

THE DREADED DESMOID: MANAGING DESMOID DISEASE IN PATIENTS DESIRING AN ILEAL POUCH IN FAMILIAL ADENOMATOUS POLYPOSIS

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Objectives

- FAP (familial adenosis polyposis) is a rare autosomal dominant inherited mutation on APC gene associated with numerous colorectal adenomatous polyps and significant risk of colorectal cancer.
- As such, prophylactic surgery is a mainstay of management; however, due to the rare nature of the disease, extracolonic manifestations of disease, and patient-specific factors, surgical decision making can often be very complex and specialized.
- This decision process often hinges at the length of small bowel after removal of colon, allowing for anastomosis to anus via ileal pouch (ileal pouch-anal anastomosis or IPAA) or rectum (ileorectal anastomosis or IR).
- In this case report, we discuss the role of desmoid disease in limiting small bowel length and the decision to perform IR anastomosis to maximize quality of life.

Case Background

We describe a 41 year old male FAP who presents for ileoanal anastomosis (IPAA) as part of a three staged procedure: colectomy and end-ileostomy, IPAA with formation of J-pouch, and reversal of loop ileostomy. With regards to pre-operative planning, prehabilitation with weight loss was pursued to improve operative success. Due to concerns for indeterminate lesions in small bowel mesentery, a plan to biopsy and attempt to perform ileal reservoir procedure was discussed pending operative findings, as patient's quality of life was significantly limited by ileostomy.



Image 1. Pre-operative CT scan indicating mesenteric mass concerning for desmoid disease

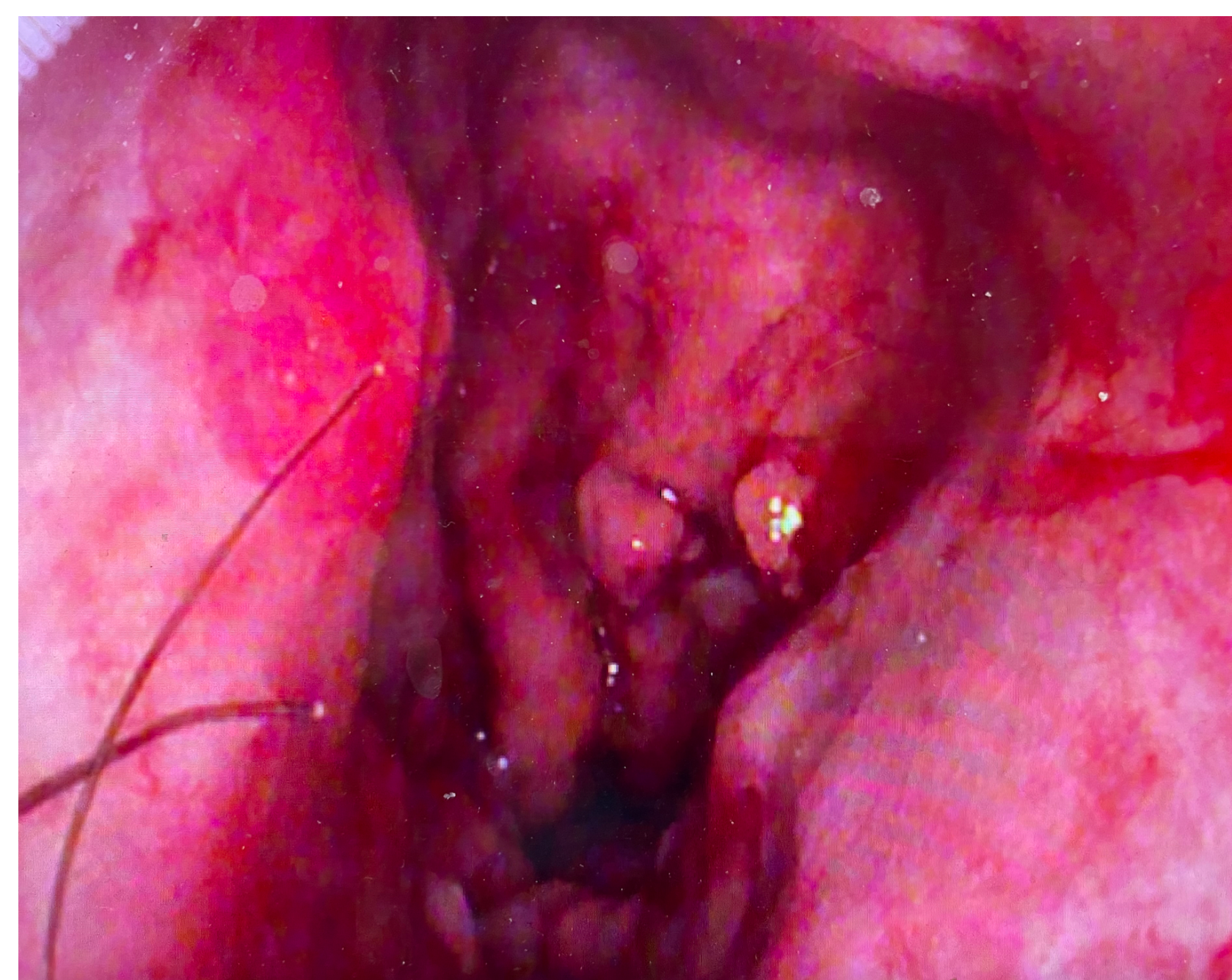


Image 2. Scope images of rectum and lesions requiring surveillance and biopsy

Intra-Operative Technique

- Intraoperatively, he was noted to have several tumors in his small bowel mesentery that significantly shortened small bowel length. Frozen biopsies were sent intraoperatively and confirmed desmoid disease.
- At that point, multiple small bowel lengthening procedures were performed and several desmoid tumors resected but the length of small bowel was insufficient to reach his anus.
- A colonoscopy was performed and biopsies sent for frozen which did not reveal any malignant rectal polyps. The decision was made to proceed with ileorectal anastomosis and diverting loop ileostomy with the acknowledgment that he will need to maintain aggressive rectal screening.

Post-Operative Course

- He is healing well from his surgery and pleased with the pursuit of gastrointestinal continuity on his quality of life. He continues to undergo surveillance sigmoidoscopy with every several months

Conclusion

- This case highlights the complexity of surgical decision making in the setting of FAP, particularly with regards to desmoid disease.
- It highlights the importance of prehabilitation, pre-operative imaging on operative planning, the importance of intraoperative flexibility as well as the patient-centric approach to care.