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# En Bloc Multivisceral Resection of a Locally Aggressive Desmoid Tumor Requiring Total Gastrectomy, Splenectomy, and Partial Pancreatectomy

**Andrea B. Henry**[1*] **and Wade O. Christopher**[2]

[1]School of Medicine, University of Mississippi Medical Center, Jackson, USA

[2]Division of Surgical Oncology, Department of Surgery, University of Mississippi Medical Center, Jackson, USA

## 1. Abstract

Desmoid tumors are rare, locally aggressive neoplasms of mesenchymal origin with significant morbidity due to their affinity for local invasion. This report describes a 36-year-old female with complex medical and surgical history who presented with obstructive symptoms and ongoing weight loss who was found to have a massive intraabdominal desmoid tumor. The mass was resected en bloc with concurrent total gastrectomy, distal pancreatectomy, and splenectomy which the patient tolerated well. This report demonstrates the feasibility of a multivisceral resection for initial management of intraabdominal desmoid tumor. In recent years, a paradigm shift from early surgical intervention to systemic therapy or active surveillance has occurred, but upfront surgical management remains an important consideration in patients with specific characteristics, and further research is required to better characterize surveillance and best initial management.

Desmoid tumors are rare, locally aggressive neoplasms of mesenchymal origin. Often termed aggressive fibromatosis, this proliferation of fibroblasts arises sporadically or in association with inherited cancer syndromes, most notably familial adenomatous polyposis (FAP). Initial presentation is extensively variable and largely dependent on location with intraabdominal masses typically discovered incidentally or because of mass effect. Diagnosis is confirmed by biopsy demonstrating nuclear β-catenin positivity. Because of their non-metastasizing nature, desmoid tumors have low mortality but exhibit high morbidity due to their affinity for local invasion. The incidence of desmoid tumors is 5-6 cases per 1 million of the population yearly with a peak age of 30-40 years. While the pathogenesis of desmoid tumors is not completely understood, the literature has confirmed alterations in the Wnt/β-catenin signaling pathway in up to 95% of cases. The current recommendations for first-line management include surveillance or systemic therapy with nirogacestat or sorafenib. Surgical resection is recommended only in those patients with large tumors where progression would be morbid, significant symptoms are present or is agreed upon by a multidisciplinary tumor board. When surgical resection is the most therapeutic option, R0 margins are preferred when significant morbidity can be avoided. R1 margins are also acceptable as Cates and Stricker found that only 42% of patients with microscopically positive margins had recurrence of disease in 5 years after resection. We present a case of a massive intra-abdominal desmoid tumor requiring radical en bloc multivisceral resection due to progressive obstructive symptoms and extensive local invasion.

The patient is a 36-year-old African American female with a past medical history significant for gastroparesis, seizure disorder, adrenal adenoma, and congenital hypertrophy of the retinal pigment epithelium (CHRPE) who presented with progressive nausea, vomiting, and a 10-pound unintentional weight loss over approximately one month. The patient has a complex surgical history including laparoscopic cholecystectomy, laparoscopic sleeve gastrectomy, and robot assisted adrenalectomy. Initial computed tomography imaging revealed a large mass in the left upper quadrant of the abdomen. Subsequent magnetic resonance imaging and core needle biopsy was obtained prior to referral to our team. Histopathologic evaluation demonstrated desmoid-type fibromatosis with nuclear β-catenin positivity by immunostaining. Notably, the patient had recently undergone colonoscopy for routine surveillance due to the close association of CHRPE with FAP. Although upfront surgery deviates from current recommendations, the patient's progressive obstructive symptoms and ongoing weight loss made neoadjuvant treatment impractical and risked further morbidity if surgical intervention was delayed.

***Corresponding author:** Andrea B. Henry, School of Medicine, University of Mississippi Medical Center, Jackson, USA

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Intraoperatively, we discovered a massive left upper quadrant tumor with dense adherence to the stomach, pancreas, and left diaphragm. The spleen was completely obscured by the mass, and no direct liver involvement was identified. We performed mobilization of the splenic flexure, medial rotation of the left colon, dissection of the inferior border of the pancreas, and mobilization of the celiac plexus. Because the splenic vessels and distal pancreas were involved, subtotal distal pancreatectomy and splenectomy was required. Mobilization of the stomach could not be performed secondary to dense adherence of the tumor, necessitating total gastrectomy. Further dissection showed involvement of the left hemidiaphragm, necessitating resection. The final specimen, including the tumor, stomach, distal pancreas, spleen, and left hemidiaphragm was removed en bloc. Hemostasis was obtained and copious irrigation performed prior to beginning the reconstruction phase of the procedure. A thoracostomy tube was placed, and primary repair of the diaphragmatic defect was performed. We continued by constructing a Roux-en-Y esophagojejunostomy and ensuring wide patency. Two 19-French Blake drains were placed in the area of the pancreatic stump and esophagojejunal anastomosis. An 18-French red rubber catheter was placed as a feeding jejunostomy. Finally, a vascularized omental pedicle was fashioned and placed in the resection bed. The patient tolerated the procedure well but experienced pancreatic leak which was successfully treated with somatostatin injections and left pneumothorax after thoracostomy was placed to water seal which resolved after three additional days to suction. The patient was deemed medically ready for discharge to an acute rehabilitation facility on postoperative day 8. By 6 weeks after the operation, the patient was tolerating sufficient intake by mouth and no longer requiring feeding via jejunostomy tube.

Final histopathology of the surgical specimen demonstrated a tumor measuring 25.4 x 18.5 x 13.1 cm and confirmed desmoid-type fibromatosis with nuclear $\beta$-catenin positivity in neoplastic cells. The fibromatosis extended to peripheral margins, indicative of an R1 resection, and involved the gastric wall, pancreatic parenchyma, and splenic capsule. Although margins were microscopically positive, this is considered acceptable because of morbidity associated with wider resection and the known prognostic significance. Postoperatively the patient was referred to medical oncology to evaluate candidacy for adjuvant treatment. Genetic testing for suspected attenuated FAP was performed and given the patient's personal history remains a consideration despite negative APC (adenomatous polyposis coli) gene testing.

The current literature describes specific scenarios when surgical resection should be considered over systemic treatment. Further research is needed to understand the molecular basis of desmoid tumor, best initial treatment for large tumors, and for surveillance in those with a genetic predisposition towards developing desmoid tumors. This case demonstrates that radical en bloc multivisceral resection can be safely performed in patients with extensive intra-abdominal desmoid tumors when severe symptoms and organ compromise preclude nonoperative management. Individualized treatment planning and multidisciplinary evaluation remain critical to optimizing outcomes.

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