



SHORT REPORT

The association between Crohn's disease and desmoid tumors: A novel case and review of the literature

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Abstract

Desmoids are rare tumors resulting from the proliferation of fibroblasts. They occur in association with familial adenomatous polyposis (FAP), but they may also occur in the post-traumatic peri-partum or post-abdominal surgery setting, and a few present spontaneously. Presenting features of desmoids are protean and largely relate to the anatomical area of involvement.

We describe a 50 year old male not known to have Crohn's disease and without FAP who presented with multiple desmoids. Investigation of post-operative diarrhea confirmed a diagnosis of Crohn's disease. This is the first report of a *male* patient, who had never undergone prior abdominal surgery, presenting with Crohn's disease and abdominal desmoid tumors. The reasons why Crohn's disease and desmoids may be associated are explored, focusing particularly on alternations in the fibrogenic cytokine TGF- β now known to be involved in the pathogenesis of both diseases.

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1. Introduction

Desmoid tumors are rare with an incidence rate of 2 to 4 cases per million per year.^{1,2} The peak incidence of desmoids is between the ages of 25–35 years and they are slightly more common in women.

Although desmoids can occur at any site in the body, three main anatomic sites are described: (i) trunk or extremity (ii)

abdominal wall and (iii) intra-abdominal. The characteristic feature of these tumors is a monoclonal fibroblastic proliferation arising from muscular or aponeurotic structures. Histology usually reveals a spindle-shaped pattern of cells, separated by thick collagen fibers.

Desmoid tumors are formally benign but with uncontrolled fibroblast growth, a mass can form which can ultimately compromise local tissue function and so desmoids

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can be as destructive as malignant tumors because of their tendency to produce local complications. Desmoids within the abdomen are often associated with bowel obstruction and/or infarction.

Although desmoids can occur sporadically they occur far more frequently in association with FAP.³ Aggressive desmoid fibromatosis affects 10–15% of all patients with FAP following colectomy, and these patients have an 852-fold increased risk of developing desmoid tumors compared to the general population. Adenomatous Polyposis Coli gene (APC) mutations in somatic cells have been reported to play a role in development of desmoid tumors.⁴ It is also well-known that desmoids can form at sites of previous surgical trauma.

Surgery for intra-abdominal desmoids, especially those involving the mesentery may be extremely difficult due to the often bulky nature of the tumor and/or attachments to vital organs. With surgery alone, desmoids have recurrence rates between 25% and 30%.⁵

Chemotherapy may be used in unresectable disease as an adjuvant to surgery.^{6,7} Medical therapy has produced some success with the non-steroidal anti-inflammatory (NSAID) sulindac, and the anti-estrogen, tamoxifen.^{8,9} The tyrosine kinase inhibitor imatinib has been used with some success in selected patients.¹⁰

Crohn's disease is rarely associated with epithelial and lymphoreticular tumors. A MEDLINE search from 1966 to 2009 revealed three papers, describing a total of 5 patients, in whom desmoid tumors presented in association with a diagnosis of Crohn's disease.^{11–13} One of these patients developed an *extra*-abdominal desmoid tumor after a diagnosis of Crohn's disease and in another 2 of these patients intra-abdominal desmoid tumor developed *after* intestinal surgery for Crohn's disease. In these cases, this suggests that the surgery was the triggering stimulus for the development of tumor.

In the remaining 2 of the 5 published cases, the diagnosis of Crohn's disease and abdominal desmoid tumor was made simultaneously. Both cases were young female patients on the oral contraceptive pill, and a high estrogen state was invoked as a possible trigger. Numerous studies have proposed a possible role for high estrogen levels in provoking the development of desmoid tumor.^{14–16}

We report the first case in which a hyper-estrogenemic state is absent, namely in a *male* patient who had never undergone abdominal surgery but was diagnosed with intra-abdominal desmoid tumors and Crohn's disease affecting the ileum and colon.

2. Case report

A 50-year old man presented to our institution with progressive abdominal pain associated with obstructive bowel symptoms, and a palpable abdominal mass. A CT scan demonstrated 3 discrete masses, involving both the colon and ileum; a fine needle aspirate confirmed a mesenchymal tumor, most in keeping with a desmoid tumor. In November 2001, because of the obstructive symptoms, the patient proceeded to laparotomy and resection of the tumor. A right hemi-colectomy and small bowel resection (4ft of terminal ileum) were both required.

Pathological examination of the surgical specimens confirmed desmoid tumors.

Post-operatively, the patient was troubled by non-bloody and watery diarrhea up to 10 times per day. This was initially ascribed to bile-salt malabsorption. He was treated with cholestyramine, which reduced the stool frequency to an average of 3 times daily. However, his stool consistency remained abnormal (liquid to semi-formed). He was referred to the gastroenterology department for further evaluation.

He recalled being anemic as a teenager and a diagnosis of irritable bowel syndrome had been made approximately 15 years previously. He described a lifelong history of watery stools. Investigations at another institution failed to define the cause of his anemia. Celiac disease was excluded based on serology and endoscopies undertaken elsewhere were normal.

In addition to cholestyramine 4 g daily, he was also taking celecoxib 200 mg daily and a multivitamin. The celecoxib was started in November 2001 as prophylaxis against recurrence of the tumor, an approach that has been reported by others.¹⁷

The initial impression was that of on-going bile-salt diarrhea and his cholestyramine was initially increased to 4 g in the morning and 2 g in the evening. This caused some abdominal bloating but did improve the consistency of his stools.

Subsequent investigations confirmed iron-deficiency anemia (hemoglobin 120 g/L, mean corpuscular volume 70.9 fL and ferritin 5 µg/L) with normal white blood cell and platelet counts. His serum B12, folate, and liver enzymes were normal. In June 2002, esophagogastroduodenoscopy was unremarkable, except for a small, non-bleeding antral angiodysplasia. Colonoscopy revealed multiple aphthoid erosions interspersed with normal mucosa throughout the colon and neo-terminal ileum. Biopsies showed mild chronic active ileitis and a moderate chronic active colitis with cryptitis and crypt abscess formation but no granulomata. Based on these endoscopic and histologic findings, Crohn's disease was formally diagnosed.

At the time of his clinical presentation with desmoid tumor the focus had clearly been on management of this tumor and the microscopic evidence of Crohn's disease in the surgically resected bowel was not fully realised at the time. A further careful histological analysis of the resected colon and ileum confirmed microscopic changes consistent with a diagnosis of Crohn's disease. In the small bowel there was a mild chronic inflammation of the mucosa and submucosa, as well as focal mild to moderate acute inflammation with erosions. In some areas, there was prominent glandular architectural distortion with well-formed lymphoid follicles in the mucosa and submucosa (Fig. 1). Rare foci of pyloric metaplasia, suggestive of healed ileitis were also seen (Fig. 2). The presence of these changes in the original surgical specimen is consistent with a diagnosis of Crohn's disease.

As his symptoms were mild, he was started on the oral 5-ASA Asacol (Procter & Gamble Inc., Toronto, ON) 800 mg BID. He was also started on monthly vitamin B12 injections (1000 µg) given his extensive terminal ileal resection. With the addition of 5-ASA, his bowel habit normalized.

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