Original Article

DOI: 10.7860/JCDR/2015/10757.5450

Pathology Section

Paediatric Surgical Pathology – a Profile of Cases from Western India and Review of Literature

NILESH SHAH[1], AMI SHAH[2], JAYUL KAMDAR[3], JIGAR SHAH[4]

ABSTRACT

Aim: The paediatric surgical pathology specimens manifest a wide spectrum of morphological and histological features. The present work has been undertaken to know the prevalence and to describe the profile of paediatric surgical pathology specimens from western India as seen in Ahmedabad, India from 2008 to 2010.

Materials and Methods: We reviewed 140 paediatric surgical specimens, 118 specimens rendered definitive diagnosis were included for the analysis. Cases were divided in two groups, one of developmental and congenital conditions and another of acquired lesions.

Results: This study included 118 patients of which 79.3 % were male and 20.7 % were female. Age range of the patients was one day to twelve years. Children of one month to one year age group (infants) were the most vulnerable (31.3% cases). Group of developmental and congenital conditions consisted of 45.7% cases where as 55.3 % cases were of acquired lesions. Gastrointestinal tract was most frequently affected organ (43.2%) followed by head and neck region (14.4%) and testis (7.6%). Hirschsprung's disease (HD) cases (6.7%) were commonest among the group of developmental and congenital conditions followed by juvenile polyps of colon (5%), Meckel's diverticulum of small intestine (5%) and neural tube defect (5%). In acquired lesions, Appendicitis was the most frequent lesion (21.2%) followed by haemorrhagic infarct of testis due to torsion (5%) and intussuception of intestine (5%). Malignant cases were (4.2%) and the most common cancer was yolk sac tumour.

Conclusion: Paediatric surgical specimens, unlike adults, represent significant number of developmental and congenital conditions in addition to acquired lesions; accounting for wide spectrum of morphological and histological features. Study provides insight into the trends of paediatric surgical lesions in the western region of India.

Keywords: Children lesions, Developmental and congenital conditions

INTRODUCTION

Paediatric lesions requiring surgical intervention are of wide variety. They range from developmental and congenital conditions, acute emergencies, benign and malignant neoplasm and other space occupying lesions. Many of the paediatric surgical specimens, both malignant and non malignant are not common and pathologist do not come across them too often. Previous reports has generally been pertaining to case reports or detailing upon a particular lesion but there has been no study providing the overall profile of paediatric surgical specimens received in a histopathology laboratory. Search of literature reveals lack of reports, particularly in context to India, encompassing the profile of paediatric surgical specimen workload. Arguably, prevalence of these lesions is affected by factors like geographical location, tropical climate and socio-economic conditions. In this study authors attempt to study epidemiological characteristics and comprehensive review of histomorphological features of paediatric surgical cases in western India.

MATERIALS AND METHODS

We reviewed 140 cases of paediatric surgical specimens and biopsies that were sent to Rushabh Pathology and Endocrine centre, Ahmedabad, India, during October 2008 to September 2010. Of the initial 140 cases, 118 cases were entered for the study. Five follow-up resection specimens of large bowel from known cases of Hirschsprung's disease whose biopsies had been previously received and diagnosis established were omitted from the study. Two specimens of colostomy closures in patients of Hirschsprung's disease who had undergone proximal diverting colostomy done at the time of diagnosis to relieve the obstruction were excluded. Colorectal biopsies from 14 patients with complain of constipation and were suspected for Hirschsprung's disease, in which, no abnormality was detected were excluded from the study. One colonoscopic biopsy specimens in suspected case of colitis revealing non-specific morphological changes was not included in the study.

Clinical information collected included patient age, sex, chief complaints and its duration, and radiological findings. To correlate development of child with frequency of paediatric surgical lesions, patients were grouped as of age less than or equal to one month (neonate), more than one month to one year age (infants), more than one year to five years age (toddler and preschool), more than five years to ten years age (school going), and more than 10 y age group (preadolescent). Pattern and characteristics of paediatric surgical lesions affecting different age groups was recorded and analysed. The surgically resected specimens and biopsies were fixed in the 10% formalin. Gross examination findings of specimen recorded and representative areas of tissue were subjected to routine tissue processing, paraffin embedding and 5-7 µm thick tissue sections stained with haematoxylin and eosin were prepared and were examined for histomorphological features.

To obtain better understanding of aetio-pathogenesis, based on age of patient at the time of occurrence of the lesion and the histological interpretation, lesions were divided in two groups as developmental and congenital conditions and acquired lesions.

RESULTS

This study included 118 patients of which group of developmental and congenital conditions consisted of 45.7% cases where as 55.3 % cases were of acquired lesions [Table/Fig-1,2]. Age and sex details were available in 113 and 116 patients respectively; 79.3 % were male and 20.7 % were female. Age range of the patients was one day to twelve years. Children of one month to one year age group (infants) were the most vulnerable (31.8% cases). The cohort of patients with developmental and congenital conditions

	n	Male	Female	Age				
				<1 month	1 month to 1 Year	>1 year to 5 years	>5 years to 10 years	>10 years
Juvenile polyps	6	5	1	0	1	3	0	1
Meckel's diverticulum	6	5	1	0	3	3	0	0
Hirschsprung's disease	8	4	3	1	4	2	0	0
Chylous lymphangioma	1	1	0	0	0	1	0	0
Duplication cyst	1	0	1	1	0	0	0	0
Choledochal cyst	2	2	0	0	0	1	1	0
Renal dysplasia	2	2	0	0	2	0	0	0
Spleenectomy for Thalassaemia	1	1	0	0	0	0	1	0
Suprarenal sequestration of lung	1	1	0	1	0	0	0	0
Sacro-coccygeal YST	1	1	0	0	1	0	0	0
Sacro-coccygeal mature teratoma	2	2	0	1	0	1	0	0
Sacro-coccygeal NTD	5	4	1	5	0	0	0	0
Scalp Meningocele	1	1	0	0	1	0	0	0
Atrophy of testis	2	2	0	0	1	1	0	0
Supraorbital Dermoid cyst	4	4	0	1	1	0	2	0
Oral congenital granular cell tumour	1	0	1	1	0	0	0	0
Capillary haemangioma of Parotid gland	1	1	0	0	1	0	0	0
Branchial cyst and sinus	4	4	0	0	2	0	2	0
Bronchogenic cyst	1	1	0	0	0	0	1	0
Thyroglossal duct cyst	1	0	1	0	1	0	0	0
Lymphangioma	1	0	1	1	0	0	0	0
Chest wall dermoid cyst	1	1	0	0	1	0	0	0
Cavernous haemangioma axilla	1	1	0	0	1	0	0	0
Total	54	43	10	12	20	12	7	1

[Table/Fig-1]: Characteristics of cases with developmental and congenital conditions. Age and sex details were available in 51 and 53 patients respectively; n=54

showed two peaks of incidence, one peak in infants (38.4% cases) and another peak in neonates (23%), thus 61.5% of children with developmental and congenital condition were of or below one year age. Group of patients with acquired lesions showed two peaks in incidence, one in school going children (37.7% cases) and another peak in infants (26.2% cases).

Gastrointestinal tract was most frequently affected organ (43.2%) followed by head and neck region (14.4%) and testis (7.6%) [Table/Fig-3].

Hirschsprung's disease cases (6.7%) were commonest among the group of developmental and congenital conditions followed by juvenile polyps of colon (5%), Meckel's diverticulum of small intestine (5%) and neural tube defect (5%). In acquired lesions, appendicitis was the most frequent lesion (21.2%) followed by infarct of testis due to torsion (5%) and intussuception of intestine (5%).

We received 29 specimens related to Hirschsprung's disease; of which 21 were colo-rectal biopsies from patients with complain of constipation and were suspected for Hirschsprung's disease; eight of these biopsies were positive for Hirschsprung's disease. Rectal and colon biopsies were taken from single or multiple sites and number of biopsies per case varied from 1 to 5. All except for two

	n	Male	Female	Age				
				<1 month	1 month to 1 Year	>1 year to 5 years	>5 years to 10 years	>10 years
Intussuception of intestine	6	4	2	0	5	1	0	0
Pseudocyst of omentum	1	1	0	0	0	0	1	0
Cholelithiasis	1	1	0	0	0	0	0	1
Coagulative necrosis of liver	1	1	0	0	0	1	0	0
Microvesicular steatosis of liver	1	1	0	0	1	0	0	0
Hernia contents of Gastric tissue	1	1	0	0	0	1	0	0
Reactive follicular hyperplasia of lymph node	3	3	0	1	0	1	1	0
Tuberculosis lymphadenitis	1	1	0	0	1	0	0	0
Post BCG adenitis of axilla	1	1	0	0	1	0	0	0
Metastatic YST of inguinal node	1	1	0	0	0	0	1	0
Torsion & infarct of ovary	1	0	1	0	0	0	1	0
Testis YST	1	1	0	0	1	0	0	0
Torsion & infarct of testis	6	6	0	1	2	3	0	0
Pyogenic granuloma lip	2	1	1	0	0	1	1	0
Deep folliculitis with fibrosis of cheek	1	1	0	0	1	0	0	0
Hodgkins Lymphoma of Lung and mediastinum	1	0	1	0	0	0	0	0
Tuberculosis cavity of lung	1	0	1	0	0	1	0	0
Suppurative abscess cavity of lung	2	1	1	0	1	1	0	0
Hydatid cyst of lung	1	1	0	0	0	1	0	0
Pleural biopsy, Empyema	2	2	0	0	2	0	0	0
Ewing's Sarcoma of chest wall	1	1	0	0	0	0	1	0
Appendicitis	25	18	7	0	1	1	17	5
Suppurative abscess	2	1	0	0	2	0	0	0
Tubercular abscess	1	1	0	0	0	1	0	0
Total	64	49	14	2	18	12	23	6

[Table/Fig-2]: Characteristics of cases with acquired lesions. Age and sex details were available in 62 and 63 patients respectively; n=64

cases, biopsies were full thickness and included both submucosa and muscularis propria, and rest of the cases biopsies were sero-muscular.

Juvenile polyps were reported in 6 (5%) cases. The age range was 1 to 12 years and maximum patients were from preschool age group (3cases, 50%). There was preponderance of male patients; male female ratio was 5:1. Four patients had single polyp, and the two patients with multiple polyps had two and three polyps respectively. The size of the polyps varied from 0.4 cms to 2.5 cms.

Meckel's diverticulum was reported in 6 (5%) cases. The age range was 7 month to 5 years; half patients were infants and rest of the three cases was from preschool age group. There was preponderance of male patients; male to female ratio was 5:1. Ectopic gastric mucosa was present in three cases (50%) and ectopic pancreatic gland in one case (20%). Complication of intussusception and gangrene was present in three (50%) cases of Meckel's diverticulum.

	n	Male	Female	Developmental and congenital conditions	Acquired lesions
Gastrointestinal tract	51 (43.2)	36	14	20	31
Head and neck region	17 (14.4)	12	3	14	3
Testis	9 (7.6)	9	0	2	7
Sacro-coccygeal region	8 (6.7)	5	3	8	0
Lymphnodes	6 (5)	6	0	0	6
Lung	5 (4.2)	3	2	0	5
Chest wall and pleura	5 (4.2)	5	0	2	3
Renal and suprarenal	3 (2.5)	3	0	3	0
Gall bladder	3 (2.5)	3	0	2	1
Liver	2 (1.69)	2	0	0	2

[Table/Fig-3]: Organ wise distribution of lesions

In this study six (5%) cases were of intussusception of intestine, five patients were below one year age and male: female ratio was 2:1.

Lesions affecting head and neck region consisted of 17 cases; the major bulk was of congenital and developmental cysts (n=10) of neck region, consisting of supraorbital dermoid cyst (4 cases), branchial pouch anomalies (4 cases), thyroglossal duct cyst and bronchogenic cyst one case each.

In five cases (4.2%) the lesions were malignant in nature; the most common cancer was yolk sac tumour (3 cases), one case of Ewing's Sarcoma of chest wall infiltrating the mediastinum and one case of Hodgkins Lymphoma of mediastinum infiltrating the lung. Site of origin for yolk sac tumour was testis and sacrococcygeal region in one case each. The third tumour was detected as metastatic yolk sac tumour in inguinal node biopsy, the primary was not known at the time of diagnosis.

DISCUSSION

Our study retrospectively analyzed three years of paediatric surgical pathology case data in Ahmedabad city which is an important centre catering medical facilities to two large states of western India, Rajasthan and Gujarat.

Of the 21 colo-rectal biopsies from patients of chronic constipation, eight biopsies were positive for Hirschsprung's disease based on absence of ganglion cells and presence of nerve fibre hyperplasia as observed on histologic examination of serial H&E stained sections. Twelve to eighteen levels were studied before confirming the absence of ganglion cells. Koletzko et al., [1] studied interobserver variability in rectal biopsy specimens from patients suspected of colonic motility disorder; except for Hirschsprung's disease, they reported high interobserver variation for different morphological features and for diagnosis of intestinal neuronal dysplasia; and have advocated that rectal biopsy for diagnostic purposes should be performed in constipated children, only to diagnose or to rule out Hirschsprung's disease. Despite the availability of special techniques such as acetylcholinesterase histochemical analysis and S-100 protein and neuron-specific enolase (NSE) immunohistochemical analysis, histologic examination of serial H&E stained sections remains the standard method for evaluating colorectal specimens establishing the diagnosis of Hirschsprung's disease [2]. The male to female sex ratio was 4:3 in present study, however western literature reports male preponderance with male to female sex ratio of 4:1 [3]. The incidence of Hirschsprung's disease varies significantly among ethnic groups [4].

Histologicaly, 90% of colonic polyps in children are juvenile polyps [5]. In this study all the polyps were histologicaly juvenile polyps. Juvenile polyps are generally thought to be hamartomatous lesions with little malignant potential [6]. Three or more than three polyps have been the criteria to label the patients as juvenile polyposis coli, a condition with neoplastic potential [7]. Out of the six cases of juvenile polyps in present study, four patients had single polyp; rest of the two patients had two and three polyps each respectively. But none of the specimen of juvenile polyp revealed adenomatous morphological features. Some case reports have reported osseous metaplasia in juvenile polyps [8], but none of the polyps in our study had metaplastic change.

Meckel's diverticulum is the most common cause of bleeding in the paediatric age group [9]. In present study, Meckel's diverticulum cases showed preponderance of male patients; male to female ratio was 5:1. Previous reports in literature states equal incidence in both sexes [9], but these reports differed from our study as they included both children and adult patients, where as our study was focused on the paediatric cases only. Ectopic gastric mucosa and ectopic pancreatic gland were present in three (50%) and one (20%) specimens of Meckel's diverticulum respectively. In previous reports frequency of presence of ectopic gastric mucosa in Meckel's diverticulum varies from 20% to 57% [10]. Complication of intussusception and gangrene was present in three cases of (50%) Meckel's diverticulum cases which is in congruence with other studies. Complications are more common in males with a diverticulum larger than 2.0 cms [9]. All diverticulums in present study measured more than 2.0 cms in size ranging from 2 to 3.5 cms, and with preponderance of male patients (83.3%) explains high incidence of complications in our cases of Meckel's diverticulum.

Intussusception of intestine is the most common cause of acute intestinal obstruction in infants and young children [11]. In this study six (5%) cases were of intussusception, five patients were below one year age and male: female ratio was 2:1. This is in congruence with the study reported from South India by Bhowmick et al., [12]. Association of natural rotavirus infection with intussuception is uncertain as studies during 1970 to 1980 have acclaimed rotavirus infection as cause of intussuception [13]. A live rotavirus vaccine has been withdrawn due to claims of increased cases of intussuception after its administration [14]. But recent reports and a population based study of intussuception incidence conducted in Delhi has denied any association between natural rotavirus infection and intussuception [15].

Birth Defects Registry of India have analyzed over 0.7 million births, of which, the most common congenital anomaly in the Indian population has been neural tube defect(NTD) [16]. In our study, there was preponderance of male patients; male to female ratio was 5:1. We received six specimens of neural tube defect of which four specimens were of meningocele and two were of meningo-myelocele; of these six lesions, five were from sacrococcygeal region and one from occipital region. However, in this study, NTD does not form the commonest congenital lesion because unlike the birth defects registry, our study cases consisted of only those NTD cases in which foetus were viable. The incidence of NTD has markedly reduced following mandatory administration of folic acid to pregnant mothers [17,18].

Of the eight cases of testis lesion, five cases were of infarct due to torsion of testis and one case each of atrophic testis, of undescended atrophic abdominal testis and of yolk sac tumour. Torsion of the testis carries a poor prognosis; all acutely painful testes need urgent exploratory surgery to salvage the testis. Loss of testis in various series due to torsion has been reported from 55% to 80% [19]. Bennett et al., [20] has attributed delay by the patient or his parents in seeking medical attention as the main reason for the poor prognosis in cases of testis torsion.

The limitations of this study are that due to retrospective nature of the study it does not allow us to identify the aetiology of the lesions. Secondly, despite of judicious search we could not find studies pertaining to profile of paediatric surgical cases in other geographical locations, so as not allowing us to comment upon variations in regional trends.

CONCLUSION

The uniqueness of the present study of paediatric surgical pathology cases lies in the fact that despite of extensive online literature search, similar reports could not be revealed. Paucity of such data is probably because congenital and developmental paediatric surgical pathology specimens and paediatric neoplasm are not frequently encountered in histopathology laboratories and are confined to certain specialised centres only. This study imparts useful insight in epidemiological trends in paediatric surgical pathology cases and provides baseline for the frequency of occurrence of paediatric surgical lesions in western India. Secondly, this study also highlights the challenging diagnostic scenarios in paediatric surgical pathology because of the different disease spectrum encountered in children and adolescents compared with older patients and emphasising the fact that "children are not little adults." This data audit report provides profile of specific diagnostic categories and impart knowledge of common entities to be remembered for differential diagnosis, having an important role in error reduction by the pathologist.

REFRENCES

[1] Koletzko S, Jesch I, Faus-Kebetaler T, Briner J, Meier-Ruge W, Müntefering H, et al. Rectal biopsy for diagnosis of intestinal neuronal dysplasia in children: a prospective multicentre study on interobserver variation and clinical outcome. *Gut.* 1999;44:853–61.

[2] Karim S, Hession C, Marconi S, Gang DL, Otis CN. The identification of Ganglion Cells in Hirschsprung Disease by the Immunohistochemical Detection of ret Oncoprotein *Am J Clin Pathol.* 2006;126:49-54.

[3] Russell MB, Russell CA, Fenger K, Niebuhr E. Familial occurrence of Hirschsprung's disease. *Clin Genet.* 1994;45:231-325.

[4] Torfs CP. An epidemiological study of Hirschsprung disease in a multiracial California population. The third international Meeting: Hirschsprung disease and related neurocristopathies, Evian, France, 1998.

[5] Gelb A, Minkowitz S, Tresser M. Rectal and colonic polyps occurring in young people. *N Y State J Med.* 1962;62:513–18.

[6] Ujjal Poddar, BR Thapa, K Vaiphei, KLN Rao, SK Mitra, Kartar Singh. Juvenile polyposis in a tropical countr. *Arch Dis Child*. 1998;78:264–66.

[7] Giardiello FM, Hamilton SR, Kern SE, et al. Colorectal neoplasia in juvenile polyposis or juvenile polyps. *Arch Dis Child.* 1991;66:971–75.

[8] Brian R. Oduma, Matthew L. Bechtoldb, C, Alberto Diaz-Ariasa. Osseous Metaplasia in an Inflammatory Polyp of the Rectum: A Case Report and Review of the Literature. *Gastroenterology Research*. 2012;5:74-78.

[9] Sagar J, Kumar V, Shah DK. Meckel's diverticulum: a systematic review. *J R Soc Med.* 2006;99: 501-06.

[10] Cserni G. Gastric pathology in Meckel's diverticulum. Review of cases resected between 1965 and 1995. *Am J Clin Pathol.* 1996;106:782–85.

[11] WHO vaccines and biologicals. Acute intussusceptions in infants and children. Incidence, clinical presentation and management: a global perspective. Geneva:*World Health Organization,* 2002:1-98.

[12] Bhowmick K, Kang G, Bose A, Chacko J, Boudville I, Datta SK, Bock HL. Retrospective Surveillance for Intussusception in Children Aged Less than Five Years in a South Indian Tertiary-care Hospital. *J Health Popul Nutr.* 2009;27:660-65.

[13] Mulcahy DL, Kamath KR, de Silva LM, Hodges S, Carter IW, Cloonan MJ. A two-part study of the aetiological role of rotavirus in intussusception. *J Med Virol.* 1982;9:51–55.

[14] Murphy TV, Gargiullo PM, Massoudi MS, Nelson DB, Jumaan AO, Okoro CA, et al. Intussusception among infants given an oral rotavirus vaccine. *N Engl J Med.* 2001;344:564–72.

[15] Bahl R, Saxena M, Bhandari N, Taneja S, Mathur M, Parashar UD, et al. Population-Based Incidence of Intussusception and aCase-Control Study to Examine the Association of Intussusception with Natural Rotavirus Infectionamong Indian Children. *The Journal of Infectious Diseases.* 2009;200:S277–81.

[16] Shamnas M, Arya PS, Thottumkal VA, Deepak MG. Congenital anomalies: a major public health issue in India. *IJPCBS.* 2013;3(3):577-85.

[17] Czeizel AE. Periconceptional folic acid and multivitamin supplementation for the prevention of neural tube defects and other congenital abnormalities. *Eur J Obstet Gynecol Reprod Biol.* 1998;78(2):151-61.

[18] Wilson RD, Johnson JA, Wyatt P, Allen V, Gagnon A, Langlois S, et al. Preconceptional vitamin/folic acid supplementation 2007: the use of folic acid in combination with a multivitamin supplement for the prevention of neural tube defects and other congenital anomalies. *J Obstet Gynaecol Can.* 2007;29:1003-26.

[19] Chapman RH, Walton AJ. Torsion of the testis and its appendages. *Br Med J.* 1972;i:164-66.

[20] Bennett S, Nicholson MS, Little TM. Torsion of the testis: why is the prognosis so poor? *Br Med J (Clin Res Ed).* 1987;294:824.

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Pathology, GMERS Medical College, Sola, Ahmedabad, Gujarat, India.
2. Assistant Professor, Department of Pathology, GMERS Medical College, Sola, Ahmedabad, Gujarat, India.
3. Consultant Paediatric Surgeon, Department of Paediatric Surgery, Jaydeep Hospital, Ahmedabad, Gujarat, India.
4. Consultant Pathologist, Department of Pathology, Rushabh Pathology and Endocrine Centre, Ahmedabad, Gujarat, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Nilesh Shah,
17 Hans Residency, Opp ICICI Bank, Prahladnagar, Ahmedabad-380015, Gujarat, India.
E-mail : setu_n_shah@yahoo.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Aug 04, 2014**
Date of Peer Review: **Sep 27, 2014**
Date of Acceptance: **Oct 06, 2014**
Date of Publishing: **Jan 01, 2015**