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Adult Intussusception: A Disease Often Uncovered in the Operating Room

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Authors' contributions

This work was carried out in collaboration between both authors. Author VJ had reviewed the literature pertaining to the discussed topic and had compiled it in a logical and chronological order. Author MB had reviewed and revised the composition with changes regarding structure and content. Both authors read and approved the final manuscript.

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ABSTRACT

Intussusception is the prolapsing of one bowel segment into another. It is a common entity in the pediatric population, but is rare in adults representing only 5% of cases. A variety of causes are attributable to this disease process, with benign lesions predominating in small bowel cases and malignancy being largely responsible for intussusception of the colon in adults. It is often difficult to diagnose due to the variable onset, presentation of symptoms and possibly intermittent nature, and thus is often revealed at emergency laparotomy. Most surgeons agree that the optimal approach to management is surgical resection with oncological principles due to the high incidence of malignancy-associated intussusception in adults.

Keywords: Intussusception; bowel obstruction; surgery.

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1. INTRODUCTION

Intussusception is the prolapsing of a proximal segment of bowel (intussesceptum) into the lumen of the distal segment (intussuscipiens). It is relatively common and benign in the pediatric population, with only 5% of all cases occurring in adults [1]. In this subset, intussusception is the cause of 1% to 5% of bowel obstructions [1,2] and the published literature pertains predominantly to case reports with brief reviews. In one case series conducted by Pavlidis et al. [2] it was encountered in 1.9% of operated intestinal obstructions in adults. It is an important topic for review due to relative inexperience with such a disease resulting in later diagnosis. Early identification may improve plans of management and reduce complications associated with delay.

It has been estimated that 2 to 3 cases occur in a population of 1,000,000 per year, accounting for 0.1% of all hospital admissions [3]. In the adult population, the mean age at diagnosis is in the sixth decade of life [4-6] with no preponderance for either sex, [5] supporting an association with certain malignancies of the gastrointestinal tract.

Presentation differs considerably from the classic-triad of abdominal pain, bloody diarrhea and palpable mass with tenderness characteristic in pediatric patients, [1] occurring in only a fraction of adults and often ambiguous in nature. The non-specific symptomology and variable presentation has made this entity difficult to diagnose, and is often incidentally revealed during emergency surgery.

2. ETIOLOGY AND PATHOGENESIS

The causes of adult intussusception span a wide spectrum of insults, statistically dependent on specific location. The pathogenesis of intussusception among these causes seem to share a common mechanism. In general, organic pathology serves as a leading point in 70% to 90% of cases, [1,7] with the remainder attributable to physiologic abnormalities [3] or idiopathic [1]. It is estimated that of these organic lesions, primary or secondary malignant lesions comprise 6% to 30% of overall insults inducing intussusceptions in the gastrointestinal tract [3,7].

It is generally believed that a lesion or irritant of the bowel wall alters peristalsis [1]. As a result, a local area is unable to contract in a normal fashion, and unbalanced peristalsis rotates the intestinal wall inwards [Fig. 1] [8]. Usually this occurs with an organic lesion serving as a lead point, demonstrated by the overall incidence of malignancy found at the apex of an intussusception (43-56%) [4]. Intussusceptions without a leading point, as in the malabsorption syndromes, produce prolapse through dilated bowel loops with increased secretion altering peristaltic motion [8]. Subsequently, telescoped bowel prevents the free passage of contents in often-associated resulting the bowel obstruction [9] at the same time as compromising vascular flow in the mesentery [Fig. 1] [1].



Fig. 1. Demonstration of ileocolic intussusception with a polyp acting as the leading point

Note: Inflammation secondary to ischemia and vascular compromise are illustrated with compression of mesenteric vasculature. Bowel obstruction may occur as the telescoping results in narrowing of the bowel lumen

Malignancy is an important factor in management intussusception. Such lesions include of carcinoma. lymphoma, malignant fibrous histiocytoma [2] and metastatic disease [1]. Benign etiologies include a wide array of offending insults such as polyps, Meckel's diverticulum. colonic diverticulum, stricture. lipomas, leiomyoma, hemangioma, Peutz-Jeghers syndrome, [2] inflammatory bowel disease, postoperative adhesions, abdominal trauma, [3] and iatrogens like intestinal, feeding tubes [1] and even gastric bypass [10].

Benign lesions predominate in the small bowel comprising 60% of enteric intussusception, followed by malignancy (30%) and idiopathic causes (10%) [11]. This is likely because tumors of the small bowel are rare, contributing to only 1% to 2% of gastrointestinal malignancies [12]. However, this small fraction of small bowel malignancies may occur with metastatic melanoma, metastatic esophageal carcinoma, lymphoma, leiomyosarcoma, carcinoid tumor, neuroendocrine, stromal tumor [13] and even metastatic lung carcinoma [1,10].

In contrast, the majority of lesions inducing colonic intussusception are associated with malignancy (60%) [11]. This coincides with the relatively more frequent incidence of colorectal cancer as compared to malignancies of the small bowel [8]. In the colon, the most common benign lesion is the lipoma, [14] but even rare lesions from mycobacterial infections can also precipitate this process [15]. In such cases, the majority are solitary submucosal lesions with 75% of them localized to the right colon [14].

Overall, intussusceptions are generally classified into four categories: enteroenteric involving solely the small bowel, colocolic involving solely the large bowel, ileocolic involving prolapse of the ileum into the ascending colon and ileocecal involving the ileocecal valve as the leading point. [1] Most often, it occurs at the communications between free segments, regions fixed to the retroperitoneum or by adhesions [1]. Approximately 90% are in association with the small bowel and colon, with the minority in the stomach or surgically placed stoma [3]. The least common type of intussusception is gastroduodenal, associated with a benign gastric tumor [3]. Of special note, the incidence of intussusception following gastric bypass ranges from 0.1% to 0.3% [10]. Interestingly, in such cases no leading points are found, and it is believed that the cause is disruption of the natural gastric pacemaker and thinning out of mesentery as a lack of resistance to intussusception once it begins [10].

3. CLINICAL MANIFESTATION

In the pediatric population, the classic triad of abdominal pain, bloody diarrhea and a palpable mass with tenderness is often reported in bowel intussusception [1]. This presentation is rarely seen in adults and has been described in 20% of cases [14]. More specifically, a palpable abdominal mass is evident in 10% to 42% of cases, [3,16] with melena or positive fecal occult blood test in another 30% of cases [5].

In adults, the presentation is variable in terms of symptoms, onset and duration. The most common symptom is abdominal pain, present in 90% of adult intussusception [3]. This pain is of non-specific nature however, and a diagnosis is only reached preoperatively in 40% to 65% of patients [4]. The next most common symptoms in adults include nausea, vomiting, gastrointestinal bleeding, bowel habit changes, constipation and abdominal distension in association with obstruction [1]. Organic lesions causing leading points have been correlated with bowel obstruction of persistent and relapsing nature, requiring definite surgical management [1]. Lesions not associated with a leading point, such as celiac sprue or Crohn's disease tend to present with non-obstructing intussusception of transient nature, and often cumulates in spontaneous resolution [1].

In addition to the lack of specificity for the presentation of this disease, it frequently manifests in a periodic manner, contributing to the delay in diagnosis [3]. Not only does the degree of intermittency vary, but also so does duration. Symptoms can endure from 1 day to 365 days, [17] with a mean of 37.4 days in one study [5]. Intussusception tends to endure for longer periods of time in benign or enteric lesions when compared to malignancies or cases of the colon [3,5]. To the best of our knowledge, Begos et al. [18] reported the longest duration in literature at five years. The non-specific nature of presentation warrants investigation of more common entities before suspicion for intussusception is entertained [4]. Thus, intussusception has proven difficult to diagnose accurately and time-effectively.

4. DIAGNOSIS

The difficulty in diagnosing adult intussusception is exhibited through the preoperative diagnostic rate of 50% and 40.7% as reported by Riejnen et al. [19] and Eisen et al. [6] respectively. This is predominantly due to the often-negative findings on physical examination, leading to the frequent diagnosis of irritable bowel syndrome [3]. As a result, this condition is most often correctly diagnosed intraoperatively by the surgeon in the emergency setting for intestinal obstruction [3,7].

Abdominal roentgenography is often the first modality entertained in diagnosis. It may demonstrate signs of obstruction and the location. Upper gastrointestinal series may elucidate the typical signs of a "stacked coin" or "coil spring" appearance [1]. The use of barium enema may pertain specifically to the filling defects apparent with colocolic or ileocolic intussusceptions [1]. However, this is not the most accurate modality available, providing an estimated diagnostic yield of only 41% [20]. Similarly, Balik et al. [21] conducted a study over a 10-year period, concluding air-fluid levels attributable to bowel obstruction in 68.7% of patients presenting with acute symptoms secondary to intussusception.

Ultrasonography has proven useful in determination of certain abdominal disease processes. It is used in the pediatric and adult population alike for detection of intussusceptions [22]. The hallmark is the appearance of the "target" or "donut" signs [1]. Diagnostic yield for this modality is estimated at 32% by Huang and colleagues [20]. However, other studies had determined a much higher diagnostic yield for intussusception using ultrasonography, as high as 100% [23] as reported by Lim et al. [24] and Balik et al. [21]. Although this modality is easily employed in diagnosis, sensitivity varies and has associated disadvantages. For instance, it typically requires an experienced practitioner for manipulation and interpretation is limited by patient factors such as obesity [1] and dilated bowel loops with air causing poor transmission of signal [23]. Ultrasonography has a userdependent variability and bias in identification of intussusception results in a generally accepted range of sensitivity from 50% to 75% [10]. However, the advantages advocating for its utilization include relative speed, lack of exposure to ionizing radiation, low cost and ability to assess vascular compromise with Doppler studies [21].

The most widely employed diagnostic modality for this disease in the preoperative setting is computed tomography (CT) scanning. It is considered the most accurate modality for diagnosis of adult intussusception outside of the operating room [1]. Sensitivity has been demonstrated with a diagnostic rate of 58% to 100% of cases [1,5]. The characteristic features appreciated are similar to those of in ultrasonography. A "target sign" or soft-tissue density in the "sausage" shape with layering of tissue are the predominant features [1]. This is attributable to the dense nature of edematous bowel and mesentery [23]. With the use of intravenous contrast, mesenteric vasculature may also be appreciated to assess the degree of ischemia and inflammation [17]. Lesions with compromised mesenteric vasculature are more likely to be associated with non-neoplastic insults, and these patients are candidates for reduction prior to surgical resection of the bowel [25]. Of similar importance, a study by Kim et al. [26]

determined that CT was able to distinguish between intussusception with lead points and those without fairly accurately. The implication of this pertains to planning management and whether reduction may be attempted prior to surgical intervention. For patients with renal disease or unable to tolerate intravenous contrast, CT findings with a hypodense layer in the returning wall of the intussusception, fluid and gas accumulated in the space surrounding this wall are indicators of vascular compromise detectable without the use of contrast [26,27].

Endoscopy has been considered invaluable in the evaluation of subacute or chronic lower bowel obstruction [1]. Confirmation, localization and demonstration of organic lesions are achieved effectively with this modality of diagnosis [1]. However, it is useful only for the lower gastrointestinal tract, in which many leading points may be caused by polyposis [1]. The complication associated with such polyposis is that snare polypectomy in the setting of chronic vascular compromise is high risk for perforation due to ischemia and necrosis [1,28].

The elusive nature of this disease has resulted in a large portion of cases diagnosed at exploratory laparotomy, and is thus often managed in the same setting.

5. MANAGEMENT

Preoperative CT scan may be effectively used to ascertain the necessity of surgery based on imaging findings that demonstrate benign lesions [26]. With such etiology, reduction is safer when signs of inflammation or ischemia of the bowel wall are not present, [29] and is achieved by milking the bowel from the distal segment in the proximal direction [30].

Given the nature of etiologies in adult intussusception, 70% to 90% require surgery as definitive treatment [1]. However, most surgeons have limited experience with such an entity, and there is debate with regards to whether reduction should be attempted prior to surgical intervention [3,24]. It is believed that with a significant fraction of cases secondary to malignancy, reduction increases the risk for seeding of the omentum or abdominal cavity per continuitatum. То circumvent such a risk, it is advocated that resection without any attempts at reduction eliminate a perceptible complication. However, when caused by benign lesions, reduction is believed to be safe as intervention for the intestinal telescoping and minimizes the incidence of associated complications of surgery [1].

One such complication associated with bowel resection is short bowel syndrome, particularly in cases requiring multiple resections such as in Peutz-Jeghers syndrome [31]. Special consideration for Peutz-Jeghers syndrome suggests that a combined approach employing both polypectomy via endoscopy and bowel resection in moderation minimize short bowel syndrome [1,31].

The surgical resection of involved bowel often requires oncological principles be employed, [3] as a large portion of cases are in relation to malignancy. Otherwise, the procedure itself is not more complicated than the conventional resection and re-anastomosis of the bowel [4] given that negative margins are achieved with regards to malignant lesions. Azar et al. [5] had described more specific approaches pertaining to the location of intussusceptions in the colon. Right-sided telescoping requires resection and primary anastomosis in even unprepared bowel, while left-sided or recto-sigmoid cases often need a Hartmann's procedure, in which reanastomosis is executed at a later date when inflammation is no longer present [1,5].

6. CONCLUSION

In conclusion, intussusception of the adult bowel is a rare occurrence that is difficult to diagnose and has a variety of causes. Given that malignancy is a prominent cause, it is important to approach this disease entity with suspicion and perform surgical resection when appropriate to avoid complications.

CONSENT

Consent was not applicable for the publication of this paper or the accompanying image.

ETHICAL APPROVAL

Ethical approval was not applicable for the publication of this paper or the accompanying image.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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