Management of Desmoid Tumor in a Patient with Familial Adenomatous Polyposis

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INTRODUCTION

- Desmoid tumor can occur sporadically or secondarily as a complication of Familial Adenomatous Polyposis (FAP).
- This locally invasive tumor rarely metastasizes, but can cause significant adverse effects due to rapid growth and compromise of surrounding organ architecture.
- Due to its variable presentation, location and disease course, optimal management of desmoid tumor is controversial.

BACKGROUND

- The patient is a 50 year old female with a prior medical history significant for Familial Adenomatous Polyposis (FAP), Gardner Syndrome, and Human Immunodeficiency Virus (HIV).
- Importantly, at the time of her presentation, the patient was two years status-post open total proctocolectomy with an ileo-anal pouch and diverting ileostomy, due to a prior diagnosis of FAP. The patient reported that since her colectomy, she also suffered from loose non-bloody stools.

CLINICAL COURSE

<table>
<thead>
<tr>
<th>Chief Complaint</th>
<th>Clinical Exam</th>
<th>Preliminary Imaging</th>
<th>Disease Course</th>
<th>Continued Course</th>
<th>Change in Management</th>
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<tbody>
<tr>
<td>Sharp left sided abdominal pain without radiation</td>
<td>Tender abdomen with marked distension</td>
<td>CT of the abdomen and pelvis showed a hypo-dense mass centered within the mesentery, extending from the pelvis to upper abdomen.</td>
<td>Because the tumor was initially deemed inoperable due to size, medical management was initiated (antibiotics, systemic steroids), to which initial response was positive.</td>
<td>Medical management was sufficient for five years until the patient returned presenting with recurrent episodes of abdominal pain and fever.</td>
<td>Recurrent infection raised suspicion for a desmoid abscess (figure 1).</td>
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<td>Increased abdominal girth</td>
<td>Large, firm mid-abdominal mass identified</td>
<td>CT-guided needle biopsy of the mass showed spindle cell proliferation</td>
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<td>Early satiety, worsening post-prandial pain and vomiting</td>
<td>Trace pedal edema</td>
<td>Multiple trials of image-guided drainage and J-tube insertion were required for infected desmoid abscess</td>
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Surgery

- Goals were for adhesiolysis, abdominal wall fistula and desmoid tumor excision.
- Several bowel loops were found within the desmoid tumor.
- Many attempts were made for dissection while avoiding enterotomy.
- After significant dissection, it was determined that further resection would risk short gut and malnutrition; surgery was discontinued.

Overall surgery resulted in removal of the abdominal wall fistula and adhesiolysis; partial desmoid tumor remained. Post-operative treatment with IV antibiotics and patient controlled analgesia pump. Home bicitravir was also restarted for HIV infection.

RADIOLOGY

Panel A: CT showing an abscess within an unresectable desmoid tumor almost 5 years after initial diagnosis of tumor Panel B & C: CT with contrast 3 days following adhesiolysis and removal

CASE DISCUSSION

- Post-operative course was uneventful until day three, when the patient developed a fever to 102.7°F and abdominal pain, despite a clean and dry surgical wound.
- CT showed pneumoperitoneum (figure 2), suspicious for intestinal perforation.
- On day four, another exploratory laparotomy was done and no perforation, infection or other acute intra-abdominal pathology were identified.
- Following procedure, no acute events developed post-operatively and the patient recovered.
- Antibiotics and anti-retroviral therapy were resumed.

CONCLUSIONS

- Desmoid tumor size can range from small and indolent to large and aggressive.
- While there is no definitive cure for desmoid tumors, surgery should be considered with wide resection and adjuvant therapy.

REFERENCES