Intussusception is one of the most common surgical emergencies encountered in infancy and early childhood. This is a condition where the proximal segment of the bowel telescopes into the distal segment causing obstruction. The classic triad of symptoms consists of abdominal pain, vomiting and blood in stools. It is often seen in children aged four months to two years, with a peak incidence during four to nine months of age. However, infants may present with non-classical symptoms such as listlessness, pallor, decreased feeding, and being non-specifically unwell. Three cases of intussusception in young infants who presented with being listless and had some or no features to suggest a clinical diagnosis of intussusception are described which are designed to highlight the non-classical features of intussusception likely to be encountered in very young infants.

Case 1

A healthy six-month-old male infant, presented with intermittently looking very pale and being poorly responsive over an eight-hour period. He had non-bilious vomiting on six different occasions and his mother reported blood spots in the nappy earlier in the day. He was reported to be fully immunised and had been weaned early on to solid food at around four months of age. Intussusception is one of the most common surgical emergencies encountered in infancy and early childhood. This is a condition where the proximal segment of the bowel telescopes into the distal segment causing obstruction. The classic triad of symptoms consists of abdominal pain, vomiting and blood in stools. It is often seen in children aged four months to two years, with a peak incidence during four to nine months of age. However, infants may present with non-classical symptoms such as listlessness, decreased feeding, non-specifically appearing unwell, looking pale, etc. We present three cases of infants with intussusception, who presented with being listless and pale, with a view to heightening the awareness of this condition.

The initial assessment revealed a very pale looking infant with a heart rate of 158/min, temperature of 36.1°C, tachycardia and prolonged capillary refill time. He was given a fluid bolus of 20mL/kg with 0.9% sodium chloride. He was intermittently responding to his parents but was not irritable. His cardiovascular and respiratory examination was otherwise normal.

He was distressed by an abdominal examination and was found to be drawing up his legs and crying while being examined. The provisional diagnosis at that point was either sepsis, gastroenteritis or a suspected intussusception. A chest and abdominal film (FIGURE 1) was organised which showed clear lung fields. The abdominal X-ray looked very abnormal with paucity of gas. Subsequently he passed fresh mucousy blood per rectum which resembled a 'redcurrant jelly' stool, hence a clinical diagnosis of intussusception was made. He needed a fluid bolus of 20mL/kg with 0.9% sodium chloride.
Further fluid bolus of 20mL/kg of 0.9% sodium chloride and was started on intravenous cefotaxime. The blood investigation results showed a C-reactive protein (CRP) of 9mg/L, white cell count of 14.5 x 10^9/L, platelets of 395 x 10^9/L and haemoglobin of 11.7 gm/dL.

He was referred to the regional paediatric surgical unit where a diagnosis of intussusception was confirmed. He had a failed trial of reduction of the intussusception by hydrostatic pressure and needed an open surgical reduction. He made an uneventful recovery and was reported to be well at further follow-up.

**Case 2**
A healthy four-month-old female infant, presented with intermittently looking very pale and listless for about six hours. She was reported to be not feeding well. She had no history of vomiting or diarrhoea, no cough or cold, and did not have a fever. She had had the first set of primary immunisations and had been weaned early on to solid food at around 3½ months of age. The initial assessment revealed a very pale looking infant with a heart rate of 148/min, temperature of 36.9°C, respiratory rate of 48/min, oxygen saturation of 94% in air, blood glucose of 5.8 mmol/L and a central capillary refill time of <2 seconds. She had a patent airway and was breathing comfortably in air. She was responding appropriately to her parents and was not irritable. The cardiovascular and respiratory examination was otherwise normal. She disliked an abdominal examination and was found to be drawing up her legs and crying while being examined. The provisional diagnosis at this point was one of sepsis or a suspected intussusception.

An abdominal film (FIGURE 2) was organised which showed multiple loops of small bowel, with a soft tissue impression in the right side which was suspicious for an intussusception mass.

She remained stable and was given a fluid bolus of 20mL/kg of 0.9% sodium chloride. The blood investigation results showed a CRP of <1 mg/L, white cell count of 12.2 x 10^9/L, platelets of 765 x 10^9/L and haemoglobin of 10.9 gm/dL.

She was referred to the regional paediatric surgical unit where a diagnosis of intussusception was confirmed on abdominal ultrasound scan. She had a successful reduction of the intussusception by hydrotstatic pressure and made an uneventful recovery. She was reported to be well at further follow-up.

**Case 3**
A healthy 4½-month-old male infant presented with intermittent shrill crying episodes for about eight hours. He had had pyloric stenosis at five weeks of age and had laparoscopic pyloromyotomy, but represented a week later with vomiting which resolved with open pyloromyotomy.

He had had non-bilious vomiting on two occasions during his present illness and a blood streaked offensive stool earlier in the evening.

The initial assessment by the casualty officer revealed a pale looking infant, listless with a heart rate of 158/min, temperature of 36.1°C, respiratory rate of 36/min, oxygen saturation of 99% in air, blood glucose of 6.7 mmol/L and a central capillary refill time of two seconds. There was no increased work of breathing.

The review by the paediatric team revealed an irritable but listless child with normal systemic examination. He disliked an abdominal examination and had tenderness on deep palpation. The provisional diagnosis at that point was gastroenteritis or a suspected intussusception. An abdominal X-ray (FIGURE 3) was abnormal with paucity of gas.

Overnight he passed blood and faeces on one more occasion. He was kept nil by mouth and given intravenous maintenance fluids. His CRP was 12mg/L, white cell count 8.4 x 10^9/L, platelets 383 x 10^9/L and haemoglobin 11.2 gm/dL. He had an abdominal ultrasound scan which confirmed the diagnosis of intussusception (FIGURES 4 and 5).

He was referred to the regional paediatric surgical unit where he had a failed trial of reduction of the intussusception by air enema and needed an open surgical reduction. He made an uneventful recovery and was reported to be well at further follow-up.
Discussion

Historical background

Intussusception was first described by Barbotte of Amsterdam in 1674. Hunter in 1789 discussed the surgical pathology of intussusception. The exact year of surgical reduction of intussusception remains controversial – 1784 according to Hutchinson but 1672 according to Ashhurst. In 1876 Hirschsprung first described the reduction of intussusception by hydrostatic pressure methods. In July 1999, reports of intussusception associated with the rotavirus vaccine led to suspension of the programme.

Epidemiology

Intussusception is the commonest cause of acute intestinal obstruction in children under two years of age with less than 25% of cases occurring in children aged more than two years. Intussusception has a male predominance with a male to female ratio of 4:1. If left untreated intussusception has been reported to be uniformly fatal within two to five days. No clear racial predilection is reported, however children with sickle cell disease may be more prone to the condition.

Aetiology and associated conditions

The aetiology of intussusception remains unclear and is idiopathic in most cases. There are two main theories forwarded viz dietary and infective. The dietary theory arises from the observation that intussusception largely occurs around the time of weaning and early weaning has been considered to be a risk factor. The infective theory is probably explained by a history of mild respiratory or gastrointestinal tract infection preceding the onset of intussusception, which may cause inflammation of abdominal lymph nodes and that of the Peyer’s patch (FIGURE 6).

Many authors believe that a lead point in the intestine allows a bowel segment (intussuscipiens) with its mesentery to telescope into the adjacent distal segment (intussusceptum) causing the obstruction (FIGURE 7). When these segments become strangulated, mucosal bleeding can occur thus producing the classic ‘redcurrant jelly’ stool.

However a specific lead point is not found in 90% of cases and thus the dietary and infection theories may play a role. Cases where a lead point may be identified can be seen in Henoch-Schönlein Purpura.

vasculitis, appendix, Meckel’s diverticulum, intestinal polyp, coeliac disease, etc. Intussusception may also occur in starvation (during religious fasts in certain communities), dehydration secondary to severe gastroenteritis, cystic fibrosis and in sickle cell crisis.

Interestingly a lead point is often demonstrated in older children presenting with intussusception.

Foreign body ingestion in young children may lead to intussusception and specific caution needs to be taken in the event of multiple magnet ingestion.

Clinical presentation

The classic triad of intussusception consisting of abdominal pain, vomiting and bloody stools, is seen in less than one-third of patients. Interestingly 75% of children have two symptoms while another 13% will have just one symptom. Abdominal pain typically precedes the vomiting, occurs in paroxysms every 15 to 30 minutes and can last up to 15 minutes during which time the child becomes inconsolable and draws up the legs. Abdominal examination may be normal early in the disease course. Later the abdomen may become distended and tender, and palpation may reveal a sausage shaped mass in the right upper quadrant while the lower quadrant feels empty (Dance sign). Neurologic manifestations are becoming a recognised entity in intussusception especially in very young infants and these generally present early.

Patients often present with non-specific symptoms such as looking mottled or pale, irritability, decreased appetite, listlessness or lethargy. Clinicians may face a diagnostic dilemma in children presenting solely with neurologic manifestations and may investigate for causes of sepsis or neurological conditions thus delaying the diagnosis of intussusception. In this group the physical examination may be entirely normal except for the infant being lethargic or listless.

Intussusception should be considered in the differential diagnosis of young children presenting with lethargy and/or hypotonia, even though the classical signs may be absent.

Chronic intussusception presents with a varying combination of symptoms including abdominal pain, vomiting, weight loss/failure to thrive, diarrhoea, and blood per rectum and is a treatable cause of failure to thrive. It is an uncommon condition and often diagnosed late. An abdominal mass may or may not be palpable and the classic triad of symptoms is uncommon.

Diagnosis

Laboratory investigations are not helpful in diagnosing intussusception. However, these may be helpful in differentiating the condition from other causes like sepsis, metabolic disorders, etc. Also use vomiting may result in electrolyte imbalance which can be detected in laboratory tests.

Radiological investigations are the mainstay for diagnosing intussusceptions. Plain radiographs are often the fastest investigation available and may show the typical target sign (in right upper quadrant, appears as a faint circle within a circle, similar to a doughnut), absence of the liver edge, crescent sign and bowel obstruction. A normal abdominal X-ray does not exclude intussusception.

Contrast or air enemas have traditionally been the diagnostic test and often can be therapeutic in reduction of the intussusception (FIGURE 8).

Ultrasonography is fast becoming the investigation of choice and is often used as it is non-invasive with no radiation involved and can also detect other pathologies. In a study of 141 children in Australia, 95 (67%) cases were diagnosed with ultrasonography and 31 cases (22%) were diagnosed with air or barium enema.
Management
A child presenting with intussusception needs assessment and appropriate resuscitation as the initial presentation may be unclear and may mimic other serious pathologies. Some children presenting with shock will need fluid resuscitation and in non-specific sepsis-like presentations or cases where peritonitis or perforation is suspected, antibiotic therapy should be started. A nasogastric tube insertion may be helpful.

Non-surgical management includes reduction of intussusception with air enema or with contrast enema. Any type of hydrostatic reduction resolves the intussusception by exerting pressure on the apex of the intussusceptum to restore it to its original position. Different studies highlight success rates between 50-95%. Air enema can have success rates up to 90% however 10% of patients go on to develop intussusception again later. The success rates decrease with repeated attempts at using air enemas and surgical intervention may be necessary.

Surgical management involves exploratory laparotomy where the intussusception is manually reduced (FIGURE 9). However, delayed presentations can cause bowel ischaemia and necrosis necessitating a resection of the bowel segment. Recurrence of obstruction after a surgical reduction is uncommon.

Complications
With treatment the mortality rate may vary between 0 to 3% depending on the duration of onset of intussusception and its reduction and also associated shock and sepsis. There is a subsequent risk of small bowel obstruction caused by adhesions in up to 7% of cases.

Conclusion
Intussusception can present a diagnostic dilemma especially in younger children presenting with non-classical symptoms. Early weaning practices need to be addressed with caution by the healthcare professionals and the current national guidelines of weaning on to solid foods around six months of age may help to decrease the occurrences of intussusception. An early diagnosis is associated with a better outcome.

References

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