



3 Anal Mass in a 4-month-old Girl

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PRESENTATION

A 4-month-old previously healthy girl presents to the emergency department with a 1-day history of a mass protruding through the anus, bloody stools, and nonbilious emesis. She was born at term and has no prior health issues.

On physical examination, the afebrile infant has a heart rate of 117 beats per minute and blood pressure of 118/52 mm Hg. Her growth parameters are within normal ranges for age. She appears listless and mildly dehydrated, with dry mucous membranes. Her diaper is filled with a jelly-like, bloody stool, and purplish mucosa that appear similar to a cluster of small grapes protrudes from the anus. Abdominal examination reveals normoactive bowel sounds and no tenderness or masses upon palpation.

Laboratory evaluation documents normal sodium, potassium, and chloride values, with bicarbonate measuring 19 mEq/L (19 mmol/L). Complete blood cell count reveals hemoglobin of 11.3 g/dL (113 g/L), with normal white blood cell and platelet counts. An abdominal radiograph shows a nonobstructive bowel gas pattern. Given the child's ill appearance, surgical consultation is obtained, and she is hospitalized for further evaluation.

During admission, the girl develops bilious emesis. An upper gastrointestinal (GI) radiograph series is done to evaluate causes of bowel obstruction, the findings of which prompt an emergency laparotomy that confirms the final diagnosis.

DISCUSSION

An upper GI contrast study showed spiral-appearing proximal jejunal loops, with the duodenal-jejunal junction located to the right of midline, findings that are concerning for malrotation with volvulus (Figure). Emergency laparotomy confirmed malrotation with volvulus. Surgical exploration also revealed ileocolic intussusception to the level of the anus. The volvulus was untwisted, exposing viable bowel. A Ladd procedure was performed, and the intussusception was manually reduced. The patient recovered uneventfully and was healthy at a 1-month follow-up evaluation.

Differential Diagnosis

The differential diagnosis for a prolapsed rectal mass includes not only simple prolapse of rectal mucosa, commonly caused by constipation, diarrhea, malnutrition, and cystic fibrosis, but also prolapse of polyps, hemorrhoids, or intussusception. The

Figure. Upper gastrointestinal tract radiograph series image for the girl in the vignette.



appearance of prolapsed intussusception varies but can be differentiated from prolapsed rectal mucosa. With intussusception, the tissue of the protruding mass can be separated from the anal wall, which cannot be accomplished with prolapsed rectal mucosa. Rectal mucosal prolapse typically has the shape of a beehive, which contrasts with the cluster-of-grapes appearance of the prolapsed mass in this case. The appearance in this case is likely explained by venous congestion and bowel edema.

Because simple rectal prolapse failed to explain this child's listlessness, the index of suspicion was high for a more sinister diagnosis. Even in the absence of a clear cause, admission for monitoring was warranted for this ill-appearing infant. The onset of bilious emesis alerted clinicians to an underlying bowel obstruction and the need for additional testing. In this case, an upper GI radiographic series made the diagnosis of midgut volvulus, and the additional diagnosis of intussusception was discovered during surgery.

The Condition

Intussusception may involve the intestine from the jejunum to the colon. The ileocolic type is most frequent in children and is defined as a telescoping of proximal bowel into distal bowel. Invagination of proximal into distal bowel interferes with venous drainage, leading to edema of the bowel wall.

Bloody stools result from mucosal bleeding, and intestinal necrosis may develop in severe untreated cases.

Intussusception is a common cause of bowel obstruction in infants and most cases are idiopathic. The classic triad of intussusception is **colicky abdominal pain, a palpable abdominal mass, and bloody currant jelly stools**. However, the complete triad is present in fewer than 50% of cases. In infants, lethargy is a common but nonspecific finding.

This patient had a rare presentation in which the intussusceptum prolapsed through the anus and mimicked simple rectal prolapse. This presentation occurs more often in developing nations. Intussusception masquerading as rectal prolapse is remarkable on its own, but the simultaneous midgut volvulus makes this case all the more exceptional.

Volvulus occurs when a malrotated midgut twists on a narrow mesenteric base, inhibiting circulation from the superior mesenteric artery. Vomiting, possibly bilious, often results from bowel edema caused by ongoing vascular compromise. If left untreated, bowel ischemia progresses to infarction with necrosis and catastrophic loss of viable intestine.

The combination of malrotation and intussusception has been described in small case series and is known as Waugh syndrome. Abnormal fixation of the mesentery is implicated as a predisposing factor for intussusception in these cases. The incidence of this association is not well established and not

every case presents with both volvulus and intussusception. However, given the known association, clinicians should be alert to the possibility of simultaneous presentations.

Diagnosis

Intussusception is reliably diagnosed by ultrasonography. Plain abdominal radiographs may show an obstructive pattern but are not sensitive enough to reliably rule out intussusception in cases of high clinical suspicion. Air, saline, or water-soluble contrast enema may be both diagnostic and therapeutic.

An upper GI contrast study is the gold standard for diagnosing malrotation and volvulus. Ultrasonography can demonstrate suggestive findings but is not as sensitive. Plain radiographs are of limited value, although they may be useful to evaluate peritoneal free air resulting from intestinal perforation. Exploratory laparotomy or laparoscopy can be helpful in select cases of high clinical suspicion with equivocal imaging results.

Management

Volvulus requires emergent laparotomy. While preparing for surgery, the child's hemodynamic status should be optimized, antibiotics administered, and the intestines decompressed. In the operating room, the volvulus is untwisted and the Ladd procedure is performed to correct the underlying malrotation and remove any nonviable bowel.

Intussusception can be reduced with air, saline, or water-soluble contrast enema under fluoroscopy or ultrasonography. Nonoperative reduction is contraindicated in cases that present late and have signs of peritoneal irritation or circulatory compromise, and a pediatric surgeon should be consulted emergently.

In this case, ultrasonography may have led to the diagnosis of intussusception with or without the additional diagnosis of volvulus. Had nonoperative reduction of intussusception been undertaken without knowledge of the volvulus, persistent symptoms would be expected. Persistent symptoms should alert the clinician not only to the possibility of recurrent intussusception but also simultaneous volvulus, especially given the association of malrotation and intussusception.

Lessons for the Clinician

- Intussusception rarely presents as rectal prolapse if the intussusceptum extrudes through the anus.
- Presentation of a child who has rectal prolapse and appears ill should alert the clinician to the possibility of intussusception.
- Intussusception can be associated with intestinal malrotation; intussusception and volvulus may present simultaneously.

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