Unusual presentations of acute appendicitis in the neonate

A report of 2 cases

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Summary

Two neonates with acute appendicitis have been treated at the Paediatric Surgery Clinic, Tygerberg Hospital, during the past 6 months. One presented with red blood in the stool and had had persistent anorexia for 1 week. The second baby presented with a large tender swelling of the scrotum which was indistinguishable from a torsion of the testis or strangulation of an inguinal hernia. Both babies made an uneventful recovery.

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Diess, cited by Hemalatha and Spitz, first described acute appendicitis in a neonate in 1903, and as recently as 1952 Meigher and Lucas reported the first neonate to survive this. In 1960 Schaupp et al. reviewed the English literature and could find only 19 cases of neonatal appendicitis, adding 5 cases of their own; up to 1980 a total of 44 cases had apparently been reported. A recent review by Srouji and Buck revealed that a total of 106 cases of acute appendicitis in neonates had been reported between 1901 and 1975.

An extremely rare form of appendicitis in infants is the scrotal type, of which Alvear and Rayfield could only find 1 case reported in the literature before 1976, adding 5 cases of their own (in a 5-year-old child). In neonates this rare condition has a mortality rate of 70-83%.

We present our 2 cases in order to caution that acute appendicitis should always be considered in the differential diagnosis of acute abdomen in the neonate. The only means of lowering the high mortality rate are early diagnosis and appendicectomy.

Case reports

Case 1

A 1-week-old White male infant weighing 2300 g was referred because red blood had been seen on the nappy. The pregnancy and vaginal delivery had been normal and no defects had been noted at birth. The baby remained listless and would not suck adequately. A tentative diagnosis of ‘infection’ was made by the practitioner and oral antibiotic therapy commenced. The baby’s general condition improved, but when 10 days old he passed red blood in the stool. There was no diarrhoea at any stage.

On examination no cyanosis, jaundice, lymphadenopathy, cardiac failure or congenital defects were detected. The baby was apyrexial and abdominal examination revealed no tenderness or free fluid, a 1 cm hepatomegaly, mild distension of the abdomen, no blood on rectal examination and no palpable abdominal mass. After sedation there seemed to be a ‘fullness’ in the right iliac fossa. A barium study showed that the ileocaecal junction was displaced, suggesting pressure from outside, but intussusception could not be excluded.

At laparotomy a mass was found which involved most of the appendix vermiformis and pressed onto the medial wall of the ascending colon. The appendix and accompanying mass were resected. Histological examination confirmed an acute purulent inflammation of the appendix. The postoperative course was uneventful.

Case 2

A 23-day-old Coloured infant was referred to us, having had a reducible right inguinal hernia since birth. The baby had been crying intermittently for 3 days and the mother had observed peristaltic waves across the abdomen which worsened with feeding and were associated with pain, vomiting and a mild fever. The scrotal swelling had increased in size and was irreducible. The baby passed a normal stool the day before admission.

On examination the baby was 10% dehydrated and there was no cyanosis, hepatosplenomegaly or lymphadenopathy, but he was irritable with a large, very tender right scrotal mass with a dark to bluish discoloration of the overlying skin. The abdomen was mildly distended with absent bowel sounds. The right testicle could not be palpated because of the scrotal tenderness and swelling.

A radiograph of the abdomen showed an ileus. Haematological examination revealed a leucocyte count of 16 x 10^9/l, a haemoglobin concentration of 10.5 g/dl, a base excess of 5 and a pH value of 7.39. The rest of the special investigations were negative. The clinical picture was that of incarceration of the inguinal hernia, and a transverse inguinal incision was therefore made and the appendix was sent for histological examination, which subsequently confirmed the diagnosis of acute appendicitis with perforation at the tip. Escherichia coli was cultured from the pus. The infant made an uneventful recovery.

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Discussion

In children and adults acute appendicitis is characterized by Murphy's triad of pain, vomiting and fever. As regards the neonate, there is a communication barrier, and abdominal distension, fever and vomiting may be the only symptoms signifying acute appendicitis (among other surgical and medical emergencies). Peritonitis followed by ileus may present with bile-stained vomiting or abdominal distension (as in case 1), with the features of peritonitis being difficult to elicit, thus making it difficult to distinguish peritonitis from septicaemia or other conditions associated with an ileus.

These vague abdominal symptoms directly contribute to the difficulty in establishing an early diagnosis; gangrene and perforation of the appendix may occur before a diagnosis is made. The literature indicates that the perforation rate in neonatal appendicitis is 79.4%, with an attendant mortality rate of 82.3%. The appalling mortality is probably related to: (i) the inability of an undeveloped omentum to isolate and wall off intra-abdominal infection; (ii) increased mobility of the caecum in some cases; (iii) the relatively large size of the appendix in the neonate; (iv) underdevelopment of the baby's immune system; and (v) failure to consider acute appendicitis in the differential diagnosis of the acute abdomen in the neonate.

The rarity of appendicitis in neonates is believed to be a result of the funnel shape of the appendix in the first year of life. This precludes occlusion of the lumen by lymphoid hyperplasia, fecoliths, oedema, parasites, kinking or a foreign body, but the configuration of the neonatal appendix is probably only one of a multitude of factors preventing occlusion of the lumen.

The ingestion of breast-milk by the neonate furnishes a significant quantity of antibodies, the function of which is the curtailment or minimization of infection in general, and of the gastro-intestinal tract in particular. Antibodies provided by the mother are supplemented by the baby's own immunoglobulins, particularly that fraction secreted by the gastro-intestinal tract. The neonatal infant is therefore adequately furnished with the necessary defence mechanisms against most intestinal infections.

Obstruction of the colon, as in Hirschprung's disease, is a known cause of perforation of the appendix in the neonate, as illustrated by the cases reported by Martin and Perrin and Gästrin and Josephson. In our 2 cases no associated disease of the small or large bowel could be demonstrated. Some authors believe that perforation of the appendix is a complication of necrotizing enterocolitis, which should always be excluded before a diagnosis of isolated acute neonatal appendicitis is entertained. In our experience incarceration of inguinal hernias, especially on the right side, is common during the first few months of life, but the presence of the appendix in the sac is a very rare cause of the 'acute scrotum' in infancy.

In our second case the appendix had become strangulated in the hernial sac with subsequent perforation, rendering the clinical picture quite indistinguishable from that of torsion of the tests or epididymo-orchitis. Acute appendicitis presenting with a scrotal mass is so rare that Alvear and Rayfield could find only 1 reported case (an infant aged 6 weeks) in the literature before 1976. They added another case of their own (in a 5-year-old). Srouji and Buck published a similar case report in 1978, the patient being a 12-day-old infant, and also refer to 3 neonates with hernial appendicitis presenting as a scrotal sinus or fistula.

Diagnosis and pitfalls

Important problems in the diagnosis of neonatal appendicitis are the communication barrier and the extent to which other medical and surgical emergencies present with the same symptoms as those most commonly seen in neonatal appendicitis, i.e. vomiting, irritability, abdominal pain, anorexia and fever. It is therefore mandatory to exclude medical conditions in arriving at a correct surgical diagnosis.

Careful four-quadrant examination of the abdomen of the infant usually reveals point tenderness in the right iliac fossa, but because of congenital malrotation of the bowel the tenderness may be localized in any of the four quadrants. Distension of the abdomen is common in neonatal appendicitis, with local tenderness and a mass being found in about 50% of cases and bowel sounds ranging from normal to absent. Rebound tenderness can be expected in the late case, when the inflammatory process extends with involvement of the peritoneum and adjacent structures, and the tenderness is confirmed by rectal examination.

Full haematological examination is mandatory because an increase in neutrophils indicates acute peritonitis or some other pyogenic infection, while viral disease is more likely to result in an increase in the mononuclear fraction. The prodromal phase of measles is known to mimic acute appendicitis in the older child, and unnecessary appendicectomy is often performed during this phase of the disease.

The urinary tracts of all neonates presenting with vague abdominal symptoms should be investigated in order to exclude congenital abnormalities which may be complicated by infection, pain and/or tenderness. Mesenteric adenitis is rarely diagnosed in the neonate. Meckel's diverticulitis has a course similar to acute appendicitis, and the patient may present with blood in the stool, tenderness, fever, an abdominal mass and vomiting; the diagnosis is not infrequently confirmed only by surgical exploration.

Pneumonia of the right lower lobe may cause referred pain with spasm of the abdominal muscles, but it is important to bear in mind that acute appendicitis may coexist with pulmonary infection. In these cases point tenderness is absent and a chest radiograph will often establish the diagnosis.

A similar clinical picture is sometimes seen in septicaemia or early tetanus, but point tenderness is absent and a septic umbilical stump may alert the clinician to this possibility. A septic umbilical stump may also cause general peritonitis, pelvic vasculitis and pelvic abscess formation. Examination of a blood culture will be required to exclude septicaemia.

Neonatal intussusception is rare, but colicky pain, an abdominal mass and blood in the stool (as in case 1) are classic findings; the diagnosis can be confirmed by barium examination.

Necrotizing enterocolitis is common in premature babies, or may follow exchange transfusion in normal-for-dates babies or infants with the respiratory distress syndrome or shock, and may produce perforation of the appendix. A diagnosis of acute appendicitis can sometimes be established only by carefully monitoring the course of the disease; in some cases the final diagnosis will only be established at laparotomy.

A marked difference exists between intra-abdominal and hernial appendicitis with or without incarceration. Hernial appendicitis in the neonate has a survival rate far superior to that of intra-abdominal appendicitis, this being attributable to earlier diagnosis of the former; for obvious reasons, intra-abdominal appendicitis is usually diagnosed late.

The blood in the stool in case 1 was probably caused by the inflammatory nature of the mass and its proximity to the medial wall of the caecum. The inflammatory process probably caused oedema and venous congestion with resultant intraluminal collection of blood-stained fluid which was passed per rectum, mimicking the clinical picture of neonatal intussusception.

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Tuberculosis of the distal colon
A case report

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Summary
A case of tuberculosis of the distal colon is described. The presenting features and the sites of the lesions are unusual even in a country where gastrointestinal tuberculosis is common.

Attention is drawn to the diagnostic difficulty encountered, as the lesion is often clinically and radiologically indistinguishable from amoebiasis, carcinoma or Crohn’s disease.

Gastro-intestinal tuberculosis is not uncommon among the lower socio-economic groups in this country. It most commonly involves the ileocaecal region. Involvement of the colon, especially the distal colon and rectosigmoid region, is rare.

The clinical spectrum of gastro-intestinal tuberculosis is wide, and the patient may present with abdominal pain, a mass, obstructive symptoms, diarrhea or malabsorption. It may also closely mimic diseases such as Crohn’s disease, carcinoma and amoebiasis.

Barium enema examination of the patient with colonic tuberculosis may reveal ulcerative disease or a segmental thickening of the bowel wall giving rise to a localized constriction, and the lesion may even resemble the so-called ‘apple-core’ appearance of carcinoma.

In this report a case of tuberculosis involving two separate segments of distal colon is described. The clinical and radiological findings are discussed and the diagnostic difficulties emphasized.

Case report
A 38-year-old Black man presented with a history of intermittent diarrhea for 2 years and severe bloody diarrhea for 1 week, associated with malaise, anorexia and marked weight loss. He denied symptoms of night sweats or coughing.

Physical examination revealed an ill and dehydrated man with a temperature of 37.4°C. A mass of matted lymph nodes was palpable in the right axilla and supraclavicular region. Apart from a pulse rate of 108/min and a blood pressure of 100/55 mmHg, examination of the cardiovascular, respiratory and central nervous systems revealed no other abnormalities. The abdomen was distended and tympanic, with tenderness and guarding in the left loin and both iliac fossae. The liver was enlarged 4 cm. Rectal examination revealed a thickened anterior wall mucosa, and the test for occult blood was positive. Laboratory tests on admission showed a white cell count of 5.0 x 10^9/l, a platelet count of 500 x 10^9/l, and a haemoglobin value of 11.7 g/dl with hypochromic cells on the peripheral smear. The ESR was 7 mm/1st h. Serum electrolyte, urea and creatinine values were within the normal range. The total protein level was 58 g/l (normal 60-80 g/l) and the albumin level 23 g/l (normal 35-50 g/l). Amoebic complement fixation and Widal tests were negative and the Mantoux test was positive. Stool culture grew Shigella flexneri, type 3.

A chest radiograph showed old tuberculous foci in one lung with no evidence of re-activation. A plain film of the abdomen revealed air-filled large bowel and segmental narrowing in the descending colon. A single-contrast barium enema examination demonstrated two markedly abnormal areas. There was circumferential narrowing with ‘shouldering’ in the descending colon and an extensive lesion of similar character in the rectosigmoid region. (Figs 1 and 2). The intervening segment of colon appeared normal.

Sigmoidoscopy revealed only oedematous and indurated mucosa anteriorly at 6 and 12 cm. A biopsy of the lesion showed infiltration of lymphocytes and plasma cells with no evidence of caseation or malignancy. However, biopsy of the supraclavicular

REFERENCES