A Case Report of a Complex Abdominal Wall Desmoid Tumor in a Patient with Familial Adenomatous Polyposis Syndrome

Timothy Cho, DO[1], Nicholas Rawson, DO[2]

[1]Department of General Surgery – Carle Health System

Introduction

Desmoid tumors are clonal fibroblastic proliferations that arise in the deep soft tissues and are characterized by infiltrative growth and a tendency towards local recurrence but an inability to metastasize (2). They occur in 10-20% of patients with familiar adenomatous polyposis (FAP) syndrome (1). Histologically, they are benign processes. However, given their aggressive biologic behavior, they represent the second most common cause of death in patients with FAP (1).

Patient Presentation

We present an interesting case of a 59-year-old male who presented for surgical consultation with a painful enlarging abdominal wall mass.

He first noticed it in 2022 when it was the size of a silver dollar, but it grew significantly over time. He denied any obstructive symptoms. On exam, there was an approximately 20 cm x 15 cm fixed well-circumscribed mass encompassing the left hemiabdomen. There was no involvement of his loop ileostomy.

PMHx: FAP syndrome, necrotizing soft tissue infection of left gluteus

PSHx: total abdominal colectomy (age 14), sharp excisional debridement with diverting loop ileostomy (Feb. 2022), wide local excision of a back mass (desmoid tumor on pathology)

The mass was consistent with a desmoid tumor. He completed an abdominal MRI for pre-operative planning (Figure 1) which revealed a 13.3 cm (transverse) by 11.1 cm (anterior posterior) by 14.5 cm (cranial caudad) anterior abdominal wall mass with surrounding edema and enhancement of the rectus musculature and appeared limited by the rectus sheath.

A multidisciplinary approach was utilized given the complexity of his abdominal wall mass and oncologic history. He was seen by medical oncology for which discussion yielded no significant role for any medical treatment. Plastic surgery discussed their availability for complex abdominal wall reconstruction if necessary. As he also relayed interest in ileostomy reversal, he was seen in consultation with colorectal surgery.

Figure 1: Sagittal view of pre-operative MRI depicting abdominal wall desmoid tumor measuring 13.3 cm x 11.1 cm x 14.5 cm

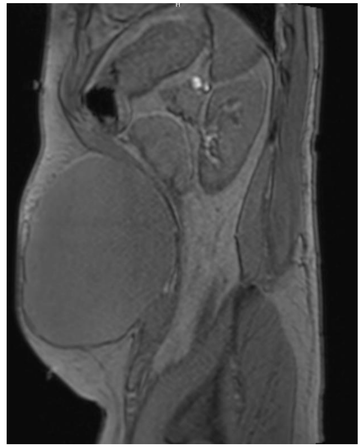
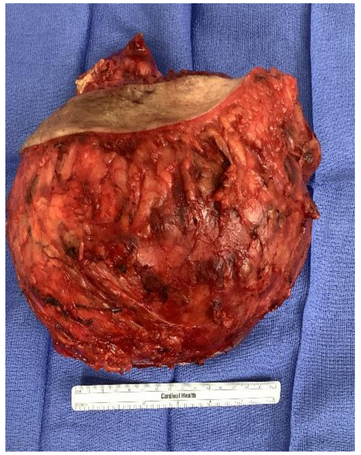


Figure 2: Intraoperative photo of en bloc resection of specimen (abdominal wall, desmoid tumor, and small bowel)



Intervention & Follow-Up

He proceeded with surgical intervention in conjunction with colorectal surgery and underwent exploratory laparotomy, lysis of adhesions, excision of abdominal wall tumor with en bloc small bowel resection with primary anastomosis (Figure 2), complex ventral hernia repair with 20 cm x 25 cm Phasix™ underlay mesh placement and left anterior component separation, complex wound closure, and closure of loop ileostomy. His pathology returned as a desmoid tumor.

His postoperative course was complicated by development of a deep space infection requiring IR drainage and antibiotics with ultimate resolution of his infection. He was seen in clinic post-operatively doing well without an incisional hernia or additional complications.

Discussion & Conclusion

Desmoid tumors can vary in presentation from anatomic location and severity of symptoms. Multiple modalities exist in the treatment of desmoid tumors, including surgery, radiation, NSAIDs, anti-hormonal therapy, tyrosine kinase inhibitors, and chemotherapy (2). One phase II randomized study (DESMOPAZ) demonstrated findings showing clear benefits of tyrosine kinase inhibitors in treatment of desmoid tumors (3). However, assessment of treatment effects remains an unresolved issue and no standard validated response criteria exists (2). In the interim, there should be consideration of a multidisciplinary approach when evaluating and treating desmoid tumors to discuss potential treatment options with patients. Surgical intervention may be necessary for select symptomatic patients.

References

(1) Righetti AE, Jacomini C, Parra RS, de Almeida AL, Rocha JJ, Féres O. Familial adenomatous polyposis and desmoid tumors. *Clinics (Sao Paulo)*. 2011;66(10):1839-1842. doi:10.1590/s1807-59322011001000027

(2) Desmoid Tumor Working Group. The management of desmoid tumours: A joint global consensus-based guideline approach for adult and paediatric patients. *Eur J Cancer*. 2020;127:96-107. doi:10.1016/j.ejca.2019.11.013

(3) Toulmonde M, Pulido M, Ray-Coquard I, et al. Pazopanib or methotrexate-vinblastine combination chemotherapy in adult patients with progressive desmoid tumours (DESMOPAZ): a non-comparative, randomised, open-label, multicentre, phase 2 study. *Lancet Oncol*. 2019;20(9):1263-1272. doi:10.1016/S1470-2045(19)30276-1

Carle Health

HRES032222