

# The role of surgery in the treatment of intussusception in celiac disease

## Çöliyak hastalığında gelişen invajinasyonun tedavisinde cerrahinin yeri

Ayşenur CERRAH CELAYİR<sup>1</sup>, Sinan CELAYİR<sup>2</sup>, Tufan KUTLU<sup>3</sup>

Zeynep Kamil Maternal and Child Hospital, Department of Pediatric Surgery<sup>1</sup>, Cerrahpaşa Medical Faculty, Departments of Pediatric Surgery<sup>2</sup> and Pediatric Gastroenterology<sup>3</sup>, Istanbul

*Small bowel intussusceptions comprise fewer than 10% of all pediatric patients with intussusceptions and most of them are secondary to another pathology. In this report, we discuss the role of surgery in the treatment of intussusception in celiac disease. A 13-month-old girl was admitted with a three day history of progressive abdominal distention and vomiting of bile. There were air-fluid levels on supine abdominal X-ray and ultrasonographic examination demonstrated an intussusception. At surgery, two separate small bowel intussusceptions were encountered. The postoperative course was uneventful. Due to a history of frequent diarrhea and mild abdominal distention developing after the age of seven months, further studies for celiac disease were initiated. Antigliadin and antiendomysium antibodies were found to be strongly positive. Celiac disease was also confirmed by endoscopic small bowel biopsy. Children who present with chronic or transient intestinal obstruction should also be evaluated for underlying celiac disease. Nevertheless, the surgical decision should be based upon clinical observation in this group of patients.*

**Key words:** Small bowel, intussusception, celiac disease.

## INTRODUCTION

Nonobstructive small bowel intussusception (SBI) was found in up to 18% of cases of celiac disease with contrast radiology in the 1960's and 1970's (1,2). However obstructive SBI in celiac disease is uncommon and there are only a few reported cases of children with this pathology (2-4). Although intestinal obstruction is a very rare presentation of celiac disease, rare clinical forms and some underestimated complications, sometimes those requiring emergency surgical treatment, may cause diagnostic difficulties (5-7). If the association of this complication with celiac disease remains undiagnosed, life-threatening conditions may occur (2). We describe a 13-month-old girl

*Çocukluk çağındaki invajinasyonların %10'undan daha azı ince barsak invajinasyonudur ve çoğunlukla başka bir patolojiye ikincil olarak gelişirler. Bu yazıda Çöliyak hastalığında gelişen invajinasyonun tedavisinde cerrahinin yeri tartışıldı. 13 aylık bir kız çocuk, üç gündür giderek artan karın şişkinliği ve safralı kusma yakınması ile başvurdurdu. Ayakta direkt karın grafisinde geniş hava sıvı seviyeleri mevcuttu. Ultrasonografik incelemede invajinasyon gösterildi. Ameliyatta, iki ayrı yerde ince barsak invajinasyonu saptandı ve dezinvajinasyon yapıldı. Ameliyat sonrası seyri problemsizdi. Öyküsünde 7 aylıktan itibaren sıklıkla karında şişkinlik ve ishal yakınması olması nedeniyle, ileri tetkiklere başlandı. Antigliadin ve antiendomysium antikorları kuvvetli pozitif bulundu. Endoskopik ince barsak biyopsisinde çöliyak hastalık teyid edildi. Kronik veya geçici barsak tıkanıklığı gelişen çocuklarda çöliyak hastalık olasılığı da düşünülmelidir. Bu tür olgularda cerrahi tedavi kararı klinik izleme göre verilmelidir.*

**Anahtar kelimeler:** İnce barsak, invajinasyon, çöliyak hastalık.

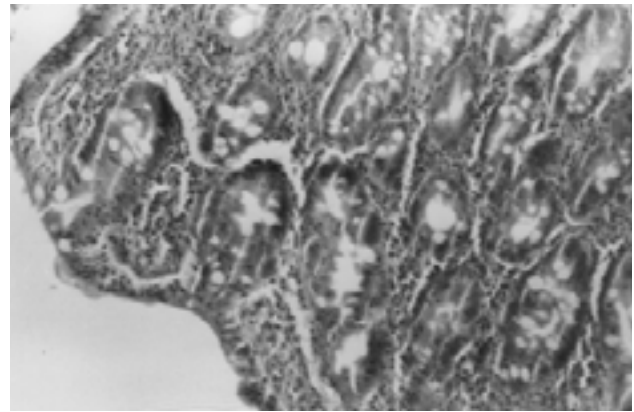
with complicated obstructive intussusception needing surgical correction and with underlying celiac disease.

## CASE REPORT

A 13-month-old girl was admitted with abdominal distension and bilious vomiting and diarrhea. There had been failure to thrive and intermittent abdominal distention during the previous months. She also had a four-week history of passing loose stools. On physical examination, there was dehydration and severe abdominal distention, with apparent episodes of colicky abdominal pain. Her plain abdominal X-rays showed air-fluid levels



**Figure 1.** Typical ultrasonographic appearance of an ileocolic intussusception demonstrating the target sign.



**Figure 2.** Total villous atrophy and crypt hyperplasia with increased intraepithelial lymphocytes (HE x 200)

with dilated intestinal loops, and on ultrasound (US) examination, signs of intussusception was seen. No stools were passed after administration of a rectal enema and her abdominal distention increased progressively, even after continuous nasogastric decompression. The follow-up serial US examination demonstrated persistence of the bowel intussusception (Figure 1). Her symptoms progressively worsened and signs of intestinal obstruction became more evident.

Thirty six hours after admission, the patient underwent emergency laparotomy. Two ileo-ileal intussusceptions were found and manual reduction was performed. There was no abdominal pathology other than the mesenteric lymphadenopathies. Postoperative oral feeding was commenced on the fourth day and abdominal distention resolved during this period. Due to the patient's history, further investigations for celiac disease were performed, and antigliadin and antiendomysium antibodies were found to be strongly positive. She was referred to a pediatric gastroenterology unit, where celiac disease was confirmed by endoscopic small bowel biopsy, which showed total villous atrophy and crypt hyperplasia with increased intraepithelial lymphocytes (Figure 2). A gluten free diet was recommended, following which she made a rapid recovery and had no further symptoms to suggest intussusception. She is currently well after three years of follow-up.

## DISCUSSION

In the past, there were very few reports of

children with celiac disease and intussusception (2-4). A SBI may complicate various conditions that disturb the small bowel mucosal structure or small bowel motility (8,9). Mucosal inflammation and disturbances of gut motility are important in the pathogenesis of intussusception complicating celiac disease (2-4). Mushtag suggested that SBI in a child with celiac disease should initially be managed medically rather than by early surgical reduction (2). In this study, the intussusceptions resolved spontaneously with medical therapy in all of the patients with celiac disease. The condition was either an isolated SBI resolving within hours or after several days, or transient, multiple SBI occurring over several days (2). These reports in the literature suggest that conservative medical treatment should be attempted first in cases with a history of celiac disease (2-4).

We treated our patient medically, but the clinical symptoms did not resolve; on the contrary, there were signs of increased intestinal obstruction during this period.

It is therefore suggested that pediatric surgeons should consider these critical questions: how should the pediatric surgeon diagnose and handle intussusception in a child with celiac disease, when should the surgeon operate on a child for intussusception when he/she was unaware of the patient's celiac disease as in this present case? In a context of celiac disease should one still wait if obstruction is complete and the clinical condition deteriorates?

Although reports advise that SBI resolves spontaneously in all patients with celiac disease, we

believe that these should not be critical in surgical decision-making. The management of intussusception should be based mainly on clinical observation and progression of the obstruction. The decision to continue medical treatment must be undertaken very cautiously because of the potentially fatal consequences of strangulation. In this child, prompt operative reduction was required due to complete intestinal obstruction and clinical deterioration. We consider that in cases with undiagnosed celiac disease, surgical decision mak-

ing regarding intussusception should be based on clinical observation, as in the present study.

In conclusion, children who present with chronic or transient intestinal obstruction due to intussusceptions should be evaluated for underlying disease such as celiac disease. However, a decision on surgical treatment of intussusception should be based on clinical observation and radiological investigation whether celiac disease is present or not.

## REFERENCES

1. Burrows FGO, Toye DKM. Barium studies. *Clinics in Gastroenterol* 1974; 3: 91-107.
2. Mushtaq N, Marven S, Walker J, et al. Small bowel intussusception in celiac disease. *J Pediatr Surg* 1999; 34:1833-5.
3. Germann R, Kuch M, Prinz K, et al. Celiac disease: an uncommon cause of recurrent intussusception. *J Pediatr Gastroenterol Nutr* 1997; 25: 415-6.
4. Gonzalez JA, Gonzalez JB, Crespo MJ, Sancho CI. Acute gallbladder distention and recurrent small bowel intussusception in a child with celiac disease. *J Pediatr Gastroenterol Nutr* 1998; 27: 444-5.
5. Cohen MD, Lintott DJ. Transient small bowel intussusception in adult coeliac disease. *Clin Radiol* 1978; 29: 529-34.
6. Masterson JB, Sweeney EC, Path MC. The role of small bowel follow-through examination in the diagnosis of coeliac disease. *Br J Radiol* 1976; 9: 660-4.
7. Riccabona M, Rossipal E. Sonographic findings in celiac disease. *J Pediatr Gastroenterol Nutr* 1993; 17:198-200.
8. Stringer MD, Pablot SM, Brereton RJ. Pediatric intussusception. *Br J Surg* 1992; 79: 867-76.
9. Cucchiara S, Bassotti G, Castellucci G, et al. Upper gastrointestinal motor abnormalities in children with active celiac disease. *J Pediatr Gastroenterol Nutr* 1995; 21:435-42.