

Case Report / Olgu Sunusu

**Inflammatory Myofibroblastic Tumor Leading to Intestinal
Obstruction in an Infant**

**İnfanтта İnflamatuar Myofibroblastik Tumor Nedeniyle Gelişen
İntestinal Obstrüksiyon**

Bethany J. Slater¹, Andrea K. Sotelo¹

¹ Rocky Mountain Hospital
for Children, Denver,
Colorado, USA

ÖZET

İnfanlarda çoğunlukla konjenital sebeplerle olmak üzere bir kısım sebeplerden dolayı ince barsak obstrüksiyonu gelişir. İnflamatuar myofibroblastik tumor (IMT) çocuklarda çok nadir görülen barsak tıkanıklarından biridir. IMT pediatrik popülasyonda bulunan nadir ve benign solid tümörlerdir. Bu makalede laparoskopik reseksiyon ve barsak anastomozu yapılan jejunal IMT ile barsak tıkanıklığı olan 4 aylık kız hasta sunulmuştur.

Anahtar Kelimeler: infant, inflammatory myofibroblastik tumor, pediatrik, ince barsak obstrüksiyonu

ABSTRACT

There are a number of causes of small bowel obstruction in infants, most commonly from congenital causes. Inflammatory myofibroblastic tumor (IMT) of the alimentary tract is a very unusual cause of intestinal obstruction in children. IMT is a rare, benign solid tumor that presents mostly in the pediatric population. We present a case of a jejunal IMT causing bowel obstruction in a four-month-old female treated with laparoscopic resection and bowel anastomosis.

Keywords: infant, inflammatory myofibroblastic tumor, pediatrics, small bowel obstruction

Corresponding Author:

Bethany Slater
Rocky Mountain Hospital for
Children, Denver, Colorado,
USA

Telefon : 303-839-6001

Faks : 303-839-6033

Email:

bjslater1@gmail.com

INTRODUCTION

There are a number of causes of small bowel obstruction in infants, most commonly from congenital causes. These include hernias, intussusception, volvulus, as well as intramural and extramural masses. Inflammatory myofibroblastic tumor (IMT) of the alimentary tract is a very unusual cause of intestinal obstruction in children. IMT is a rare, benign solid tumor that presents mostly in children. We present a case of a jejunal IMT causing bowel obstruction in a four-month old female

CASE

A four-month female was referred who had difficulty with feeding after birth that had gotten progressively worse. At 3 weeks of age, she developed emesis and irritability with feeds. At that time she was worked up with an ultrasound and upper GI which reportedly revealed reflux and a possible duodenal stricture. She was put on antireflux meds with no significant improvement. The mother also maintained a restrictive elimination diet to avoid any allergens in her breast milk. The patient continued to have intermittent emesis, weight loss, and irritability. She was thus sent to Pediatric Gastroenterology who directly admitted her to the hospital for workup and feeding tube placement. The physical exam was unremarkable, and she had no other medical or surgical history. An upper contrast study was performed demonstrating a very dilated duodenum and proximal jejunum with a transition to decompressed small bowel suggesting a focal obstructing lesion in the proximal jejunum. (Figure 1) The patient was taken to the operating room the next day and a small bowel mass was identified (Figure 2). A laparoscopic resection with intracorporeal stapled side to side, functional end to end anastomosis was performed with an endoscopic 5-mm stapler (Just Right Surgical, Boulder, CO). (Figure 3A, B) The resultant enterotomy was closed with a running suture.

The patient had an unremarkable hospital course and was discharged on POD # 4 tolerating full feeds. The pathology showed an inflammatory myofibroblastic tumor with negative margins. (Figure 4)



Figure 1 – Upper GI image demonstrating transition zone with dilated proximal bowel and decompressed distal bowel



Figure 2 – Gross picture of jejunal mass

At two-week follow-up, she was doing well with good oral intake and normal stools. Approximately one month post-operatively, she underwent an MRI of the abdomen and pelvis and CT scan of the chest with no residual or metastatic disease. In addition, inflammatory markers were obtained which were normal. Another MRI was obtained at six months post-operatively with no recurrent mass or dilation of the bowel. At that time, she was eating well, stooling, and gaining appropriate weight. She will be followed up with MRIs every 6 months for 2 years.

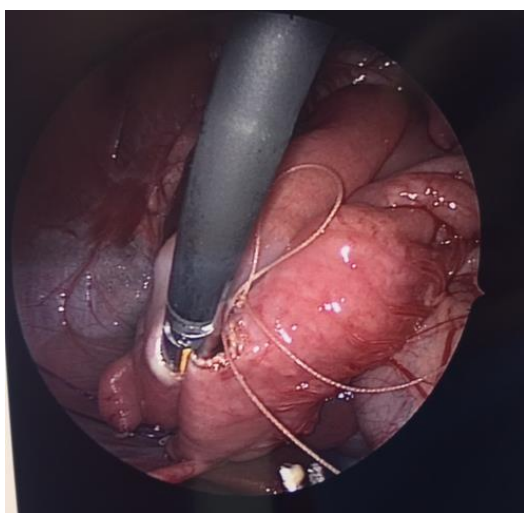


Figure 3A – Intra-operative picture demonstrating anastomosis with 5mm stapler

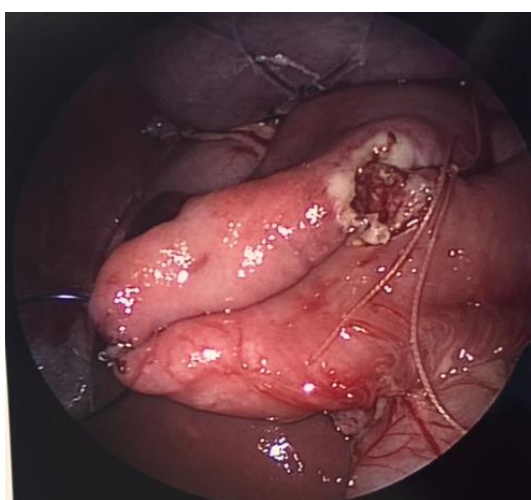


Figure 3B – Intraoperative Picture demonstrating stapled anastomosis

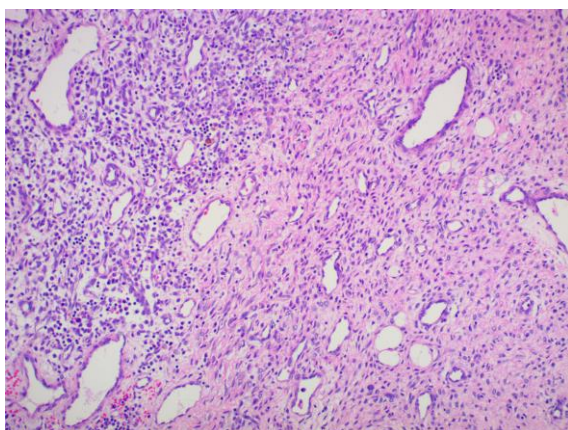


Figure 4 – Histology of IMT demonstrating an infiltrate of lymphocytes and plasma cells

DISCUSSION

Inflammatory myofibroblastic tumor (IMT) is a rare, solid tumor of unknown etiology seen mostly in children and young adults. It has been reported in a variety of sites including lung, which is most common, abdomen and pelvis, and head and neck.(1) The clinical presentation depends on the anatomic site from where the tumor originates. They may be asymptomatic and found incidentally on imaging or cause symptoms from mass effect, such as abdominal pain or respiratory distress, or from an inflammatory response, such as fever, weight loss and fatigue. Some patients can have laboratory abnormalities, such as microcytic anemia, thrombocytosis, and elevated white blood cell count. Rarely bowel obstruction can result when tumors arise from the mesentery or small intestine, as in this case. There have been a few other cases of obstruction from these masses reported, but IMT of the alimentary tract in children is rare.(2, 3)

Although most IMTs have a benign course with survival rates greater than 90%(1), some have invasive or malignant features, and there have been reports of malignant transformation.(4) Due to the potential invasive nature of this lesion, they may be confused for sarcomas or lymphomas pre-operatively. Cross sectional imaging, such as CT scan or MRI, may aid with diagnosis and operative planning, however diagnosis is typically made only with pathology. Histological diagnosis is made by the presence of variably cellular, bland, spindled, fibroblastic-myofibroblastic cells with an infiltrate of lymphocytes, plasma cells and occasional eosinophils.

Immunohistochemistry for ALK-1 is present in approximately 50-60% of cases.

Complete surgical resection is the treatment for IMT. In this case, complete resection of the mass was performed with a laparoscopic bowel resection and intracorporeal anastomosis. Local recurrence is fairly high with incidences reported to be between 15-40% in large series of children.(5, 6) As such, close follow-up is required to identify recurrences with physical exam and imaging studies. For cases of unresectable or recurrent lesions, adjuvant treatment with steroids or chemotherapy has been used with variable success.(7-9) Additionally, NSAIDs may be used to treat or shrink these tumors.(10) It is hypothesized that NSAIDs may interfere with vascular endothelial growth factor signaling by cyclooxygenase 2 inhibition.

REFERENCES

1. Dalton BG, Thomas PG, Sharp NE, Manalang MA, Fisher JE, Moir CR, et al. Inflammatory myofibroblastic tumors in children. *J Pediatr Surg.* 2016 Apr;51(4):541-4
2. Ciftci AO, Akcoren Z, Tanyel FC, Senocak ME, Caglar M, Hicsonmez A. Inflammatory pseudotumor causing intestinal obstruction: diagnostic and therapeutic aspects. *J Pediatr Surg.* 1998; 33(12): 1843-5.
3. Sanders BM, West KW, Gingalewski C, Engum S, Davis M, Grosfeld JL. Inflammatory pseudotumor of the alimentary tract: clinical and surgical experience. *J Pediatr Surg.* 2001;36(1):169-73.
4. Zavaglia C, Barberis M, Gelosa F, Cimino G, Minola E, Mondazzi L, et al. Inflammatory pseudotumor of the liver with malignant transformation. Report of two cases. *Ital J Gastroenterol.* 1996;28(3):152-9.
5. Karnak I, Senocak ME, Ciftci AO, Caglar M, Bingol-Kologlu M, Tanyel FC, et al. Inflammatory myofibroblastic tumor in children: diagnosis and treatment. *J Pediatr Surg.* 2001;36(6):908-12.
6. Souid AK, Ziemba MC, Dubansky AS, Mazur M, Oliphant M, Thomas FD, et al. Inflammatory myofibroblastic tumor in children. *Cancer.* 1993;72(6):2042-8.
7. Bertocchini A, Lo Zupone C, Callea F, Gennari F, Serra A, Monti L, et al. Unresectable multifocal omental and peritoneal inflammatory myofibroblastic tumor in a child: revisiting the role of adjuvant therapy. *J Pediatr Surg.* 2011;46(4):e17-21.
8. Dagash H, Koh C, Cohen M, Sprigg A, Walker J. Inflammatory myofibroblastic tumor of the pancreas: a case report of 2 pediatric cases--steroids or surgery? *J Pediatr Surg.* 2009;44(9):1839-41.
9. Williams ME, Longmaid HE, Trey G, Federman M, Crosson AW. Renal failure resulting from infiltration by inflammatory myofibroblastic tumor responsive to corticosteroid therapy. *Am J Kidney Dis* 1998;31(6):E5.
10. Applebaum H, Kieran MW, Cripe TP, Coffin CM, Collins MH, Kaipainen A, et al. The rationale for nonsteroidal anti-inflammatory drug therapy for inflammatory myofibroblastic tumors: a Children's Oncology Group study. *J Pediatr Surg.* 2005;40(6):999-1003