Journal of Pediatric Surgery (2013) **48**, 677–680

ELSEVIER

Journal of Pediatric Surgery

www.elsevier.com/locate/jpedsurg

Obstructing apple core lesion of the rectum: A case report of inflammatory pseudotumor masquerading as colorectal carcinoma

Shevonne S. Satahoo[a,*], Carole Brathwaite[b], James S. Davis[a], Cathy Burnweit[c,d]

[a]*Department of Surgery, University of Miami Miller School of Medicine, Miami, FL 33136*
[b]*Department of Pathology, Miami Children's Hospital, Miami, FL*
[c]*Department of Pediatric Surgery, Miami Children's Hospital, Miami, FL*
[d]*Department of Surgery, Herbert Wertheim College of Medicine, Florida International University, Miami, FL*

Received 5 November 2012; revised 2 January 2013; accepted 9 January 2013

Key words:
Inflammatory pseudotumor;
Apple core lesion;
Rectum;
Pediatric;
NSAIDs

Abstract Inflammatory pseudotumor is a rare lesion which can occur, typically in children and young adults, in many different organ systems. The tumor often clinically behaves like a cancer but without histological evidence of malignancy. This case study of a 14 year-old boy is the first report in the literature of an inflammatory pseudotumor presenting as an obstructing apple core lesion, mimicking a rectal carcinoma. A six-week course of non-steroidal, anti-inflammatory drugs (NSAIDs) led to complete resolution of the mass, and following resection of a residual stricture, the patient has been recurrence-free for seven years.

Inflammatory pseudotumor is a rare lesion, often occurring in the first two decades of life [1]. While it is most commonly seen in the lungs, it has been reported in almost every organ of the body with specific symptoms in each case relating to the site of origin [1–9]. Previously considered benign, inflammatory pseudotumor has been reclassified as an "intermediate neoplasm" by the World Health Organization [1,8]. This designation is supported by the tumor's capacity to infiltrate surrounding tissue, metastasize, and recur locally [7,10,11]. Diagnosis is often challenging and delayed because these tumors often resemble a malignancy without confirmatory microscopic or serological evidence. Management usually involves resection, but a correct pre-operative diagnosis may avert an overly aggressive resection or oncologic therapy as well as their associated morbidities.

This study presents the first report of inflammatory pseudotumor presenting as an apple-core rectal lesion. Found in a teen-aged boy, the lesion masqueraded as an obstructing adult-type colorectal carcinoma. This case further illustrates how non-steroidal, anti-inflammatory drugs (NSAIDs) effectively treated the tumor once the proper diagnosis was made.

1. Case presentation

A 14 year-old male, with no past medical or surgical history, presented to a clinic in the Bahamas with a 3-week

* Corresponding author. Tel.: +1 305 585 1280.
E-mail address: sssatahoo@med.miami.edu (S.S. Satahoo).

0022-3468/$ – see front matter © 2013 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.jpedsurg.2013.01.034

history of tenesmus, bloody stools, obstipation and a 5 kg weight loss. He denied trauma, anal instrumentation, or history of chronic constipation. A barium enema revealed an apple core lesion, 6 cm in length, consistent with rectal carcinoma (Fig. 1). The patient was referred to our institution for further management.

On examination, the boy was well nourished, with a height of 173 cm (85th percentile) and a weight of 60 kg (55th percentile). His abdomen was benign with no palpable mass, tenderness or organomegaly; and his genitalia and perineum were normal. Digital rectal exam revealed a nontender, firm, fixed lesion approximately 8 cm from the anal verge, a tight luminal stricture, and Hemoccult positive stool.

Laboratory data included a white blood cell count of 12,100/mm^3, hemoglobin of 13.3 g/dL, globulin of 3.6 g/dL, C-reactive protein of 9.6 mg/dL and sedimentation rate of 58 mm/h; CEA and AFP were normal. Computerized tomography (CT) of the abdomen and pelvis showed a 6.5 cm circumferential rectal mass with near obliteration of the lumen, infiltration of surrounding fat, and significant local lymphadenopathy. Attempted colonoscopy confirmed a shaggy, white, necrotic lesion that appeared to be rectal cancer and a 4 mm luminal stenosis that precluded advancement of the scope (Fig. 2). Biopsy specimens revealed marked chronic inflammation and what was initially interpreted as fibrosis without evidence of malignancy. The patient subsequently underwent a transanal surgical biopsy. Multiple generous incisional tumor fragments showed extensive fibro-inflammatory changes with mucosal ulceration and ischemic necrosis, but no evidence of malignancy.

Despite negative biopsies, the clinical picture was still considered highly suspicious for rectal carcinoma. In an attempt to relieve the obstruction and provide a definitive tissue diagnosis, exploratory laparotomy with open biopsy and diverting loop sigmoid colostomy was performed. An expansive, hard, fixed mass engulfing the rectum at the peritoneal reflection was noted; there were multiple lobulations and focal infiltration, but contrary to the imaging findings, no lymphadenopathy could be identified in the pelvis or periaortic region. Histology revealed fat necrosis, focal acute and chronic inflammation, and no malignant cells. There was also a storiform proliferation of spindle to stellate myofibroblastic cells, alternating with fibrosis and almost keloid-like collagenous areas of scarring (Fig. 3). With the exception of vimentin, all immunohistochemical stains, including anaplastic large cell kinase (ALK), were negative; and fluorescent in situ hybridization was negative for the t(2:5) gene rearrangement. At this point, the diagnosis of inflammatory pseudotumor was cautiously suggested.

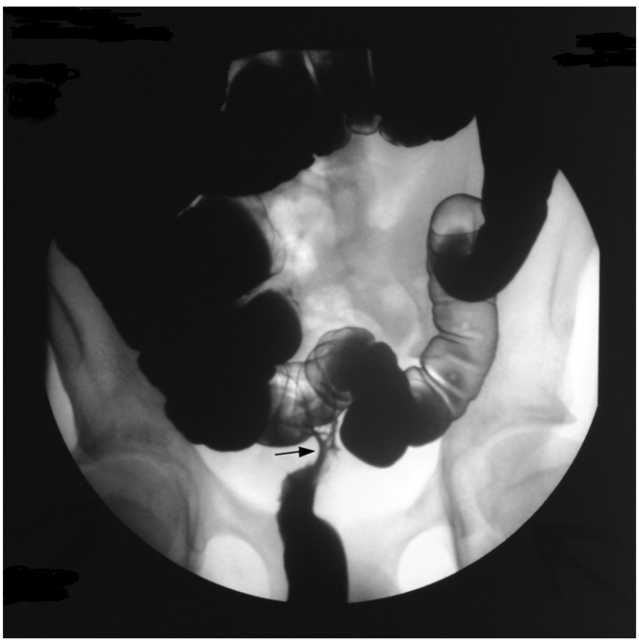

Fig. 1 Barium enema showing an apple core lesion (arrow).

The boy began a six-week course of high-dose oral NSAID therapy with Ibuprofen 600 mg four times daily. Rapid clinical improvement and weight gain ensued. Two weeks after initiation of therapy, an exam under anesthesia revealed healthy pink rectal mucosa, and a rectal stricture was dilated using a 60-French esophageal bougie. Two months thereafter, having finished his course of NSAIDs, the patient underwent a repeat CT scan demonstrating marked resolution of the rectal wall thickening and perirectal fat stranding. No lesion was palpable on rectal exam.

The patient was scheduled for an evaluation under anesthesia and colostomy closure. No residual mass was seen intraoperatively, but a localized, dense rectal stricture was found at the peritoneal reflection, above the reach of a digital exam, where the pseudotumor had been. Consequently, the patient underwent a resection of the involved short rectal segment with colostomy closure. Histology showed submucosal proliferation of spindled myofibroblastic cells in a fascicular pattern, scarring, some focal ulceration, and residual inflammation. Post-operatively, the patient remained asymptomatic; he was eating well and passing normal bowel movements. By his six month follow-up visit, he had attained his pre-illness weight. He remains without recurrence seven years after diagnosis.

2. Discussion

Inflammatory pseudotumor is a rare entity. While it occurs most often in children and young adults (mean age of 10 years), it has been documented in patients as old as 79 years [1,4]. There appears to be a slight female predilection [1,7,10,12]. Most commonly, inflammatory pseudotumor originates in the lungs [1,3,8,10–12]. Among the extrapulmonary lesions, 43% occur in the mesentery and omentum [1]. Other sites include soft tissue, mediastinum, gastrointestinal tract, pancreas, genitourinary tract, oral cavity, breast, nerve, brain and central nervous system [1–6]. Clinical symptoms depend on the lesion's location [6–9]. Up to a third of patients experience fever, growth failure,