GASTRO-INTESTINAL STROMAL TUMOUR OF RECTUM CASE REPORT

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Abstract:
Gastro-Intestinal Stromal Tumour (GIST) is a mesenchymal tumour arising from the interstitial cells of Cajal. Though it can occur anywhere in the Gastro intestinal tract, it is common in stomach accounting for nearly 90 of cases and the rest accounting for only 10 of cases. This report describes a case of Gastro-Intestinal Stromal Tumour of the rectum.

Keyword: Gastro-intestinal stromal tumour, GIST, Rectum

Case History:
A 40 years old male was admitted in the surgical department with complaint of constipation and abdominal pain for 6 months. On examination patient had a polypoidal growth rectum. Patient underwent abdominoperineal resection and the specimen was sent to department of Pathology for histopathological examination.

Gross:
Specimen of intestine included colorectal junction and anal verge measuring 15cm in length. Specimen was cut opened and showed irregular soft pinkish glistening mass measuring 4 x 3 x 2 cm closer to one resected margin (Anal verge) (Fig. 1) A solitary lymph node was dissected out from serosa measuring 1.5 x 1.5 cm. Cut section was grayish pink and firm.

Background:
The interstitial cells of Cajal is a mesodermally derived cell located in the wall of the GI tract and thought to function as “pacemaker cell” in the transmission of the stimuli leading to the coordinated contraction of smooth muscle. Cajal cells express C-KIT (also known as CD117) and CD34. GIST occurs mostly in stomach around the age group of 60 years, About 10% occur under 40 years of age but uncommon in children. Gastro-Intestinal Stromal Tumour of colorectal region is very rare.
Figure 1: Gross photograph showing a 4x3x2 cm Polypoidal mass arising from the colorectal region.

Microscopic
Sections studied showed tumor composed of spindle cells arranged in fascicles (Fig.2). Individual cells showed pale staining eosinophilic cytoplasm and pleomorphic ovoid and vesicular nuclei (Fig.3). There were areas of neuronal differentiation, with transmural infiltration of rectal wall and extension into anal verge (Fig.4). Section was immunostained for CD117, the stained section showed very strong immuno reactivity for CD117 (Fig.5). Section from lymph node showed reactive follicle and was negative for tumour deposit.

Figure 2: Photomicrographic picture shows fascicle of spindle cells with ovoid vesicular nuclei (H&E X 100x)
Figure 3: Photomicrographic picture shows spindle cells with mitotic figure (H&E X 400x)
Figure 4: Photomicrograph of spindle cells with rectal mucosa (H&EX10x )
Figure 5: Photomicrograph of immuno-reactivity – CD 117 X 100x

Discussion:
GIST appears to arise from (or) share a common stemcell with the interstitial cells of cajal, which are located in the muscularis propria (Pace maker cells for GIT peristalsis). Cajal cell express C-Kit (CD117) and CD34. Carney triad in young females includes Gastric GIST, Paraganglioma and Pulmonary chondroma. GIST is most common along with neurofibromatosis type 1. Phenotypic features of GIST includes 4 categories

1. Tumor differentiation towards smooth muscle. Largest category, Immunohistochemistry positive for smooth muscle, actin, desmin.

2. Calponin. Shows microscopically both spindle cells and epithelioid cells.

3. Tumor differentiation towards neural elements. Second large category shows microscopically spindle cells. Immunohistochemistry positive for neuron specific enolase, Leu7, S100 protein.

4. Tumor dual differentiation towards smooth muscle and neural elements.

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<th>Risk categories for GISTs based on tumor size and mitotic activity</th>
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<td>Tumour showing less than 2 cm size and less than 5 mitosis per 50 HPF are at very low risk, 2 to 5 cm in size and less than 5 mitosis per 50 HPF are low risk, 5 cm in size and less than 5 mitosis per 50 HPF are intermediate risk and more than 5 cm size and more than 5 mitosis per 50 HPF are high risk.</td>
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Colonic gist with low mitotic rates < 5 mitosis per 50 HPF, typically are CKIT positive, have spindle cells with skenoid fibres (Aggregates of extracellular collagen). Presence of skenoid fibres indicates good prognosis. The present case originated in a rare site – rectum. Though the size of tumour at the time of diagnosis was only 4 cm, cellular pleomorphism, mitotic index of 7 per 50 HPF and transmural invasion along with anal verge involvement places the tumour into a high risk malignant category.

Reference:
a common type of gastrointestinal stromal neoplasm. Ultrastruct Pathol 1997, 21; 419-424.
