Short Course

Intussusception—Current Concepts

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Introduction

INTUSSUSCEPTION IS THE INVAGINATION of a proximal bowel segment (intussusceptum) into an adjacent distal segment of intestine (intussuscipiens).[1] This telescoping of one bowel segment into another can produce obstruction, ischemia, and eventual strangulation of the invaginated portion.[2] The entity of intussusception was first clearly detailed by John Hunter[3] in 1789 in his essay "On Introsusception." Hunter reported three cases of intussusception that all resulted in death. Although his anatomic descriptions of telescoping bowels were quite accurate, his suggested therapeutic measures were rather misguided. He recommended bleeding the patient (to decrease inflammation) and inducing vomiting in an attempt "to invert the peristaltic motion of the containing gut, which will have a tendency to bring the intestines into their natural situation."[3] It is known today that such actions are not only futile but might also be harmful to these often critically ill patients.

Epidemiology

The incidence of intussusception in the United States is 2.4 cases per 1,000 live births.[2] It typically occurs in infants and young children between the ages 4 months and 2 years with a peak incidence between 6 and 9 months of age. Ninety-five percent of all intussusception cases occur in the pediatric population, making it the most common cause of intestinal obstruction in young children. Sixty-five percent of all patients with intussusception are less than 1 year old, and 80% of patients are under the age of two.[4] In addition, there is a male predominance (3:2) as well as midsummer and midwinter incidence peaks. Such observations suggest that a specific infectious etiology may be involved, but clear data to support this theory have not been forthcoming.[1]

Pathophysiology

In 90% of cases, no intrinsic abnormality is found (idiopathic intussusception). More than 80% of cases are ileocolic intussusception.[2] A popular theory attempts to explain this "idiopathic" pathophysiology. It is believed that a viral illness (upper respiratory tract infection or gastroenteritis), usually adenovirus or rotavirus, results in hypertrophy of lymphoid nodules (Peyer's patches) in the terminal ileum.[5] A hypertrophied lymphatic patch then behaves as a lead point and becomes drawn into the lumen of the terminal ileum (and then into the ascending and transverse colon).[6] Edema formation and inflammation follow, finally culminating in bowel obstruction. In addition, the mesentery can become compressed between bowel walls, with potential development of vascular insufficiency.[1] Complications such as incarceration, strangulation, and perforation of the intussusceptum can occur and can be fatal.

There are reported cases in the literature of idiopathic intussusception in which no histologic evidence of lymphoid hyperplasia was found. Nissan et al. propose a cytokine-based mechanism to explain such cases.[5] It is well known that lipopolysaccharide (LPS) activates nitric oxide (NO) synthase leading to increased NO levels. It is hypothesized that these increased NO levels alter intestinal motility and promote intussusception.[5] LPS was injected intraperitoneally into mice and 25% of the animals demonstrated intussusception in the small or large intestine. Furthermore, addition of arginine (NO precursor) increased the incidence of intussusception to 30.7%, whereas only 3.8% of the mice that received L-NAME (NO synthase inhibitor) developed intussusception. In addition, prostaglandins exert a variety of effects on intestinal muscles. For example, PGE_2 causes contraction of longitudinal muscle but relaxes circular muscle, while PGF_{2a} contracts both layers.

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Interestingly, injection of the cyclooxygenase inhibitor indomethacin completely prevented intussusception, suggesting a role for prostaglandins in intussusception induction.[5]

Clinical Presentation

Typically, an otherwise healthy infant of 6–9 months of age develops sudden severe colicky abdominal pain with intermittent vomiting.

There are periodic episodes of irritability and crying with the infant drawing up both legs onto the abdomen every 20–30 minutes.[1] Between bouts of colic, the young child may initially appear to be comfortable and even playful; but, if the intussusception is not reduced, he will become progressively more ill-appearing, demonstrating lethargy, weakness, and exhaustion.[6] A classic triad is often described, consisting of abdominal pain, vomiting (bilious in late occurrences), and currant jelly stool (feces mixed with blood and mucus).[2] Some authors suggest an alternative triad of abdominal pain, abdominal mass (sausage-shaped), and bloody stool.[7,8] Regardless of which triad is preferred, clinicians must be aware of the fact that neither one is a reliable diagnostic indicator and are present in less than one-half of the cases. In fact, in 1992, Stringer's multi-study analysis (1,164 cases) revealed that the classic triad was present in only 32% of the cases.[9]

Diagnosis

There is a pathognomonic physical finding in intussusception known as Dance's sign, named after the French physician Jean Dance.[6] It is the finding of an elongated mass in the right upper quadrant of the abdomen with absence of bowel in the right lower quadrant on palpation. A rectal examination is of utmost importance for two reasons and should not be neglected. First, in a small percentage of cases, there is actually a palpable intussusceptum that has progressed to the rectum. This diagnosis can be made on physical examination alone. Second, the detection of occult blood furthers the clinician's suspicion of the diagnosis, and 85% of patients indeed have guaiac positive stools.

The differential diagnosis of intussusception is extensive and can be divided into six categories:

1. Infectious causes—enteritis, colitis, appendicitis, pancreatitis, pyelonephritis, glomerulonephritis, and right lower lobe pneumonia.
2. Obstructive causes—malrotation with volvulus, obstructing adhesions, and incarcerated hernias.
3. Abdominal trauma.
4. Hematologic disease—leukemia and lymphoma.
5. Vasculitic disease—Henoch–Schonlein purpura.
6. Miscellaneous—ovarian/testicular torsion, hypokalemia.[10]

Daneman and Alton[7] explored some of the controversial issues related to the diagnostic imaging of intussusception and offered their recommendations. The abdominal radiograph, although a popular test, is often unhelpful. Certain signs suggestive of intussusception include presence of a soft-tissue mass, empty right lower quadrant, proximal distention with air fluid levels, and loss of visualization of the tip of the liver. However, abdominal radiographs correctly identify intussusception in only 45% of cases. Nevertheless, it is a helpful test to exclude perforation in a child with abdominal pain and vomiting. Enemas (both barium and air) are diagnostic with close to 100% accuracy. Barium outlines the leading portion of the intussusceptum giving a "coiled spring" appearance. Enemas also have the added benefit of therapeutic reduction (see Treatment section below). They are, however, contraindicated in cases involving clinical findings of peritonitis, shock, or signs of perforation on abdominal radiograph.[7]

Ultrasonography is now being used more frequently to diagnose intussusception.[11] Two recent American studies examined the diagnostic effectiveness of ultrasonography. In 1992, Bhisistkul and Verschelden independently reported 100% sensitivities with 88% and 93% specificities, respectively, when using sonography to diagnose intussusception. More recently, color Doppler sonography has been used in an attempt to determine whether a particular intussusception is likely to be reducible or not. It has been suggested that the absence of blood flow (color) might indicate the presence of ischemia and necrosis and that attempts at enema reduction in such patients should be avoided to obviate the potential risk of perforation.[7]

Ultrasonography offers several diagnostic advantages as compared to enema, namely, lack of radiation exposure, lower cost, and no harmful potential. In addition, ultrasonography can sometimes identify other potential etiologies for the patient's symptoms

including appendicitis, ovarian sources, and urinary tract sources.[2,7] Disadvantages of ultrasonography include lack of therapeutic benefit, and accuracy of results depend on the radiologist's skill. Verschelden, however, showed that a 100% diagnostic accuracy rate could be achieved by third-year and fourth-year residents who have completed a 3-month to 5-month training program in sonography.[7]

Treatment

The controversy over barium enema (hydrostatic reduction) versus air enema (pneumatic reduction) for management of intussusception is nearly universally resolved: air enema now is the recommended treatment of choice in experienced hands. What advantages does air offer as compared to barium? First, air enemas are cleaner and thus technically easier to perform.[12] Next, better reduction rates have been achieved with air enemas.[13]

A 1992 literature review by Stringer revealed a 50% to 78% success rate with barium enema versus a 75% to 94% success rate with air enema.[7] (Recurrence rates were equal for barium and air, approximately 5%.) In addition, the patient is exposed to a lower radiation dose with air enema: the average fluoroscopy time for pneumatic reduction was 94.8 seconds which was one-half to one-third less than with the hydrostatic method.[12] Finally, and perhaps most important, air enemas possess a greater safety profile than that of barium. The dreaded complication of enema reduction is perforation, and perforation rates of barium and air are about equal (<1%). However, if perforation does occur, much less fecal spillage and peritoneal contamination occur with pneumatic reduction as opposed to hydrostatic reduction. This may relate to the fact that, because of its lower viscosity, air escapes (unaccompanied by feces) much more rapidly causing less extension of the serosal tear than occurs with the more viscous barium, which moves more slowly taking stool with it.[7,14]

Because enema reduction yields such high success rates with concomitant low complication rates, surgery is not recommended as the primary treatment. Operative reduction is necessary, however, for those patients in whom radiologic reduction is unsuccessful, for those where a pathological lead point (e.g., tumor or diverticulum) is suspected, and for those with multiple recurrences. A laparotomy is performed with manual reduction (milking) of the intussusception followed by resection of nonviable bowel segments and/or lead points.[2] The intussuscepted bowel is next examined for adequate reperfusion. Finally an appendectomy is usually performed[1] in ileocolic cases to prevent the risk of developing future appendicitis. The postsurgical recurrence rate varies from 0 to 4%.[2]

Conclusion

When a 6–9 month old child presents with abdominal pain, vomiting, and bloody stool, with or without an abdominal mass, intussusception should be on the top of the differential diagnosis. In some centers, ultrasonography is recommended for diagnosis, whereas at others diagnosis is followed by treatment using barium or pneumatic reduction (air enema). Surgery should be reserved only for those cases where reduction is unsuccessful or where peritonitis is suspected.

References

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