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Case Report

Sporadic Giant Intra-abdominal Desmoid Tumor: A Case Report of a 8.0-Kg Desmoid Tumor

Muzna Al Farsi[1]*, Rashid Al Alawi[2] and Aamed Al Araimi[2]

[1]*Department of Colorectal Surgery, Royal Hospital, Muscat, Oman*

[2]*Colorectal Consultant, Royal Hospital, Muscat, Oman*

***Corresponding Author:** Muzna Al Farsi, Department of Colorectal Surgery, Royal Hospital, Muscat, Oman.

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Abstract

Introduction: Desmoid tumors are rare lesions that tend to invade locally and recur but without any metastatic potential. Due to its unpredictable behavior, the management is difficult with lots of controversies.

Case Presentation: We report an 8.0-kilograms anterior abdominal wall desmoid tumor in a 29 years old male patient with no previous history of surgery. The tumor was successfully excised as an emergency procedure. The patient had a non-eventful recovery and was followed for over 18-months without tumor recurrence.

Conclusion: DTs are often presenting a challenge in the diagnosis and management. Despite surgical excision of the tumor, recurrence rates are high and continued surveillance is crucial.

Keywords: *Intra-abdominal; Desmoid Tumors (DTs)*

Introduction

DTs are benign locally aggressive tumors, characterized by local infiltration and high recurrence rate after surgical removal. Unlike malignant neoplasms, DTs lack the potential for metastasis [1]. DTs are rare, constituting about 0.03% of all neoplasms and less than 3% of all soft tissue tumors. The reported incidence in the general population is two to four per million population per year [2]. 80% of DTs are sporadic and they occur at sites of trauma, and scars or irradiation. However, the risk increases with pregnancy, trauma and genetic syndromes such as familial adenomatous polyposis (FAP). The most common site of occurrence is the abdominal wall, with an incidence of 50% [3]. The optimal treatment of these tumors is still controversial [4,5].

Case Presentation

29 years old male was referred from a district hospital to a national referral hospital after being diagnosed with an intra-abdominal desmoid tumor.

At initial presentation, Computed tomography (CT) scans showed 20 × 16.5 × 10 cm soft tissue mass. The patient's case was discussed in tumor board and he was offered surgical resection but he refused any surgical intervention.

3 months later, the mass had grown to a size of 21 × 18.5 × 11.5 cm. Although the patient was thoroughly counselled regarding the expected risks and outcome, he willingly chose not to go on with the surgery.

2 months later, he presented to the Emergency Department with 1-day history of severe abdominal pain associated with abdominal distention, vomiting, and constipation. Abdominal pain was generalized and has been progressing in severity, associated with 8 episodes of vomiting.

On clinical admission, the patient was in pain. Vital are WNL. Abdominal examination revealed a tense and tender abdomen. There was a palpable abdominal mass extending from epigastric area to the pelvis. Digital rectal examination revealed normal anorectal mucosa with no palpable masses.

His labs showed the following: haemoglobin 13.2 g/dl, white blood cells 11.1 × 10^9/L, neutrophils 9.5 × 10^9/L, platelets 224 × 10^9/L and C reactive protein 92.4 mg/L, GFR 86 mL/min/1.73 m^2. Other lab tests results were within normal ranges.

He underwent an abdominal CT scan with finings of small bowel thickening and perforation, in view of the presence of the known huge mesenteric mass encasing some of the small bowel loops and its vasculature, the picture is most likely attributed to bowel ischemia (Figure 1).

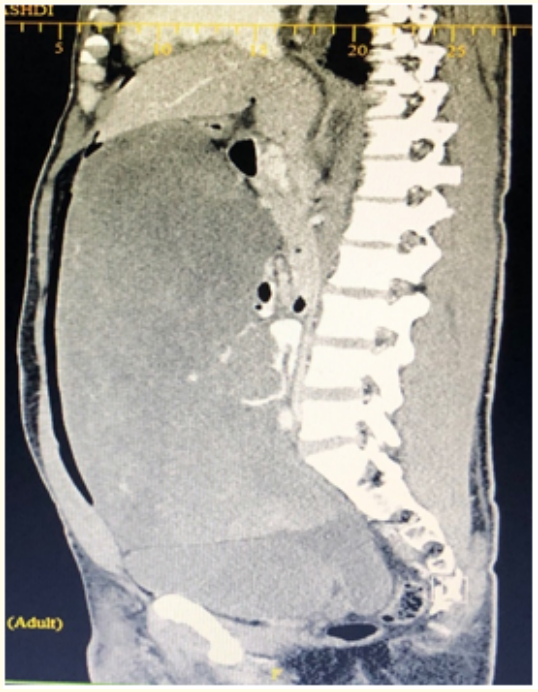

Figure 1: *Sagittal View of huge desmoid tumor with pneumoperitoneum.*

The patient was taken immediately for emergency laparotomy with high risk consent. Midline incision revealed a huge lobulated hypervascular mesenteric mass measuring 40 × 21 × 12 cm, firm in consistency, smooth surface, causing significant compression effect occupying the full intra-abdominal space.

There was a large segment of small bowel (jejunum) and right ascending colon entrapped within the lobulated space of the mass with a perforated gangrenous segment. In addition, the abdominal cavity was full of small bowel content causing generalized peritonitis. The rest of abdominal viscera looks grossly normal.

Right ureter was saved during dissection, however, there was part of the tumor tightly adherent to the ureter. Considering the high risk of local recurrence of this tumor, ureter was divided, and re-anastomosed, right DJ stent was placed (Figure 2).

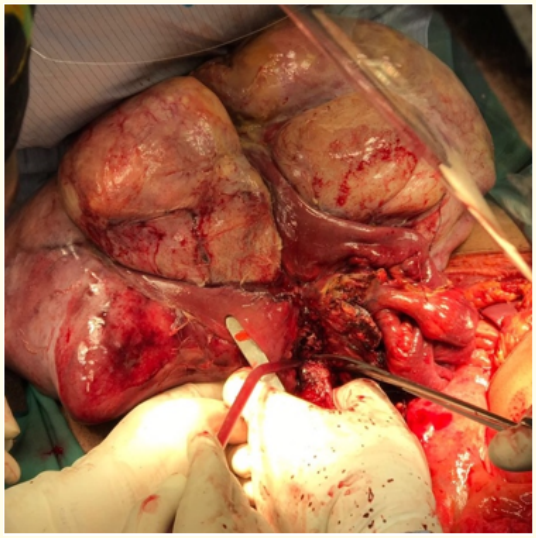

Figure 2: *Intraoperative view of the mass invading the bowel loops and the RT ureter.*

The patient was discharged 5th day postoperatively in a good condition, and he was then reviewed as an outpatient after 12 days, with smooth recovery outcome and without any significant complaints.

The final histopathology resulted as intra-abdominal mesenteric fibromatosis invading small and large bowel as well as a segment of the right ureter. All resected bowel and ureteric margins are free of tumor (Figure 3).

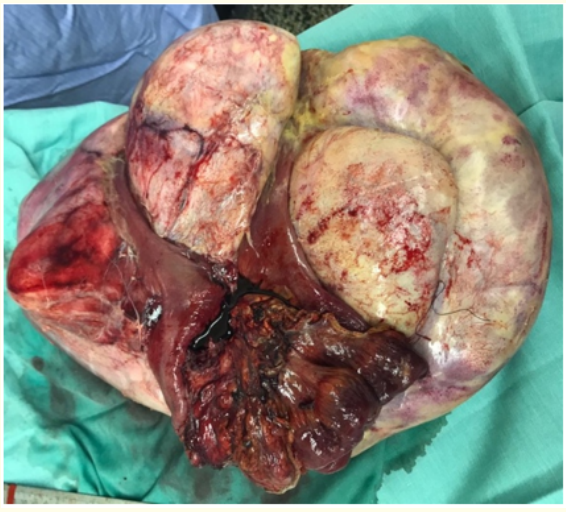

Figure 3: *The retrieved specimen, wt 8 kg.*

Outcome and follow-up

Interval follow up of the patient was done every 3 to 6 months with detailed history and clinical examination. CT scan abdomen and pelvis and colonoscopy repeated on 12th months post-operatively that excluded recurrence. Then patient resumed his full functional and physical status few weeks after surgery and currently he is working in the military service.

Discussion

Desmoid tumors are benign deep fibromatoses, originating from musculoaponeurotic stromal elements with a slowly infiltrating growth [6,7]. These tumors can be either sporadic or associated with pregnancy and trauma and others associated with hereditary cancer syndromes, unlike our patient, who had no family history of any inherited syndromes and no personal history of abdominal trauma, suggesting that it may have occurred sporadically [1,6]. Sporadic DTs are more likely to be extra-abdominal, with only 5% of sporadic DTs located intra-abdominally [8].

According to their location, desmoid tumors are divided into extra-abdominal, abdominal and intra-abdominal [9]. Many studies have shown that between 37% and 50% of desmoids are initiated in the abdominal region which was similar to the location in the case we reported [10].

In many cases -including ours-, patients usually remain asymptomatic or they may have vague abdominal symptoms until their growth and infiltration causes intestinal obstruction, organ ischemia and genitourinary compression which is evidenced by cross-sectional imaging and reveals a locally advanced tumor [4,11].

Regarding treatment, surgical resection is the treatment of choice [12]. As these tumors are usually discovered after they have already progressed in size, adjacent involved structures must be resected as well [3,13]. In the present case, the tumor was embedded in mesentery of the small and large bowel with extension to retro-peritoneal structures and showed extensive adhesions, resection of a large segment of small bowel, right ascending colon and right ureter was performed. Due to the high tumor recurrence rate ranging from 30% to 40%, regular postoperative follow-up needs to be adopted [4].

Other than surgical, medical therapy including antiestrogens, chemotherapy, and radiotherapy have been trialed for the treatment of DTs, though large randomized studies are difficult considering the rarity and varied anatomical presentation of DTs [6,10].

In summary, we encountered a rare case of giant intraabdominal DT in a young male who had no history of surgery, trauma, or inherited syndromes. It was complicated with bowel ischemia and perforation, so optimal treatment was surgery. DTs have a high rate of recurrence, so primary resection with negative margins and close follow-up are necessary.

Conclusion

DTs present a clinical challenge in their diagnosis and management. Wide margin tumor excision is the first line management of desmoid tumors. Considering the high recurrence risk, continued surveillance is crucial.

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