

A Rare Case Report Of “Mesentric Desmoid Tumour”

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Abstract

Desmoid tumors are benign but locally aggressive tumors of mesenchymal origin which are poorly circumscribed. infiltrate the surrounding tissue, lack a true capsule and are composed of abundant collagen. History of trauma to the site of tumor origin is elicited in up to 1 in 4 cases and they most commonly develop in the anterior abdominal wall and shoulder girdle but they can arise in any skeletal muscle. They constitute 3% of all soft tissue tumors and 0.03% of all neoplasms. Abdominal DTs occur sporadically or are associated with certain familial syndromes, such as familial adenomatous polyposis (FAP). A male patient with a Desmoid Tumour of the mesentery who had no relevant family history was admitted to hospital. The patient, who presented with periumbilical pain, had no history of trauma. According to the medical history, physical examination and CT report, the patient was diagnosed with Desmoid tumour. Exploratory laparotomy was performed and tumour excised. The histological diagnosis was of Desmoid tumour. The patient remains in good health and complete remission without any other treatment following surgery. DTs exhibit aggressive growth and have a high rate of recurrence. Surgery is the optimal treatment, and subsequent radiotherapy may decrease the local recurrence rate. Further research into their aetiology is required combined with multicentre clinical trials of new treatments in order to improve management of this disease. This case report provides general knowledge of DT, and may be used as a guidance for diagnosis and treatment. The clinical behavior and natural history of desmoid tumors are unpredictable and management is difficult with many issues remaining controversial, mainly regarding early detection, the role, type and timing of surgery and the value of non-operative therapies.

INTRODUCTION

- “The term desmoid, coined by Muller in 1838, is derived from the Greek word desmos, which means tendon-like”.¹
- “Desmoid tumors (DTs), also known as aggressive fibromatoses, are benign myofibroblastic neoplasms originating from muscular aponeuroses that are also classified as deep fibromatoses”².
- “They constitute 3% of all soft tissue tumors and 0.03% of all neoplasms”³.
- “Desmoid tumours usually occur in fertile females and are uncommon during the menopause; during pregnancy an increase in volume occasionally occurs in already existing tumors. This corroborates the estrogen-stimulated tumor growth hypothesis”⁴.
- “Numerous studies have demonstrated that 37–50% of DTs are initiated in the abdominal area”⁵. “Abdominal DTs occur sporadically or are associated with certain familial syndromes, such as familial adenomatous polyposis (FAP)”⁶.
- “Despite their aggressive local infiltration, DTs lack metastatic potential”⁷. However, the local infiltrations and compressions of surrounding structures demonstrate a high recurrence rate, and in anatomic locations with restricted access to surgical resection, may lead to fatalities.

GENETIC ASPECTS

- “Mutations in the CTNNB1 gene or the APC gene cause desmoid tumors. CTNNB1 gene mutations account for around 85 percent of sporadic desmoid tumors”⁸.
- “APC gene mutations cause desmoid tumors associated with familial adenomatous polyposis as well as 10 to 15 percent of sporadic desmoid tumors”⁸.

CASE SUMMARY

- A 49-year-old male was admitted on 24/10/2015.
- The patient had history of **bilateral hernia repair with meshplasty** 2 years back with **no history of trauma**
- SYMPTOMS:
- **Pain** over infraumbilical region since 15 days.
- Non-billous, non-projectile **vomiting** 2 episode.
- **Anorexia** since 15 days.

EXAMINATION

- INVESTIGATION:
- BLOOD INVESTIGATION:
Hb: 13.5g/dl, WBC: 14,400/μl, Platelets: 221×103/μl, S. Creatinine: 0.8mg/dl, S. calcium: 12.81mg/dl
- USG: Hypochoic lesion (3×3.1cm) with central area of calcification involving mesentery of anterior abdominal wall.

- CECT ABDOMEN:
 - A well defined well margined homogeneously enhancing iso to hypodense mobile lesion in infra-umbilical mesentry showing central calcification within. Possibility of Metastatic carcinoid in mesentry or mesenteric desmoids tumour.
 - Two subcentimeter calculi in right distal upper ureter leading to moderate dilatation of pelvicaliceal system.
 - Small calculus in right kidney.
- TREATMENT: Exploratory Laparotomy followed by excision of mesenteric tumour including a peripheral margin of 2 cm of healthy mesentry.
- HISTOPATHOLOGY: Histologically, composed of collagen that surround spindle cell. The cytoplasm is pale with regular nuclei , with neither mitoses nor giant cell suggestive of DESMOID TUMOUR.
- RADIOTHERAPY : No active line of Radiotherapy treatment require at present . Wait and watch with monthly USG.(As per discussion with radiotherapist)
- OUTCOME: After a follow-up of 1 month from the end of surgery, the patient remains in good health and complete remission without any other treatment.

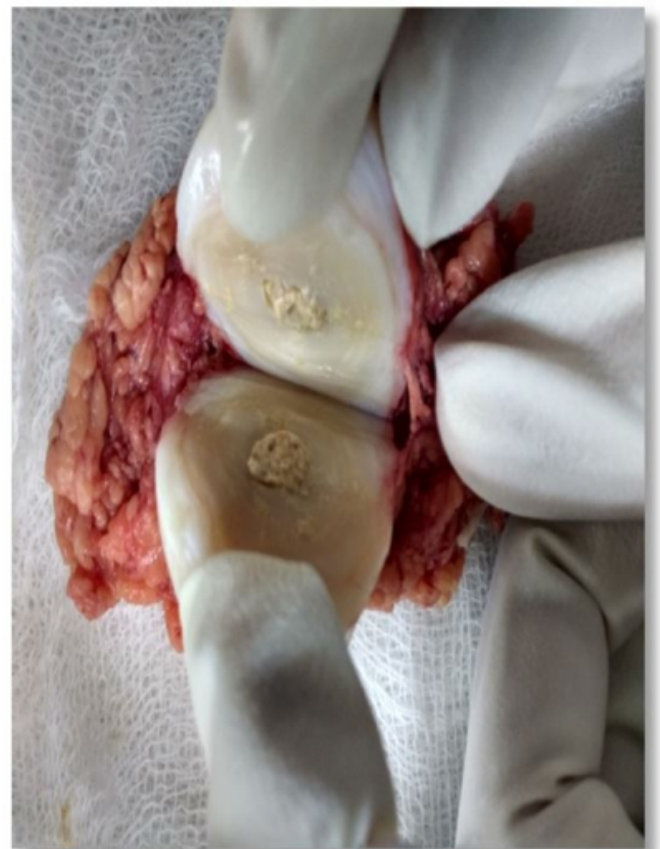
DISCUSSION

- “Incidence of mesenteric desmoid tumour range from 1 case per 200,000-350,000 population” 9.
- “Mostly it is solid or cystic lesion”9.
- It is of benign etiology.
- It is less likely to recur again.
- “Small bowel mesentery is the most common site for intra-abdominal desmoid tumours”10.
- “Additionally, unlike FAP-related cases, the incidence of which is higher in middle-aged women”11, “sporadic MDTs do not show any age or sex predilection “12,13.
- “The pathogenesis of desmoid tumour is unknown, but links have been made to hormonal, genetic and traumatic factors (e.g. previous surgery)”14 .” Notably, previous surgery has been shown to be a less important risk factor in sporadic cases, with only 10% of patients having previous surgery, compared with 83% in FAP”15.
- “Furthermore, post-resection recurrence tends to be less common in sporadic cases than in patients with FAP”10.
- “Although the treatment algorithm remains debatable, the above forms the rationale for surgical resection being preferred in sporadic cases, whereas medical therapy (chemotherapy or hormonal therapy) is advocated in patients with FAP”10,14,16.
- Cross-sectional modalities such as CT and MRI are the preferred imaging modalities for mesenteric masses. “The CT and MRI appearance of MDT is directly related to the histological characteristics, with homogeneity related to the amount of collagenous vs myxoid stroma, as well as to tumour vascularity”17-19 .
- To conclude we report the first case of mesenteric desmoid tumour in Guru Govindsingh Hospital, Jamnagar.

Figure 1



Figure 2



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