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SMALL INTESTINAL TUBULOVILLOUS ADENOMA- CASE REPORT AND LITERATURE REVIEW

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ABSTRACT
Benign small intestinal tumour, though rare, have been reported. We report a case of sessile and extensive tubulovillous adenoma in a 13-year-old girl. She presented in a private hospital with three months history of abdominal pain, abdominal distension, vomiting, constipation, weight loss and anorexia. Significant findings on examination were weight loss, dehydration, and a huge smooth, not tender**,** mobile and indentable mass which extended from the left lumbar region to right iliac fossa. Visible peristalsis coursing from left to right was seen on the mass. Erect and supine plain abdominal x-rays revealed features of partial intestinal obstruction and abdominal ultrasound scan revealed dilated and hypertrophied bowel segment but could not say the bowel segment affected. The affected segment was found to be a 55cm portion of terminal ileum at operation which was resected due to hypertrophied proximal and collapsed distal segments, features in keeping with chronic intestinal obstruction, and ileo-ileal anastomosis done. Histology report was that of benign tubulovillous adenoma and the girl has enjoyed stable health for more than a year on close follow up in surgical outpatient clinic. This case highlights the unusual presentation and unusual gross nature of this small intestinal adenoma, which was found to be a benign adenoma on histological examination.

Key Words: Small intestine, benign tubulovillous adenoma. *(Accepted 22 January 2008)*

INTRODUCTION
Benign small bowel tumours are rare. Adenoma accounts for approximately 25% of benign small intestinal tumours, with benign mesenchymal tumours (especially leiomyomas), lipomas and neuromatous lesions following in frequency [1]. Polyp is a general term used to describe any projection arising from flat mucosa into intestinal lumen. It is broadly classified into benign and malignant neoplasm and also as familial adenomatous polyp, which may be premalignant, or malignant and non familial polyp which is usually benign [2-4]. Adenoma similar to those of the large bowel can develop in the small bowel but their frequency is said to be minuscule with the duodenum and jejunum more commonly affected than ileum [5,6]. They can be single or multiple, pedunculated or sessile. Microscopically, those that have the appearance of an adenomatous polyp are said to be tubular adenoma, those that have villoglandular polyp appearance are referred to as tubulovillous adenoma or villous adenoma depending on the relative amount of villous architecture. Most adenomas are periampulary. There is a distinct familial predisposition in such cases and the common age range affected are those between 30 and 60 years [5,7,8]. Like its counterpart in the large bowel, the smalladenoma is regarded as a premalignant lesion [8-10].

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They are generally asymptomatic and some are discovered incidentally during radiological investigations. A significant number, however, produces microscopic faecal blood loss. They are rarely large enough to cause obstructive signs in the absence of malignant change. From literature search, not many publications have been done on small intestinal adenoma in this subregion. We report this case to highlight the diagnostic difficulty and strengthen clinicians' suspicion of this diagnosis when considering the differentials of intraabdominal tumours with features of intestinal obstruction.

CASE REPORT
Miss S. D, a 13-year-old girl, presented with three months history of colicky abdominal pain and distension, vomiting, constipation, weight loss and anorexia. The colicky abdominal pain was maximal at periumbilical region and minimally relieved by vomiting of bilious stale food. Abdominal distension, weight loss and anorexia were progressive but no absolute constipation. There was no history of bleeding or passage of mucoid stool per rectum. Examination revealed a wasted girl, dehydrated, afebrile, not pale and not jaundiced. Temperature was 36.8°c, pulse rate 82 per min. and B.P 100 / 60mmHg. Abdomen was distended with visible peristalsis. A huge, smooth, not tender, mobile and indentable mass was palpated extending from left lumbar region to right iliac fossa. Rectum was empty and gloved finger smeared with hard faeces. Plain abdominal X-rays revealed multiple air-fluid levels

with gas distribution down to the rectum and a huge speckled radioopaque mass. Abdominal ultrasound scan revealed a hypertrophied bowel loop with thickened wall filled with faeces and gas. Haematocrit was 10.5? and blood chemistry was normal.Laparotomy revealed dilated hypertrophied terminal ileum which was pale and rubbery in consistency. The Hypertrophy extended 8cm from ileocaecal junction, and involved 55cm of the ileum (Figure 1). There was no pedunculation, mesenteric nodes were enlarged and other intraabdominal viscera were normal. There was gross dilatation of the proximal and markedly collapsed distal segments which was in keeping with chronic intestinal obstruction that necessitated resection of about 70cm ileum with ileo-ileal anastomosis done . Histology of the specimen revealed a loop of markedly distended terminal ileum with hypertrophied wall which showed benign neoplastic lesion composed of tubular and papillary structure covered by normal mucous secreting columunar epithelium with a fibrovascular connective tissue base and moderate chronic inflammatory cells infiltrate. There was a reactive hyperplasia of the underlying lymphoid follicles. The muscularis propria was hypertrophic while the serosa contained some inflammatory cells. These features were in keeping with Tubulo-villous Adenoma (Figure 2). The girl did well and was discharged on day eight after operation. She gained weight rapidly and has been on follow up in surgical out patient clinic for more than a year.

Figure 1: **Photographs of the resected bowel segment showing different views, the arrows pointing at the pale bowel with tumour and 'X' showing normal dilated proximal bowel segment.**

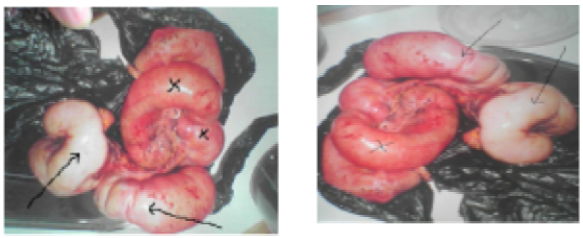
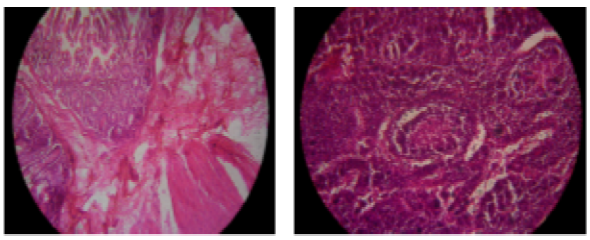


Figure 2: **Photomicrographs showing serial sections of the bowel wall with features in keeping with benign small intestinal adenoma.**



DISCUSSION

Juvenile polyps were first described by Verse in 1908 [11]. For many years all polyps in children were considered adenomas and were often treated by radicalprocedures. It was subsequently discovered, however, that only adenomatous polyps associated with juvenile polyposis syndrome have malignant potential in children [11,12]. Approximately 95% of colorectal carcinoma in adult are believed to arise from adenoma while the overall chance of developing carcinoma in small intestinal adenoma is estimated at 5% [2,9,13]. Several factors have been suggested to explain both the scarcity of adenoma in small bowel and the infrequency of their malignant transformation. Firstly, rapid intestinal transit through the small bowel limits contact time to the small bowel mucosa. Secondly, greater fluidity of small bowel chyme may dilute luminal irritants. Thirdly, alkaline pH may play a role, as may the low bacteria colony counts of the small bowel. Fourthly, higher levels of benzyl peroxidase (thought to detoxify potential carcinogens) have been detected in the small bowel and fifthly the increased levels of immunoglobulin A and widespread gut lymphoid tissue may impede the growth and development of tumours and their malignant transformation [14,15]. Haigis et al [16] reported that intestinal adenomas can develop with a stable karyotype and microsatellites even though stable karyotype has been believed to be protective against adenoma growth and progression to malignant change. Adenomatous cells are characterized by loss of normal growth control. They continue to proliferate as they reach the top of the crept, and they are not extruded into the lumen. Instead, they multiply and eventually fold back into the surrounding normal mucosa, inducing a response in the mesenchymal tissue that helps shape the microscopic architecture of the adenoma. The rate of progression of adenoma to cancer is variable but has been reported to occur within five to ten years. Patient with inherited forms of the disease such as familial adenomatous polyposis (FAP) can have a more rapid rate of adenoma formation and progression to carcinoma [2]. Steinbrecher et al [17] reported that the expression of guanylin is down regulated in mouse and human intestinal adenomas and that this gives rise to the loss of normal control and progression to malignancy. The size of the adenoma also determines its malignant potential as carcinomatous change is rare in adenoma less than 1cm and is estimated at 40-50% in villous adenoma larger than 4cm. The dysplasia-adenoma-carcinoma sequence occur in the setting of increasing loss of heterozygosity in genes involved in DNA replication accuracy (mismatch repair) chromosomes 2 and 3; tumour suppression-chromosomes 5, 18 and 17; and oncogene activation-chromosomes 5, 17 and 18.The higher the degree of dysplastic changes in adenoma, the higher the chance of progression to carcinoma formation [11]. Intracellular sphingosine kinase 1 was postulated by Kohno et al [18] to play a significant role in this progression and proliferation. Truly benign small bowel tumours are rare. If tumours with malignant potential are excluded, a small group of truly benign lesion remains.Analysis of 56 cases seen by William et al [14] revealed that lipoma was most common,

Followed by myoepithelial hamartoma, Peutz-Jeghers hamartoma, neurogenic tumours, Brunner's gland abnormalities and inflammatory fibroid polyp in decreasing order. Small intestinal adenoma usually affects patients who are between the ages of 3o and 6o years with occult blood loss in stools, or as incidental findings during routine radiological examination of the gastrointestinal tract. There is equal sex and racial distribution and the lesion is hardly seen in children [19,20]. The duodenum (especially periampullary region) and jejunum are the regions most commonly affected and very rarely the ileum. Most cases have positive family history. The index case did not fall into these groups. The lesion was seen in a 13-year-old girl, no positive family history and the terminal ileum was the only portion affected. The gross appearance of the tumour is even more contrasting as shown in figure 1. The growth permeated the bowel segment involved and it was not polypoid, contrary to the polypoid growth seen in adenomatous polyps. The diagnostic difficulty with this unusual tumour was resolved by histological examination of the bowel segment as shown in figure 2. The rapid rate of recovery and the stable health enjoyed for more than a year suggest this was a truly benign adenoma.

In conclusion, a case of unusual benign small intestinal adenoma in a 13-year-old girl who had bowel resection with ileo-ileal anastomosis; and rapid recovery, enjoying stable heath for more than a year after operation, is presented to highlight the diagnostic difficulty; and strengthen clinicians' suspicion of this diagnosis when considering the differentials of intraabdominal tumours with features of intestinal obstruction.

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