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Case Report

Rectal polypoid Inflammatory Myofibroblastic Tumor in 39-year-old liver transplant recipient with de-novo ulcerative colitis: A case report and literature review

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ABSTRACT

We describe a rare, polypoid Inflammatory Myofibroblastic Tumor (IMT) and de-novo ulcerative colitis (UC) arising concurrently in the rectum of a 39-year-old liver transplant recipient. This is the third report of an IMT or IPT (Inflammatory Pseudotumor) occurring in a liver transplant recipient, the first in an adult, and only the fifth IMT/IPT to involve the rectum. It is also the first to be identified in an individual with confirmed ulcerative colitis. We discuss the potential role of Tumor Necrosis Factor alpha (TNFα) in the pathogenesis of these rare neoplasms and consider a biological association between IMTs and ulcerative colitis. We also recognize IMTs as a rare, potentially serious complication of solid-organ transplants and discuss the significance of both an autoimmune disease and an IMT developing simultaneously in the setting of potent immunosuppression.

1. Introduction

Inflammatory Myofibroblastic Tumors (IMTs) are mesenchymal proliferations of intermediate malignant potential. While it has long been debated whether these tumors represent inflammatory processes or true neoplasms, [1–3] current literature favors that they are neoplastic in nature, and specific genetic aberrations have been identified [4,5]. However, the pathogenesis of these rare lesions is just beginning to be understood. Interestingly, infliximab, a TNFα inhibitor and mainstay of therapy in the treatment of ulcerative colitis, has also displayed effectiveness against IMTs in a single case study, implicating that TNFα could play a central role in the pathogenesis of both ulcerative colitis and IMTs [6]. We describe, for the first time, an IMT arising in the context of active ulcerative colitis.

IMTs commonly occur in the lungs, abdominal and pelvic viscera, and soft tissues of children and adolescents, but they have been known to occur in any region of the body, and in any age group [6]. They are infrequent in the gastrointestinal tract, and exceedingly rare in the rectum. This report describes a polypoid submucosal IMT arising in the rectum of a 39-year-old male who received a cadaveric liver transplant for end-stage liver failure secondary to primary sclerosing cholangitis. Although not documented before nor after liver transplant, the patient was found to have de-novo pan ulcerative colitis at the time of the IMT excision.

2. Case report

A 39-year-old man with an orthotopic liver transplantation at the age of 31 for end stage liver disease secondary to Primary Sclerosing Cholangitis (PSC) with no prior endoscopic or histologic evidence of Ulcerative Colitis (UC) (colonoscopy 2 years prior to transplantation and lower endoscopy pre-transplantation were negative for colitis), presented with a 9 to 10-month history of 4–6 non-bloody bowel movements per day, associated with urgency, nocturnal bowel movements and unintentional weight loss of approximately 8 to 10 pounds. No infectious risk factors were present. Past medical history was

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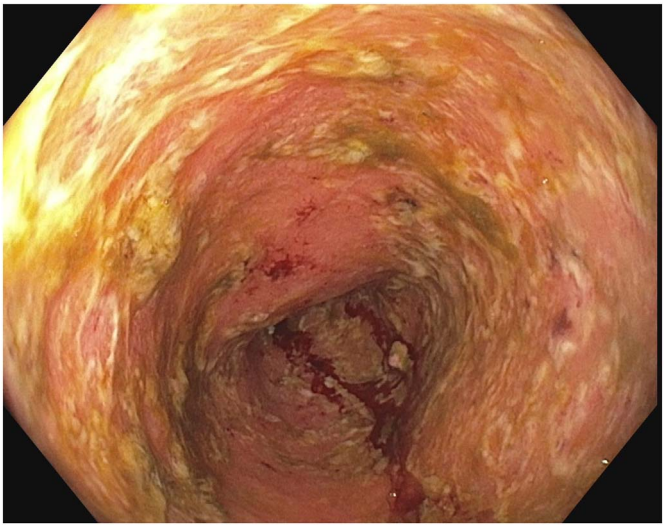

Fig. 1. Transverse colon at endoscopy showing inflammation, mucosal erythema, loss of vascular pattern, exudate, and superficial ulcerations (Mayo 3 colitis).

significant only for PSC and liver transplantation with no recurrence of PSC and adequate graft function. Medications included tacrolimus 3 mg and mycophenolate mofetil 500 mg, with no known drug allergies. Family history was non-contributory. Investigations revealed a CRP of 8.9 mg/L and negative stool cultures for *C. difficile*.

A colonoscopy was performed, which revealed pan-colonic erythema, loss of vascular pattern, mucosal friability, exudate, superficial ulcerations, and pseudopolyps (consistent with Mayo grade 3 Ulcerative Colitis) (Fig. 1). Pan-colonic biopsies were performed (Histology Figs. 2 and 3). A rectal polyp was removed with hot snare polypectomy (Fig. 4 during polypectomy).

Pathology from the biopsies revealed a pancolonic severe active chronic colitis typical of inflammatory bowel disease (IBD), in particular, chronic ulcerative colitis. There was no evidence of mycophenolate mofetil toxicity. Two of the 31 biopsies from non-polypoid mucosa showed atypical epithelium indefinite for dysplasia. Immunohistochemistry (IHC) for CMV was negative in the rectal mucosa (please see Table 1 re: IHC antibody characteristics). The large rectal polyp (2.5 by 1.5 by 1.3 cm measured in the laboratory) was within the submucosa/mucosa and no deeper with no muscularis propria in the specimen. Pathologically there was a thin rim of acutely

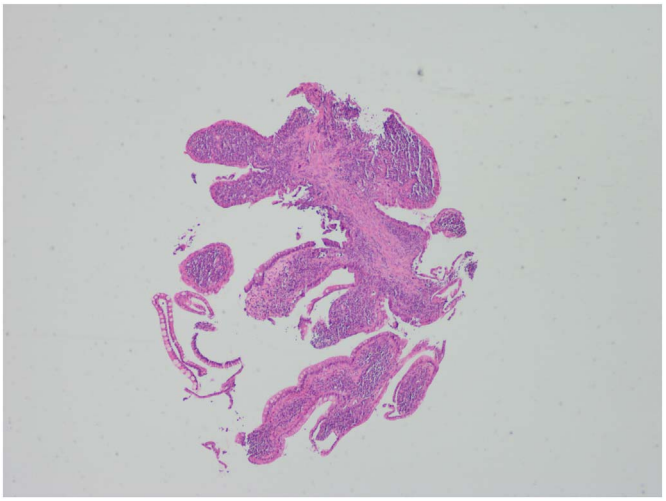

Fig. 2. Haematoxylin and Eosin ×40. Representative histologic section of pan-colitis (transverse colon) to show significant architectural distortion and dense lymphoplasmacytic lamina proprial infiltrate.

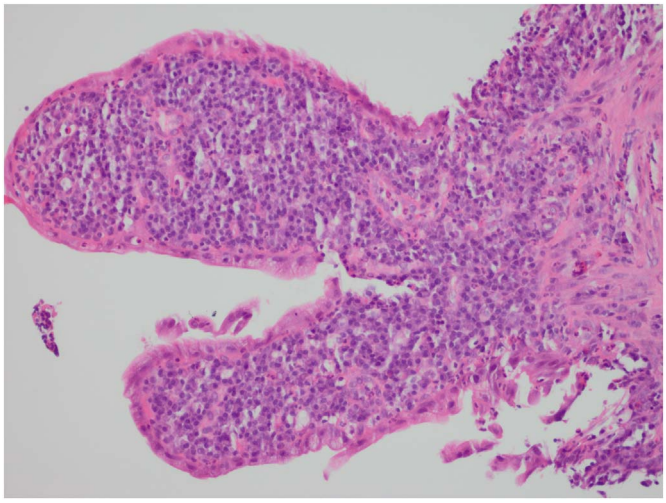

Fig. 3. Haematoxylin and Eosin ×200. Transverse colon severe active chronic colitis with surface epithelial attenuation/repair.

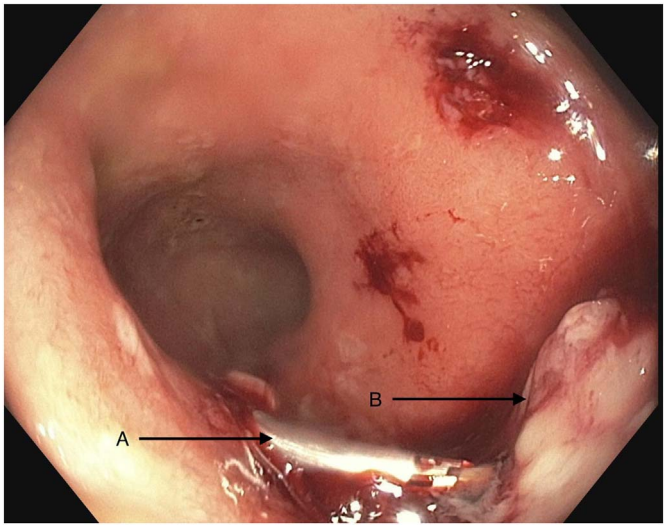

Fig. 4. Rectum: Photograph taken during polypectomy, prior to complete snare polypectomy. Residual polyp at B. Hemoclip at A.

inflamed granulation tissue surrounding a dominant homogenous core of a densely cellular large spindle cell proliferation arrayed in a storiforming architecture (non-fascicular) (Figs. 5 and 6). Throughout there was a light admixture of lymphocytes and plasma cells and occasionally macrophages. The nuclei were open with generally small nucleoli with mild nuclear membrane irregularity with an occasional larger pleomorphic nucleus (Fig. 7). Mitotic figures were few and the Ki67 proliferation rate relatively low at 10% (Fig. 8). The eosinophilic cytoplasm was ill-defined, without cross nor longitudinal striations. The cells were negative by immunohistochemistry for CD117 (gastrointestinal stromal tumor); negative for HMB45 (Perivascular epithelioid cell tumor); negative for CD34 (solitary fibrous tumor); negative for myogenin (skeletal muscle) but moderately positive for desmin (Fig. 9) with weaker but definite positivity for smooth muscle actin (Fig. 10) and caldesmon. The tumor was strongly and diffusely positive for vimentin (Fig. 11). A stain for cytokeratin was negative (pan keratin cocktail). Stains for p53 and bcl2 were negative. A stain for ALK1 per immunohistochemistry showed no overexpression (Fig. 12) and follow-up Florescent in situ hybridization (FISH) for the ALK1 translocation was negative (see Note 1); however, a clonal population that was identified in 50% of the nuclei on FISH showed a loss of one signal indicative of either monosomy of chromosome 2 or a deletion of the