Chronic intussusception in a young adult with cystic fibrosis

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Introduction

A case of chronic intussusception is described in an 18-year-old boy with cystic fibrosis (CF).

Case report

The patient was diagnosed as having CF at the age of one year on the basis of failure to thrive and a positive sweat test. Following diagnosis he was commenced on pancreatic supplements and regular physiotherapy. At the age of 12 years, *Pseudomonas aeruginosa* was regularly recovered from his sputum, and he was commenced on 3 monthly intravenous antibiotics. He was transferred routinely to the adult CF unit at the age of 16 years. At the time of transfer he was taking 2-3 Pancrease tablets with each meal, Ketovite tablets and vitamin E 100 mg daily. He had a normal dietary intake and opened his bowels once a day. He lived at home with his parents and two brothers. One of his brothers had CF and had had two episodes of intussusception. The patient was extremely fit and played fullback at rugby for his school. His weight was 74 kg and his height 1.9 m. His forced expiratory volume in one second was 4.75 l/min (100% of predicted) and his vital capacity was 5.5 l/min (100% of predicted). On 27 October 1987 he was admitted to another hospital with acute abdominal pain, constipation and episodic vomiting. His mother was sceptical of a provisional diagnosis of appendicitis. Plain abdominal radiographs showed no evidence of intestinal obstruction and an abdominal ultrasound showed sludge in the gallbladder with a normal liver and spleen. No diagnosis was made and he was treated conservatively with enemas and intravenous fluids. After 6 days he was transferred to Monsall Hospital. On admission he was apyrexial and not distressed. His abdomen was soft with a fullness in the epigastrium, bowel sounds were active and he was having episodes of intermittent colic requiring intramuscular opiates. A serum amylase and a white cell count were normal. He was treated with oral Gastrografin. On the second day a mass was distinctly felt in the epigastrium which was mobile and slightly tender. On the third day the patient volunteered the information that whilst playing rugby 2 days before being admitted to hospital he had fallen onto the rugby ball and subsequently been trampled upon. Over the next 8 days he had several ultrasound examinations and a computed tomography scan of the abdomen. Serial plain abdominal radiographs showed Gastrografin passing through the large and small bowel with a suggestion of a soft tissue mass in the epigastrium with no evidence of intestinal obstruction. A transverse ultrasonogram to the left of the umbilicus showed a mass with a target or 'bullseye' pattern with concentric rings (Figure 1).

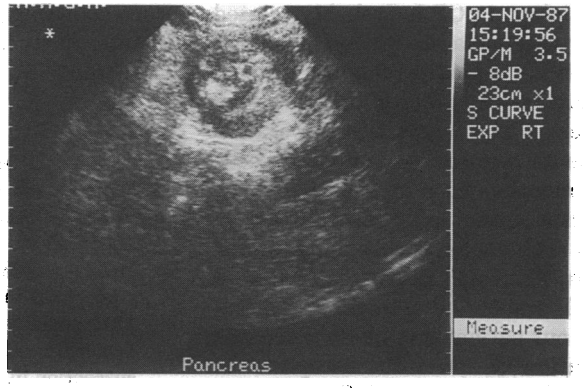

Figure 1. A transverse ultrasonogram to the left of the umbilicus shows a mass with a 'target' or 'bullseye' pattern with concentric rings. There is an echolucent or surrounding ring

Computed tomography showed a soft tissue mass in the same location as the sonogram with Gastrografin present in two segments of small bowel. The diagnosis was unclear. It was noted that Gastrografin was able to pass through the whole of the gastrointestinal tract. Over the next 8 days the mass diminished in size and he was able to open his bowels and take a light diet. He still had intermittent pain which required injections of pethidine. On the 9th day he was discharged home. At outpatient review one week later he was continuing with a light diet, and opening his bowels. A mass could still be felt to the left of the epigastrium. It was smaller and felt to be compatible with a resolving haematoma around the bowel, probably inflicted during his rugby match. On 19 November 1987 he was readmitted with severe abdominal pain and vomiting following a meal of cheese omelet. Bowel sounds were active. In view of the severity of the pain laparotomy was performed on 20 November 1987 one month after commencement of symptoms. At laparotomy he was found to have an ileo-caecal intussusception. He had a resection of 6 cm of small bowel and 15 cm of large bowel. His recovery was slow. He was immediately started on Creon capsules postoperatively and he did not open his bowels for a week. Following discharge he initially had frequent bowel motions probably due to resection of small bowel. His Creon capsules were increased to 10-12 with each meal. Over the preoperative period of one month he had a weight loss of 13 kg. Six weeks after surgery and with an increase in Creon capsules with meals his weight had returned to 77 kg. He has continued to play competitive rugby.

Discussion

Acute intussusception is a common abdominal emergency in early childhood. It accounted for

5 hospital admissions a year to one district hospital with a peak incidence at 6 months. An editorial on acute intussusception did not mention CF as one of the causes[2]. The aetiology of intussusception is idiopathic in approximately 90% of children but it is suggested that enlarged lymph nodes relating to a viral infection in the wall of the small intestine may act as a lead point for the intussusception.

In one series of 2200 patients with CF the incidence of intussusception was reported as 1%[3]. The presenting symptoms in this group were crampy intermittent abdominal pain (77%), a palpable mass (68%), vomiting (57%) and rectal bleeding (23%). Three patients had chronic symptoms lasting for longer than one month. The aetiology in CF is probably related to insufficient pancreatic replacement and abnormal intestinal mucous. It is interesting that our patient had no bowel symptoms prior to hospital admission but following surgery with increased postoperative pancreatic supplements, his body weight now exceeds his weight three months prior to surgery. It is probable that intussusception is an unusual sequel to meconium ileus. The cause of chronicity in our patient was unclear. The colon in patients with CF is larger than usual, probably due to chronic faecal overloading, and in our patient it is likely there was spontaneous resolution of his intussusception as shown by the complete passage of Gastrografin through his bowel and the return to a light diet. Diagnosis was also delayed by the consideration that the mass was related to his rugby injury and the rarity of intussusception in adults. Previously, diagnosis of intussusception was confirmed and sometimes treated by barium enema. However, it is now appreciated in occasional reports that intussusception has a characteristic appearance of a bullseye pattern on ultrasound[4,5]. It is a reliable non-invasive method to confirm or exclude intussusception, and is as sensitive as a diagnostic barium enema.

Computed tomography may show a similar appearance, but this does not have greater diagnostic value and is more expensive. With the advent of microspheres for pancreatic supplementation, the clinical incidence of meconium ileus is declining and correspondingly intussusception should become a greater rarity if the enzymes are given in an adequate dosage.

References

1 Thomas DFM. The management of childhood intussusception in a district hospital. *Br J Surg* 1980;67:333-5
2 Editorial. Acute intussusception in childhood. *Lancet* 1985;i:250-1
3 Holsclaw DH, Rockmans C, Shwachman MD. Intussusception in patients with cystic fibrosis. *Pediatrics* 1971;48:51-8
4 Mulvihill DM. Ultrasound findings of chronic intussusception in a patient with cystic fibrosis. *J Ultrasound Med* 1988;7:353-5
5 Miller JH. Randall Kemberling C. Ultrasound of the pediatric gastrointestinal tract. *Semin Ultrasound, CT, MR* 1987;8:349-65