A 5-MONTH-OLD male infant had presented with chronic nonbilious emesis at age 3 weeks and was briefly hospitalized owing to dehydration from emesis and constipation for 4 days, the latter relieved by barium enema. An abdominal x-ray film showed distended bowel loops. Two months later, emesis recurred with diarrhea and a low-grade fever. A stool sample was positive for rotavirus. Soy-based formula was administered. At 3 months of age, symptoms recurred with abdominal pain. Distended bowel loops were noted on abdominal x-ray film. He responded to volume resuscitation and broad spectrum antibiotics. At 5½ months of age, he had recurrent dehydration and foul-smelling stools. His white blood cell count was 50×10^9/L with a shift to the left.

A repeated barium enema showed an “apple core” deformity (Figure 1). An extrinsically obstructed segment was resected from the splenic flexure. The specimen was 3.5 cm in diameter and 7.0 cm long. Mucosal hyperemia of the dilated segment and polypoid hyperplasia near the obstructed region was seen. The opened bowel and histologic sections are shown in Figures 2, 3, and 4. The postoperative course was uneventful, and he was discharged 7 days postoperatively. He was asymptomatic at follow-up 6 months later.

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Diagnosis and Discussion

**Solitary Intestinal Fibromatosis**

Figure 1. Barium enema. The splenic flexure has an “apple core” deformity indicating extrinsic obstruction.

Figure 2. A longitudinal section through the stricture. Note the heaped up, pale mucosa and submucosa and a serosal nodule.

Figure 3. Panoramic view of the strictured area with submucosal collagenous transformation with fibromatous cellularity. Note the pale nodules in the serosa (Masson trichrome, original magnification x25).

Figure 4. A collection of serosal fibrous nodules. One has increased cellularity (hematoxylin-eosin, original magnification x100).

The list of diseases causing intestinal obstruction in neonates and requiring surgical intervention is long and includes incarcerated inguinal hernia, pyloric stenosis, necrotizing enterocolitis, trauma, neoplasm, meconium ileus (eg, cystic fibrosis), meconium blockage syndrome (non-cystic fibrosis), and various other anomalies (intestinal atresia and/or stenosis, malrotation with midgut volvulus, Hirschsprung disease, and duplication cysts)\(^1\). These conditions were among those considered in the clinical differential diagnosis of this infant. Confounders include the limited x-ray film assessment early on and the delayed diagnosis owing to rotavirus infection. The resected specimen represents solitary intestinal fibromatosis (SIF), a rare cause of intestinal obstruction in the neonatal and early infantile periods.\(^2\) In contrast to the disseminated variety, SIF often presents in the neonatal or early infantile period, with low recurrence and an excellent prognosis. It can involve either the small or large intestine.

The pathological differential diagnoses include leiomyosarcoma, spindle cell sarcoma, leiomyoma, malignant mesenchymoma, cellular mesenchymal tumor, and fibrosarcoma.\(^3\) The benign appearance of the lesion and its solitary confinement to a segment of bowel earned SIF its name. Well-described pathological features include discrete thickening of the bowel wall with serosal nodules, submucosal involvement, inflammatory infiltrate (vs inflammatory fibroid polyp), heterogeneous cytological components with spindle cells, and small round cells. A monophenotypic, vimentin-positive fibroblastlike cell population was seen in our patient, possibly indicating a low-grade fibrosarcoma. However, the low proliferation index and the paucity of mitotic figures argue against this possibility. The presence of focal lymphoplasmacytic inflammation is reminiscent of an inflammatory myofibroblastic tumor (IMT), a condition that can occur in infants as young as 3 months.\(^4\) This lesion invariably shows myofibroblastic markers including cytoplasmic actin. The potential for recurrence is low but is well described in IMT. However, no IMT cases have featured areas of small round blue cells, a distinctive feature of congenital SIF. Overlapping histological features such as a focal inflammatory infiltrate mixed with spindle cells may make the differential diagnosis difficult. Conversely, SIF has not entered into the differential diagnosis of IMT. It is possible, however, that some cases may represent a transition between these conditions. Both conditions are associated with an excellent response to simple excision. Regression following biopsy has been described in IMT, while none of the reported cases of SIF have escaped surgical intervention, which consists of segmental resection and end-to-end anastomosis. To our knowledge, this represents the 10th recorded case of SIF.\(^2,3,5-8\)

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