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Spectrum of Pediatric gastrointestinal tumors with special reference to clinico-pathological findings.

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Abstract:
Background: *Alimentary tract malignancies in children are rare and the incidence is less than 1% of pediatric tumors. Rapid diagnosis is essential for early surgical intervention and adjuvant therapy in case of malignant tumors .*
Aims and Objectives: *This study was undertaken to ascertain the clinical profile, anatomic distribution and histological subtypes of all pediatric gastrointestinal masses.*
Material and methods: *This prospective study was conducted in tertiary care hospital from July 2010 to June 2015. Total 21 patients with clinically and radiologically detected gastrointestinal masses were evaluated. The data of every patients including age, sex and presenting symptoms were recorded. Resected specimens were obtained for histopathological examination and categorization was done according to WHO classification.*
Results: *Among 21 cases, 16 were males (76.2%). The median age was 6 years. Maximum cases of benign lesions were mature teratoma (3 cases; 14.28%), followed by inflammatory myofibroblastic tumor (9.5%; 2 cases) and tuberculosis (9.5%; 2 cases) respectively. Other benign cases were pseudo lymphoma (9.5%; 2 cases), stomach GIST (4.8%; 1 cases) and carcinoid tumor (4.8%; 1 cases). Non- Hodgkin lymphoma (28.5%; 6 cases) was the most common malignancy.*
Conclusions: NHL (Non Hodgkin's lymphoma) is most common pediatric gastrointestinal tumor.
Keywords: *Gastrointestinal tract mass, pediatric age group, imprint cytology.*

I. Introduction

Malignant tumors of the gastrointestinal tract especially in pediatric age group are rare and the incidence is less than 1% of all pediatric tumors. [1] It ranks second to trauma as a cause of surgical morbidity in children. [2] The non-specific early symptoms combined with the rare occurrence and variable clinical presentation often poses a diagnostic challenge. [3] Most of these diagnoses were made during urgent exploration for an acute abdominal problem.This is in contrast to adults who have long-standing symptoms such as pain, weight loss, and bleeding. An abdominal mass in a neonate, young child, or adolescent patient can indicate malignancy, and is something that should heighten concern for every pediatrician. A holistic approach includes the identification of the site of localization with respect to the specific anatomy. It also calls for percepting the histopathology based on age of the patient and associated symptoms or signs.

Both adenocarcinomas and adenomas are rare in children and mostly found in adult population. But the gastrointestinal tract (GIT) is the most frequently involved by extra nodal Non-Hodgkin's lymphoma in pediatric age group. Topographically the ileocaecal region, appendix, and ascending colon are the most commonly involved. [4]

II. Material and methods

This prospective observational study was carried out in the Department of Pathology of a medical college in collaboration with Department of Pediatric Surgery from July 2010 to June 2015 including twenty one patients. Institutional ethical committee approval was taken.
Patients' selection:

Twenty one patients attending Pediatric Surgery OPD were included in the present study. Inclusion criteria were 1) patients with age ranges from 0 -12 years (childhood), 2) patients presenting with clinically symptomatic gastrointestinal mass, 3) patients who were candidate for complete surgical removal of the gastrointestinal mass. Patients presenting with peripheral lymphadenopathy, hepatosplenomegaly, altered complete blood picture, abnormal chest radiograph were excluded from the study. The data of every patients including age, sex and presenting symptoms were recorded.

Laboratory work-up & imaging:

Routine hematological investigations including complete blood count and erythrocyte sedimentation rate were carried out. Bone marrow examination along with trephine biopsy was done in suspected malignant cases to exclude marrow involvement. Biochemical investigations included serum lactate dehydrogenase and liver function test. Radiological findings were noted and topography of the tumors was documented according to ICD-10 categories. In case of gastrointestinal lymphoma, staging was done according to St. Jude staging system.

Initial surgery:

Removal of the gross tumor (complete removal) was performed in all twenty one cases. Type of surgery depended on tumor location and surgeon's preference. Treatment modalities of all patients were recorded.

Histopathological examination:

Tissue for histopathological study was obtained in the form of post-operative material. The masses were weighed and measured. After proper processing of the representative sections, slides were stained with Hematoxylin and Eosin stain. Histopathological categorization was done according to WHO classification after examining the slides under light microscope.

Statistical analysis:

Data was analyzed by entering in Microsoft excel 2007 data sheets with SPSS Statistical Software.

III. Results

Clinical profile:

We studied twenty one cases aged between 4 days to 12 years with median age of 6 years. Male to female ratio was 3.2:1.

Table -1: Showing age distribution of gastrointestinal masses in pediatric age group [n=21].

Pain was the main diagnostic symptom in most cases, followed by palpable abdominal mass in both gastric and intestinal tumors. Majority patients had a history of nonspecific chronic abdominal pain and present urgently with an acute exacerbation caused by bowel obstruction (38.1%), intussusceptions (9.5%), or appendicitis (4.8%). "B" symptoms were detected in 75% cases of NHL. Loss of weight was not noted as such but considered as caused by the underlying disease. Median time from onset of symptoms to diagnosis was the shortest for ileocaecal mass and longest for colonic involvement.

Blood parameters:

13/21(61.9%) patients suffered from severe anaemia. Hypoalbuminemia was the next significant biochemical findings. LDH level was more than 1000 U/L in 5/6(83.3%) patients suffering from gastrointestinal lymphoma.

Topographic distribution:

To define the site of origin in GI masses, we distinguished first of all between gastric and intestinal masses. Intestinal masses were subdivided as follows: (1) Duodenum, (2) Jejunum, (3) Ileocecal region, defined as involvement of terminal ileum, cecum, appendix, and/or lower part of ascending colon, (4) Colon, and (5) Rectum. We considered the ileocaecal region and the rectum as separate sites, because they allow localized treatment in curative intention by localized resection. Table 2 also shows that the most common site of gastrointestinal NHL was ileocaecal region.

Table 2: showing distribution of gastrointestinal mass

Histopathological findings:

Diagnosis were broadly classified into two major categories - benign (52.4%; 11 cases) and malignant (47.6%; 10 cases). Among the benign lesions, maximum number cases belongs to mature teratoma (14.3%; 3 cases) followed by inflammatory myofibroblastic tumors (9.5%; 2 cases) and tuberculosis (9.5%; 2 cases) respectively. Other benign cases were pseudo lymphomas (9.5%; 2 cases), stomach GIST (4.8%; 1 cases) and

carcinoid tumor (4.8%; 1 cases). Regarding the malignant categories, Non- Hodgkin lymphoma [Figure 1] (28.5%; 6 cases) was the most common malignancy among which 3 cases of Burkitt lymphoma and 3 cases of DLBCL were found. Maltoma was found in one case only. We had also received the morphological categorization of lymphoid malignancies. We also got single case of yolk sac tumor, rhabdomyosarcoma [Figure 2] and adenocarcinoma [Figure 3] each.

IV. Discussion

Present study of clinico-morphological profile and histopathological examination was done in all cases of primary gastrointestinal tumors in childhood. Primary gastrointestinal malignancies are rare in children as stated by other authors[5]. We came across only 10 such cases during five years study period.

Six of these 10 cases (28.5%) were of primary gastrointestinal lymphomas during our study period. According to different studies, primary gastrointestinal lymphoma is a rare but aggressive tumor making up to 4% of all childhood gastrointestinal malignancies malignancy [6]. Diffuse extranodal involvement is generally noted in pediatric age group [7].

Furthermore, our study included 21 cases aged between 4 days to 12 years. Regarding age distribution, we found 100% of patients of primary gastrointestinal lymphomas belonged to age group of 5-12 years. Bethel et al. [6] postulated that primary gastrointestinal lymphoma most commonly occur between 5 to 15 years age group.

The peak age for abdominal NHL in children is 5-15 years. In our series, the median age for our patients was 8 years, which is similar to the median age of patients reported by Ebeid et al[8] of eight years. As regards to sex distribution, there is a marked predominance of males in all series of childhood NHL [9]. In the present study the male to female ratio was 5:1.

In the present study, the most common presenting symptom was abdominal pain. Ahmed Morsi et al [10] and Pier Luigi Zinzani et al [11] also reported abdominal pain as commonest presenting symptoms.

Regarding topographical distribution, the ileocaecal region was the most common involved site. Amany M Ali et al [12] showed right iliac fossa mass in 25% of cases.

Meticulous microscopic examination was done and Non Hodgkin lymphoma was found as the most common pediatric gastrointestinal malignancy in our study. Our observation was consistent with different literature. [13, 14]

V. Conclusion

It has been seen that ileocaecal region is the commonest site in pediatric gastrointestinal tumors and NHL is the most common type of tumor in pediatric patients. While dealing with such lesions, one should also keep the possibility of tuberculosis particularly in ileo-cecal region.

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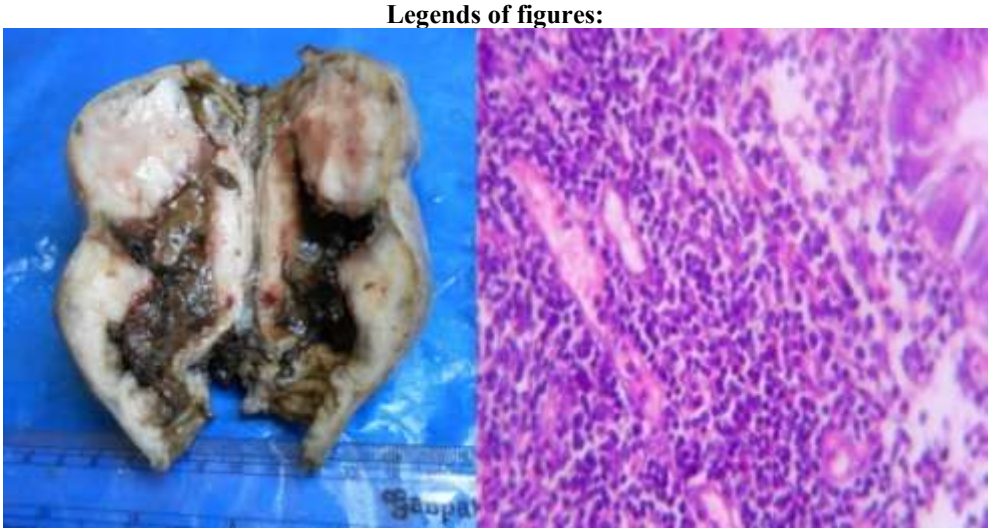

Figure 1: Grosspicture of tumor in histologically confirmed NHL [H&E X400]

Table 1: Showing age distribution of gastrointestinal masses in pediatric age group [n=21].

Age group (year)	Category	Diagnosis
0 - 4	Benign (7)	Pseudolymphoma (2)
		Carcinoid tumor (1)
		Myofibroblastic tumor (1)
		Mature teratoma (3)
	Malignant (1)	Rhabdomyosarcoma (1)
5 - 8	Benign (3)	Stomach GIST (1)
		Tuberculosis (1)
		Myofibroblastic tumor (1)
	Malignant (6)	Burkitt lymphoma (2)
		DLBCL (2)
		Maltoma (1)
		Yolk sac tumor (1)
9 - 12	Benign (1)	Tuberculosis (1)
	Malignant (3)	DLBCL (1)
		Adenocarcinoma (1)
		Burkitt lymphoma (1)

Figure 2: Gross picture of tumor in histologically confirmed Rhabdomyosarcoma [H&E X400]

Table 2: showing distribution of gastrointestinal mass [n=21].

Site	Number	Diagnosis
Stomach	4	Stomach GIST (1)
		Mature teratoma (3)
Jejunum	3	Myofibroblastic tumor (1)
		DLBCL (1)
		Yolk sac tumor (1)
Ileocaecal	11	Burkitt Lymphoma (3)
		Carcinoid tumor (1)
		Pseudolymphoma (2)
		Myofibroblastic tumor (1)
		Maltoma (1)
		Rhabdomyosarcoma (1)
		Tuberculosis (2)
Colon	2	DLBCL (2)
Rectum	1	Adenocarcinoma (1)

Figure 3: Gross picture of tumor in histologically confirmed Adenocarcinoma [H&E X400]