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A STUDY OF JUVENILE POLYPS IN Children– Diagnostic Criteria and Difficulties

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ABSTRACT

Polyps in the gastro-intestinal tract are common in children and occur in up to 1% of pre-school and school-going children. Colo-rectal polyps remain one of the most frequent causes of painless rectal bleeding in children. They can cause significant morbidity due to anaemia, protein energy malnutrition, intestinal obstruction and intussusception, which may ultimately affect the overall growth of the child. Juvenile polyps account for more than 90% of all polyps, and are a common cause of painless rectal bleeding in children. Rectum is the commonest site for these polyps. These polyps are usually solitary, but may be multiple. If more than 5, the condition is called as juvenile polyposis coli. While solitary polyps have no malignant potential, juvenile polyposis does carry a risk for malignant transformation. A retrospective study of 63 cases of colorectal juvenile polyps in the age group of 0-15years was done over a 5 year period, at a tertiary care center. This study aimed to evaluate the age of presentation, sex preponderance and common presenting symptoms of juvenile polyps, to study gross morphological features and microscopic variations of juvenile polyps and to review indications for colectomy in juvenile polyps. There were 61 cases of juvenile rectal polyps in our study. 53/61 cases were single, 2 cases had polyps more than 1, but less than 5 and there were 6 cases with more than five colonic polyps, qualifying the terminology of juvenile polyposis coli. Rectum was the commonest site for solitary juvenile polyps accounting to 90.28% of cases. Dysplastic or adenomatous change was not seen in any of the cases of solitary juvenile polyps. 4/6 cases of juvenile polyposis coli showed cauliflower like projections on gross examination, and these showed adenomatous changes on histology. Identification of adenomatous change is important as this would predispose for colonic adenocarcinoma and therefore needs definitive correction.

KEYWORDS : Juvenile polyps, child, polyposis coli

INTRODUCTION

Polyps in the gastro-intestinal tract are common in children and occur in up to 1% of pre-school and school-going children[1]. Colo-rectal polyps remain one of the most frequent causes of painless rectal bleeding in children. They can cause significant morbidity due to anaemia, protein energy malnutrition, intestinal obstruction and intussusception, which may ultimately affect the overall growth of the child. Juvenile polyps account for more than 90% of all polyps, and are a common cause of painless rectal bleeding in children. Rectum is the commonest site for these polyps. These polyps are usually solitary, but may be multiple. If more than 5, the condition is called as Juvenile Polyposis Coli. They are hamartomatous in origin. Mostly they are solitary. If more than 5 polyps are present, juvenile polyposis syndrome should be suspected. Rectum is the commonest site. Solitary polyps are thought to have almost no malignant potential but there is risk of malignant change in polyposis. This study aimed to evaluate the age of presentation, sex preponderance and common presenting symptoms of juvenile polyps, to study gross morphological features and microscopic variations of juvenile polyps and to review indications for colectomy in juvenile polyps.

MATERIALS AND METHODS

A retrospective study of 63 cases of colorectal polyps in the age group of 0-15years was done over a 5 year period, at a tertiary care center. Institutional ethics approval was taken prior to commencement of the study. History, findings on physical examination and results of laboratory, radiological and colonoscopy studies were noted. Gross and microscopic examination findings of polyps was noted. Slides stained by H& E and Masson's trichrome stain (wherever indicated) were studied. Data was tabulated and analyzed.

RESULTS

There were 61 cases of juvenile rectal polyps in our study. 77% cases were male and 27% were female. The mean age of the patients was 9 years. 53/61 cases were single, 2 cases had polyps more than 1, but less than 5 and there were 6 cases with more than five colonic polyps, qualifying the terminology of juvenile polyposis coli. Of the 6 cases with juvenile polyposis coli, 5 cases presented in the first decade, while one case presented in the second decade. Rectum was the commonest site of the juvenile polyps. Of the solitary polyps, 46 were seen in the rectum (83.64%), 3 in the recto sigmoid junction, 4 in the sigmoid colon, totalling to 53 cases. In both cases with multiple polyps < than 5 in number, the polyps were seen in the rectum (3.64%). Therefore rectal polyps were the commonest juvenile polyps accounting to 90.28%.

Painless rectal bleeding was the commonest presentation in cases with solitary juvenile polyp, seen in all the 55 cases (100%). Protrud-

ing anal mass in 14 patients (25%), abdominal pain and cramps in 2 patients (4%) and intestinal obstruction in one patient (2%) were the different presentations seen. Anaemia was found in 7 patients, but, none of these patients had diarrhoea or hypoalbuminemia.

In 52 cases (94.5%), the polyp was pedunculated, and non-pedunculated in 3 cases (5.5%). (Fig.1)The polyps were less than 1 cm in diameter in 41 cases (75%), 1 to 2 cm diameter in 14 cases (25%), but none of the patients had polyps more than 2 cm. The external surface was smooth and globular in 49 cases (89.1%), with polypoid projections in 6 cases (10.19%), greyish white and congested in 48 cases (87.3%) and mucinous and cystic in 7 cases (12.7%).

On histopathology, cystically dilated glands were seen in all the cases. Cystically dilated glands with focal loss of goblet cells and containing proteinaceous material admixed with inflammation, widened lamina propria, with dense mixed inflammatory infiltrate, with plenty of eosinophils were the microscopic features in juvenile polyps.(Fig.2) Most polyps showed surface ulceration and inflammation. Hyperplastic changes were seen in in 30 cases (54.5%), regenerative changes were seen in 40 cases (72.5%), both regenerative and hyperplastic changes in 27 cases (50%) and mucosal ulceration in 30 cases (54.4%). Dysplastic or adenomatous change was not seen in any of the cases of solitary juvenile polyps.

Diagnostic difficulty was faced in 12 cases of patients with solitary juvenile polyps. In 2 cases differentiating between severe hyperplasia and mild dysplasia was difficult. With hyperplastic glands, nuclear stratification, focal nuclei reaching up to luminal surface, distinction of severe hyperplasia from mild dysplasia was difficult.(Fig.3,4) In 4 cases there was dense inflammation and a diagnosis of inflammatory polyp was also thought of initially. But presence of few cystically dilated glands, smooth muscle bundles in lamina propria, absence of features of inflammatory bowel disease in the background and clinical supportive history helped in correct diagnosis. In 6 cases there was a diagnostic dilemma between juvenile polyps and Peutz –Jeghers polyp, wherein there is fanning of muscle bundles from muscularis propria. Peutz –Jeghers polyps, which also hamartomatous polyps, are seen more commonly in small intestine and stomach, but less frequently in colon/rectum. Grossly they are polyps with lobulations unlike juvenile polyps which are not lobulated. Histologically, the polyp is composed of branching bands of smooth muscle covered by normal/hyperplastic glandular epithelium, indigenous to the site, and dividing the polyp in to sectors, while lamina propria is normal.

There were 6 cases of juvenile polyposis. Mean age at diagnosis was 9 years, rectum being the most common site. Bleeding per rectum was present in 5 patients. 3 patients presented with abdominal pain, there was an anal mass in 2 patients, one patient each presenting with features of intestinal obstruction and intussusception. 4/6 cases showed cauliflower like projections on gross examination, and these showed adenomatous changes on histology. There was less stroma, resulting in more glands per unit area with back to back arrangement(Fig.5,6), and lack of the smooth outline of polyps. These findings were reported as focal adenomatous change. None of the cases had severe dysplasia or invasive carcinoma or co-existing pure adenomas.

Discussion

Juvenile polyps are hamartomatous polyps and are the most common type of polyps found in the GI tract[1]. They occur in young children, adolescents but can also affect adults[4]. They can be solitary polyps or less than 5 in number, usually confined to the colo-rectum and not associated with family history of polyps. When they are more than 5 in number, the patient is diagnosed to have juvenile polyposis syndrome[5]. When the patient is < 6 months age, with polyps throughout the gastro-intestinal tract and a non sex linked inheritance, it is called as 'Diffuse juvenile polyposis of infancy'[6, 7]. In a patient who is 6 months to 5 years of age and has polyps throughout the gastro-intestinal tract, it is termed as 'Diffuse juvenile polyposis'. In a patient beyond 5 years of age up to 15 years, when limited to the colo- rectum and with an autosomal dominant inheritance it is called as 'Juvenile polyposis coli'[6, 7].

Over 90% of these lesions are pedunculated[8], spherical, measuring 1-2 cm, rarely more than 3 cm in diameter. Only 10% are sessile[9]. The polyps have a stalk ranging in length from 3mm to 5mm, a smooth, red, friable surface that bleeds easily on ulceration. On cut surface they show characteristic yellowish white mucin filled cysts amidst haemorrhagic areas.

The earliest lesion is an ulceration of surface epithelium and mucosa with increase in number of capillaries and inflammatory cells, neutrophils and eosinophils. This causes secondary occlusion of neck of glands, leading to mucous retention, which causes cystically dilated glands. These cystically dilated glands lead to elevation of mucosa causing polypoidal appearance[9].

Painless rectal bleeding was the most common presentation in our series, as mentioned in literature. Protruding anal mass was the second most common presentation (25%), comparable with Kerr et al[10] & Kennedy et al[11]. Juvenile polyps were located throughout colon and rectum, however, rectum remained the most common site in our study (86.8%), followed by sigmoid colon. This is in agreement with many authors who have mentioned rectum as the commonest site. Solitary polyps were easily removed using a sigmoidoscope.

Multiple polyps were accounted in only 4% of the cases which is within the range mentioned in literature as between 4% and 36%[12, 13,14,15,16]. Polyps were easily removed using a sigmoidoscope. Recurrence rate of solitary polyps is 3% to 18% as reported in literature[12, 17, 18], but was 4% in our study and rectum was the commonest site.

Juvenile polyposis is a rare condition in children, more so in the Indian sub-continent[19]. Family history which is reported to be present in about one-third cases[20], was absent in our cases. This may be because genetic changes responsible for colorectal neoplasias are uncommon in our race or due to small sample size. Although congenital anomalies are more common in non-familial cases of juvenile polyposis than familial ones[1, 4, 21], we have not encountered any congenital anomalies in our study, either due to genetic heterogeneity or due to less number of cases studied.

The average age of diagnosing juvenile polyps is 8 years[22], while it is 15.7 years for juvenile polyps with adenomatous change[23, 24, 25, 26]. 5 out of 6 cases in our study were of age more than 10 years except one case which was in a child 3 of years. The average age in our study was 9 years. 4 out of 6 cases had multilobulated cauliflower like projections on external surface and corresponded with dysplastic changes and were reported as focal adenomatous change. Invasive carcinoma or adenoma was not seen in any of our cases even after examination of multiple polyps in resected colectomy specimens. According to literature, adenomatous changes can occur in 26 to 47%, adenomas in 2% to 15% and carcinoma in 2.5% to 20% of all cases of juvenile polyposis[27, 28, 29].

These patients require long term follow up because of the risk of developing carcinoma (17%) at an early age (mean age as mentioned in literature is 35.5 years)[30]. Colonoscopic polypectomies followed by endoscopic surveillance is a reasonable method of treatment as long as polyp clearance is possible. The choice of treatment will be influenced by the severity of associated symptoms, feasibility of achieving endoscopic clearance and of course, patient compliance [1,27,31] .2 out of 6 of our cases underwent endoscopic polypectomies and were followed up on colonoscopy. In the presence of multiple polyps, as many polyps as possible should be removed colonoscopically and studied microscopically.

There is insufficient data on juvenile polyposis to justify prophylactic colectomy[32] done for risk of carcinoma as done in cases of familial adenomatous polyposis. However in patients with intractable symptoms such as bleeding, protein losing enteropathy, poor compliance, lack of awareness of condition, inability to follow-up and difficulty in colonic clearance, partial colectomies have been reported to have been conducted[1, 27, 33]. 4 out of 6 cases in our study underwent (partial in 3 and total in 1 case) colectomies (Fig.7).

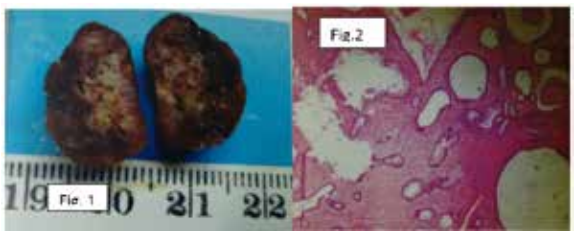
Conclusions

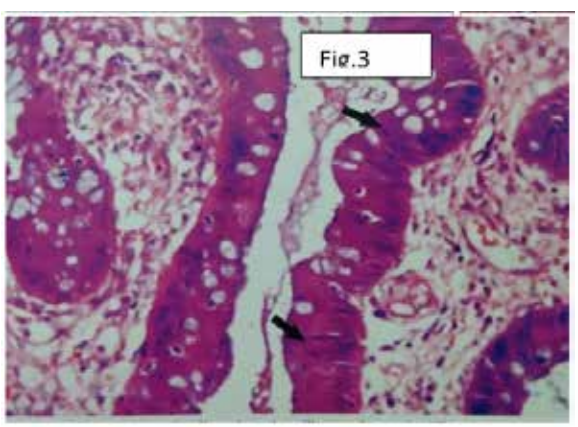
Solitary juvenile polyps usually occur below 10 years of age. There was a male preponderance and rectum was the most common site. Painless rectal bleeding was the clinical presentation in all the cases. The next most common feature was a protruding anal mass. Identification of a polyp as being juvenile polyp is important as surgery is cu-

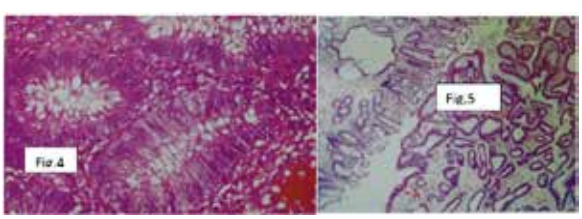
rative even in recurrent cases. Recurrence of symptoms was seen only in 4% cases with solitary polyps, while recurrence was the constant feature in cases with polyposis.

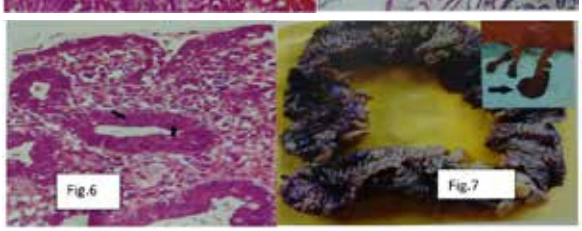
Rectal digital examination and procto-sigmoidoscopy for removal of polyp was sufficient in up to 90% of cases. However in cases of juvenile polyposis coli, intractable symptoms such as loss of blood and protein, stunting of growth, inability to clear a large number of polyps, poor patient compliance, and inability to long term follow up made it essential to carry out colectomy.

Identification of adenomatous change is important as this would predispose for colonic adenocarcinoma and therefore needs definitive correction.









Legends:
Solitary juvenile polyp maintaining smooth, spherical outline and greyish-white, congested cut surface.

A classical juvenile rectal polyp showing cystically dilated glands in edematous, inflamed stroma(H&Ex400X).

Hyperplastic glands with nuclear stratification. Focal nuclei seen reaching towards luminal surface at places (arrows). Distinction of severe hyperplasia from mild dysplasia was difficult (H&Ex400X).

Hyperplastic features in glands in a case of juvenile rectal polyp (H&Ex100X).

Specimen of total colectomy with variably sized numerous polyps, sessile and pedunculated, some showing cauliflower like projections on external surface.

View showing normal to hyperplastic glands (left) to focal adenomatoue change (right), in the same polyp in a case of juvenile polyposis (H&Ex40X).

High power view showing moderate dysplasia (focal adenomatous change) in a case of juvenile polyposis (H&Ex400X).

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