mucosa. Routine blood tests were within normal limit. CT scan revealed distension of rectum with one large polypoidal soft tissue attenuation lesion (Figure-1). The mass measured 9.1x8.7x5.2 cm, located posteriorly towards right. Post contrast image revealed heterogenous enhancement and variable areas of central necrosis with few calcified areas. There was no lymphadenopathy on CT scan. Colonoscopy was done and biopsy was taken from the rectal mass. Histopathological examination showed rectal tissue with submucosal tumor location composed of proliferation of densely packed epitheloid and spindle cells, with prominent nuclear palisading (Figure-2).

Mitotic count was of 3 mitosis/50HPF. C-KIT protein (Figure-3) and CD34 (Figure-4) were positive. Final diagnosis was GIST of intermediate risk aggressive behavior (Table-1). Patient underwent ultralow anterior resection, coloanal anastomosis and defunctioning ileostomy. Postoperative course was uneventful. The patient was discharged on postoperative day 14 with Imatinib and Gut continuity was restored by closure of ileostomy after 6 months. Follow up at 6 months with sigmoidoscopy and CT scan showed no tumor mass.

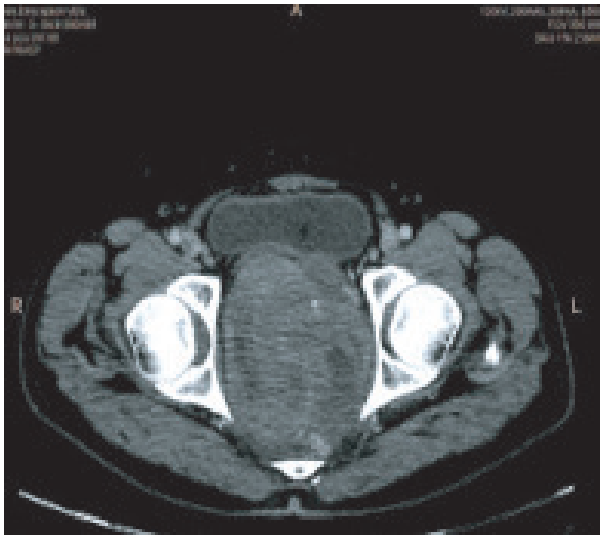

Fig.-1: *CT Abdomen- showing distension of rectum with large polypoidal soft tissue attenuation, heterogenous enhancement and areas of central necrosis with calcification*

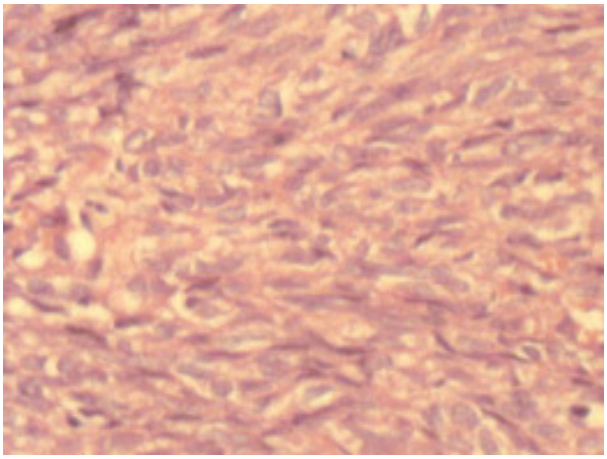

Fig.-2: *Histopathology -Tumour composed of proliferation of densely packed spindle cells, with nuclear palisading. (HE X400)*

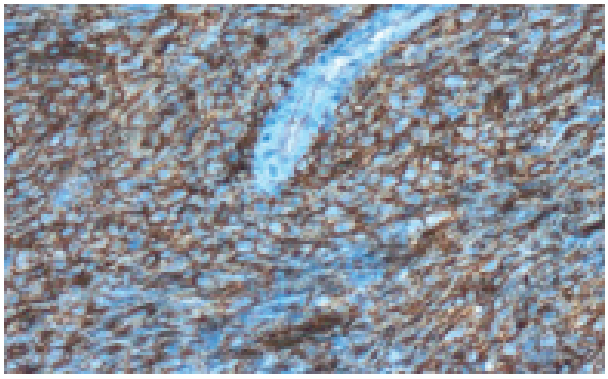

Fig.-3: *CD117 Positivity seen in tumor cells of GIST (C-KIT staining) (×400).*

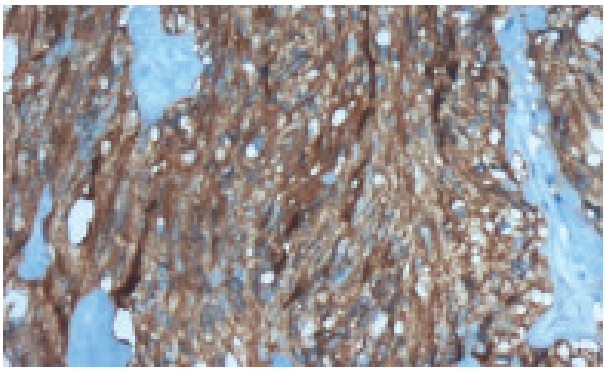

Fig.-4: *CD34 Positivity seen in tumor cells of GIST (IHCx 400)*

Table-I

Defining risk of aggressive behavior in GIST[6]

	Size	Mitotic Count
Very low risk	< 2 cm	< 5/50 HPF
Low risk	2–5 cm	< 5/50 HPF
Intermediate risk	< 5 cm	6–10/50 HPF
	5–10 cm	< 5/50 HPF
High risk	> 5 cm	> 5/50 HPF
	> 10 cm	Any mitotic rate
	Any size	> 10/50 HPF

Discussion:

The term “GIST” was introduced in 1983. It included tumors of the GI tract that could not be classified as either smooth muscle or neurogenic in origin[1]. Among GI mesenchymal tumors GISTs are the most common. GIST is an uncommon mesenchymal tumor and expresses CD117, a tyrosine-kinase growth factor receptor as important marker [7]. CD117 also serves as the target for drug therapy with imatinib, a selective tyrosine-kinase receptor inhibitor that is at present the only promising chemotherapeutic drug for the

treatment of patients with advanced GIST, although complete surgical resection remains the most effective treatment for such a tumor[8]. The symptoms of GIST in the rectum do not generally differ from those of other rectal tumors and diagnostic work-up is also similar. Digital examination of the rectum, colonoscopy and CT abdomen are essential for diagnosis. Preoperative biopsy plays a key role in the diagnosis of GIST, since it provides information on the immunohistochemical features and mitotic count. GIST typically expresses CD117, often CD34 and sometimes SMA and S-100, but its expressions vary depending on different sites. Miettinen et al[9] found that CD34 is expressed in 92% cases of rectal GIST but only 50% cases of small intestinal GIST. On the other hand, Smooth muscle antigen (SMA) is expressed in 47% cases of small intestinal GIST but only 14% cases of rectal GIST. The reason for these variations remains unexplained. Most GISTs originate within the muscularis propria and most commonly have an exophytic growth pattern[6,10] , seen on CT or MRI images. A focal, well-circumscribed mural mass is the most common finding and an infiltrated layer can be clearly assessed[11]. CT or MRI helps define local invasion and detection of metastases.

Because of lower incidence, the clinicopathological profiles of rectal GIST have not yet been accurately characterized and prognostic factors of common sites like stomach is used instead. Size and mitotic rate are common prognostic criteria[12-14]. A rate of 5 mitoses per 50 HPF is taken used as a limit to discriminate between benign and malignant lesion[15]. Tumors of 2 cm in diameter generally behave in a benign fashion. Less than 5 cm in diameter are associated with a better survival. The epithelioid phenotype has poor outcome. Symptoms lasting for a year are considered as further prognostic factors.

Complete surgical resection with negative margins is curative for primary and non-metastatic low risk tumors[6,16-18]. Local excision, anterior resection or abdomino-perineal resection for rectal GIST depends on tumor size and location[15,19]. Imatinib is useful as adjuvant post-operative treatment in high risk tumor or in patients with incomplete surgical resection. Imatinib is accepted treatment for advanced or metastatic tumors but further evidence is needed to see its effectiveness in high risk tumors and to evaluate its role as neoadjuvant therapy.

Conclusion:

Rectal GIST is rare. The diagnostic workup is same as other rectal tumors. Biopsy of rectal GIST is essential for preoperative diagnosis. CD117 and CD34 are important tumor markers. Rectal GIST should be considered as a differential diagnosis for exophytic lesions in rectum.

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