International Journal of Pharmaceutical research and Applications
Volume 8, Issue 6, Nov.-Dec. 2023, pp: 1853-1858 www.ijprajournal.com ISSN: 2456-4494

A Rare Case of Desmoid Tumor Secondary to Previous Traumatic Jejunal Perforation Repair

Dr. Omkar R. Khandekar
Final Year PG scholar

Dr. Rashmi A. Kale
Associate professor

Department of Shalyatantra ,Sumatibhai Shah Ayurved Mahavidyalaya ,Hadapsar,Pune 411028

Date of Submission: 04-12-2023 Date of Acceptance: 17-12-2023

Abstract
Desmoid fibromatosis is a rare benign neoplasm of the soft tissue. Primary desmoid neoplasms rarely occur in the small bowel and are primarily found in patients with a previous abdominal surgery or irradiation history. They are challenging to diagnose at the time of presentation due to a lower incidence and non-specific presentation. Making it difficult to distinguish from other intra-abdominal neoplasms, such as gastrointestinal stromal tumors (GISTs), may present with similar symptoms. A case of a 43 year old male, c/o a one week history of pain in abdomen and distension with severe vomiting and rigid abdomen since one day. A computed tomography (CT) scan of abdomen and pelvis showed the presence of mesenteric lesion with solid and cystic component in the left lumbar and iliac region and another lesion in right iliac region causing compression and showing features of small bowel obstruction. The patient was immediately taken for emergency laprotomy with all investigation and fitness for surgery the tumor was completely removed. The specimen collected was sent for histopatholgy with diagnosis of desmoids tumor. The aim of this case review is to highlight the importance of keeping a broad differential diagnosis in a patient with acute abdomen.

KEYWORD- Desmoid tumor, Laprotomy,*Mansarbuda*

I. INTRODUCTION –

Desmoid tumors are locally aggressive but benign soft tissue tumors originating from the mesenchymal cells with a high recurrence rate but without the propensity to metastasize [(1)]. The estimated incidence rate of desmoid tumors is two to four cases per million in the general population. It occurs more commonly in females and ages between 15 to 60years [(2)]. It is found to occur in the intra-abdominal soft tissues with a reported incidence of 8% [(3)]. Studies have found a strong association between familial adenomatous polyposis and desmoid neoplasms [(1,4)]. An association has also been found between desmoid neoplasms and Gardner syndrome [(5)]

In the classical Ayurved reference text a condition ressembles to desmoids tumor mention as *Mansarbuda*. The etiological factor such as traumatic causes are mention which causes *Dusthi* of *mansa* lead to the swelling which is *snigdha*(soft) ,*twakasavarna*(colour of skin) , *Ashmopanam*(Consistency hard like stone), *Avedanam*(painless), *Sthira*(slow growing) & *Apaki*(non-suppurative) it is mentioned as *Asadhya*(not curable) in text[(17)].

II. CASE PRESENTATION –

A 43 year old patient came to OPD with complaint of pain in abdomen and five episode of projectile vomiting with bilious vomitus. On examination the abdomen was rigid and distended at left and right lumbar region with a mass felt at the right iliac region of abdomen. At PR examination the rectum was empty and motion not passed since two days having obstipation and peristalsis was absent

International Journal of Pharmaceutical research and Applications
Volume 8, Issue 6, Nov.-Dec. 2023, pp: 1853-1858 www.ijprajournal.com ISSN: 2456-4494

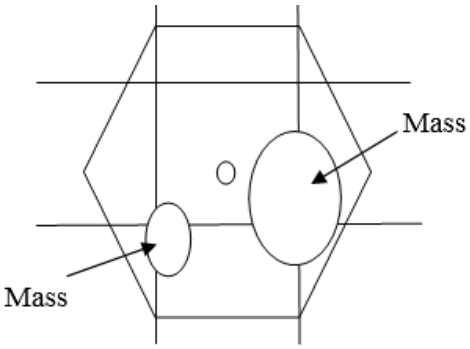


TABLE No. 1 – History of past illness.

HISTORY	
medical history	No any medical history
surgical history	Exploratory laprotomy for abdominal trauma(july2020)
Personal history	Diet – Mixed , Addiction- Tobacco , Sleep – Proper Occupation – Sedentary
Family history	Not Significant
Drug history	Not any

On Examination (On admission)–

1. Nadi /pulse -102/min
2. Mala (stool) – Malavshtambha (Obstipation)
3. Mutra (urine) – Peetavarniya
4. Jihva (tongue)- Samata
5. Kshudha (appetite)-Mandya
6. Shabda (speech)-Prakrut (normal)
7. Sparsha (skin)- Sheetha (Afebrile)
8. Akruti – Madhyam
9. Bala – Madhyam
10. Druk (eyes) – Shweta Varniya

Table No. 2 General Examination-

General condition	Fair ,Patient conscious , alert, oriented to time ,place and person
Temperature	Afebrile
Pulse	102/min
BP	140/80 mmHg
P/A	Tender and Rigid
Nourishment	Poor

Local Examination –

Per/Abdomen –Tenderness present all over abdomen and distended with a hard mass felt in the right iliac region and rigidity noted

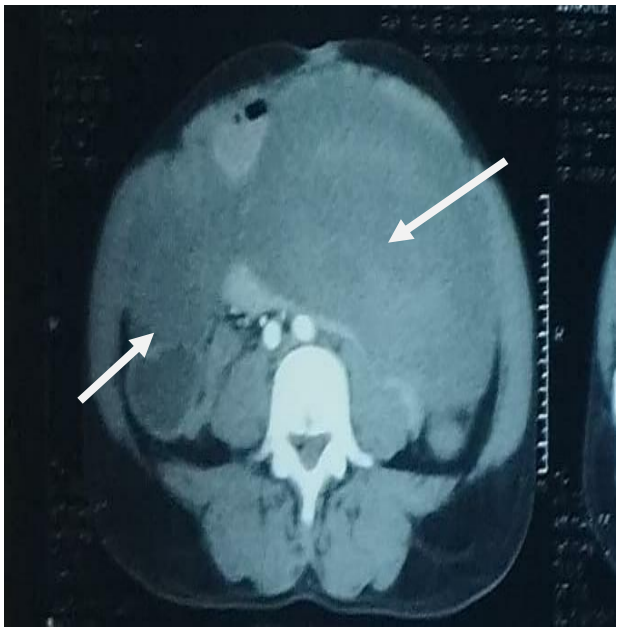
Per/ Rectal –Rectum was empty .

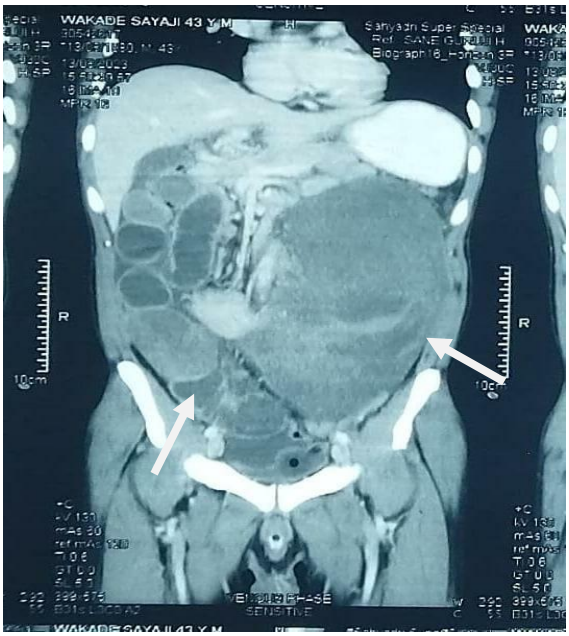
Radiological Examination-

A erect X-ray of the chest and the abdomen were done and it shows multiple fluid level suggesting small bowel obstruction. Then the decision to proceed with a CT abdomen and pelvis was made which showed a well defined oval hypodense mesenteric lesion with solid and cystic components in the left lumbar and iliac region of size approximately 9.8x18x23 cm (AP x TR x CC) and similar lesion in the right iliac region of approx 7.6x6.5x8.1cm (AP x TR x CC) suggesting a benign soft tissue mesenteric tumor and shows dilated distal jejuna , proximal and mid ileal loops

International Journal of Pharmaceutical research and Applications
Volume 8, Issue 6, Nov.-Dec. 2023, pp: 1853-1858 www.ijprajournal.com ISSN: 2456-4494

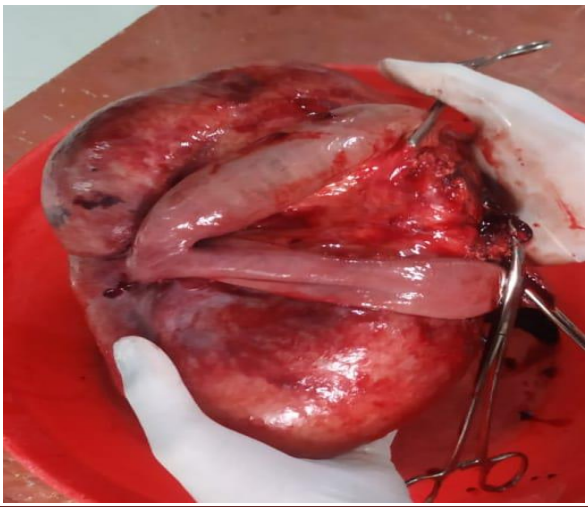
with fluid level and probable zone of transition at the junction of mid and distal ileal loop suggestive of small bowel obstruction.

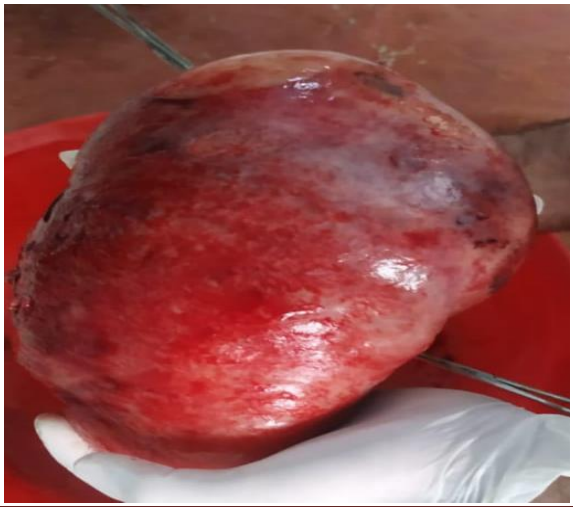




TREATMENT –

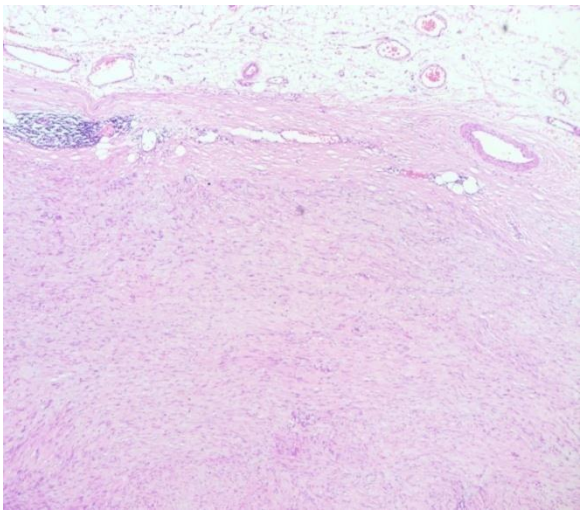
The patient was admitted in ward and Ryles tube insertion and Foleys catheterization done (200ml urine drain). Pre-operative investigation were done and thenThe Patient underwent Exploratory laprotomy which showed the presence of a large intra abdominal mass arising close to the distal ileal mesentery root with the involvement of a segment of the ileum loop from left iliac region of approx 10* 15*20 cm and another mass from right lumbar region adherent to abdominal wall of approx 6*6*10 cm . The ileal segment was densely adherent to tumor and could not be separated from the mass the tumor was resected from the root of the mesentry along with the affected segment of the ileum .After resection of the whole tumor mass the affected segment of the intestine end to end anastomosis was done using PDS(suture matarial). The mass in right lumbar region adherent to abdominal wall was also resected completly. The resected segment was sent for histopathology. A thorough abdominal lavage was done with warm normal saline the abdomen was closed with two drain one in pelvis and other in the Morrison's pouch.

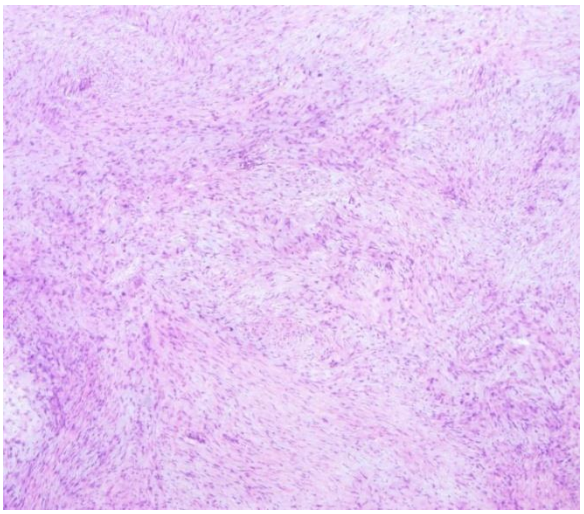




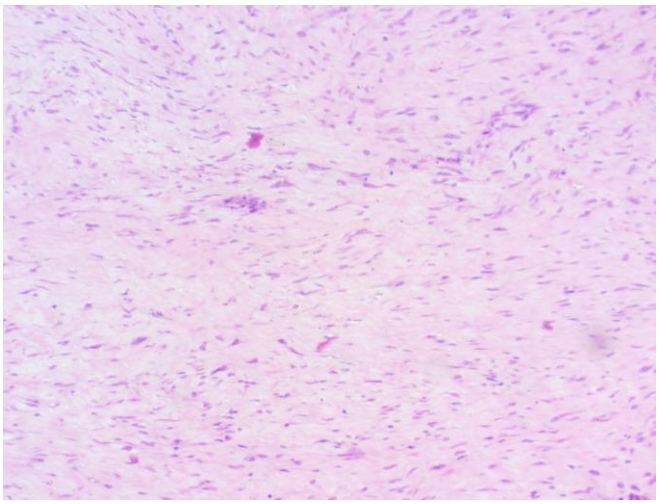
International Journal of Pharmaceutical research and Applications
Volume 8, Issue 6, Nov.-Dec. 2023, pp: 1853-1858 www.ijprajournal.com ISSN: 2456-4494

MICROSCOPIC EXAMINATION –





Histopathology Slides



Immuno histochemistry slide

The peritoneal fluid sent for culture and sensitivity shows many pus cells and the organism isolated was Escherichia coli and showed sensitivity to Piperacillin /Tazobactam which was administered there on.

The Histology report of the collected sample showed an low –grade spindle cell Neoplasm with no necrosis and cut margin unremarkable further Immunohistochemistry was advised and performed and it showed lesional cell express SMA (Smooth Muscle Actin). They are negative for S100, CKit . Desmin and nuclear Beta Catenin is expressed in few cell suggesting Desmoid fibromatosis the patient was stable without any active symptoms at the time of discharge

III. DISCUSSION-

Desmoid tumor are rare , accounting for 0.03% of all tumor overall and 3% of all soft tissue tumor [(6)]. Extra-abdominal desmoids tumor fibromatoses most commonly occur in the abdominal wall (50%) and in exta-abdominal soft tissue of the trunk or limb (40%) [(3)]. However ,abdominal tumor are less common (8%) and mainly affect the mesentry of the small intestine , the ileocecal region and the mesocolon[(3)]. Intra-abdominal desmoids tumors most commonly occur

International Journal of Pharmaceutical research and Applications
Volume 8, Issue 6, Nov.-Dec. 2023, pp: 1853-1858 www.ijprajournal.com ISSN: 2456-4494

after abdominal surgery performed in the past for various reasons[7]. A history of abdominal wall trauma has been associated with the development of desmoids[9]. It is suggested that this may be due to the molecular connection between the wound healing process and the development of fibroproliferative pathologies in the mesenchymal tissue[9]. This could be a possible cause of cancer development in this patient who underwent laprotomy .Other causes associated with the development of desmoids tumor include a history of radiation exposure, genetic mutation such as familial adenomatous polyposis and Gardner syndrome, and a positive family history of desmoids tumor[10-13].This has been reported to due to intranuclear accumulation of Beta-catenin resulting from mutation in the Wnt/Beta-catenin gene . The immunostaining marker Beta- catenin has recently contributed to the detection of antigens capable of distinguishing desmoid tumors from other fibroblastic tumor that may have similar finding in terms of general appearance and histology[14]. Patients histology was typical of a desmoids tumor and his immune-histochemical staining was also strongly positive for Beta-catenin , allowing the diagnosis Early-stage abdominal tumor do not cause any symptoms . Common signs and symptoms therefore include abdominal pain, vomiting, gastrointestinal bleeding and a palpable mass, which is usually only discovered late in the course of the disease.common complications associated with abdominal cancer include enterocutaneous fistulas, intestinal perforation, intestinal bleeding, intestinal obstruction, and ureteral obstruction.[4]. Our patient presented late with an enlarging mass; Consequently patient come with classical case of acute abdomen with intestinal obstruction . Due to the unclear etiology and life-threatening condition, surgical treatment is planned in our case. However,it has emerged that there are various options available for treating patient with desmoids tumors. Chemotheraphy ,radiation therapy and hormone therapy with non-steroidal anti-inflammatory drugs are effective. It is recommended to start treating these tumors with less toxic medication and then switch to more effective medication as needed. Patient are offered a wait and see strategy and active intervention is recommended only in patient with acute symptoms[15]. Nevertheless the mainstay of management in the past involved using low dose of chemotheraphy. However recent development have found good responses to tyrosine kinase inhibitors like sorafenib and pazopanib[16]

IV. CONCLUSION-

Thorough history of patient can lead to provisional diagnosis. Though certain tumours are rare still need to go for biopsy before going for any laprotomy. Prior Histopathological diagnosis can avoid major laprotomy and could be benifited with other feasible non operative measures. In view of the above case many abdominal neoplasm can be seen. Being a less common among the patient it should be included in the differential diagnosis of patient with signs of intestinal obstruction.

REFERENCES-

[1]. Fukayama M, Goldblum JR, Miettinen LA: Mesenchymal tumors of the digestive system. WHO Classification of Tumours - Digestive System Tumours. WHO, Lyon, France; 2019. 1:446-7.
[2]. Dinauer PA, Brixey CJ, Moncur JT, Fanburg-Smith JC, Murphey MD: Pathologic and MR imaging features of benign fibrous soft-tissue tumors in adults. Radiographics. 2007, 27:173-87. 10.1148/rg.271065065
[3]. Antonescu CR, Bridge JA, Cunha IW, et al.: Soft tissue tumors. WHO Classification of Tumours - Soft Tissue and Bone Tumours. WHO, Lyon, France; 2020. 3:93-6.
[4]. Haddad FG, El Bitar S, Barakat I, Deeb L: Desmoid tumor as an initial presentation of familial adenomatous polyposis: a review of the literature. Cureus. 2018, 10:e2297. 10.7759/cureus.2297
[5]. Braschi-Amirfarzan M, Keraliya AR, Krajewski KM, et al.: Role of imaging in the management of desmoid-type fibromatosis: a primer for radiologists. Radiographics. 2016, 36:767-82. 10.1148/rg.2016150153
[6]. Papagelopoulos PJ, Mavrogenis AF, Mitsiokapa EA, Papaparaskeva KT, Galanis EC, Soucacos PN: Current trends in the management of extra-abdominal desmoid tumours. World J Surg Oncol. 2006, 4:21. 10.1186/1477-7819-4-21
[7]. Damiani G, Mikhael R, Tzanis D, El Zein S, Bonvalot S: Desmoid tumors arising on the mesenteric surgical scar of abdominal sarcomas. Cureus. 2022, 14:e21727. 10.7759/cureus.21727
[8]. Robinson WA, McMillan C, Kendall A, Pearlman N: Desmoid tumors in pregnant

and postpartum women. Cancers (Basel). 2012, 4:184-92. 10.3390/cancers4010184

[9]. V. Ravi, S. R. Patel, C. P. Raut, et al.: Desmoid tumors: Epidemiology, risk factors, molecular pathogenesis, clinical presentation, diagnosis, and local therapy. UpToDate. Ted. W. Post (ed): UpToDate, Waltham, MA; 2023.

[10]. Lynch HT, Fitzgibbons R Jr: Surgery, desmoid tumors, and familial adenomatous polyposis: case report and literature review. Am J Gastroenterol. 1996, 91:2598-601.

[11]. Ezhapilli SR, Moreno CC, Small WC, Hanley K, Kitajima HD, Mittal PK: Mesenteric masses: approach to differential diagnosis at MRI with histopathologic correlation. J Magn Reson Imaging. 2014, 40:753-69. 10.1002/jmri.24690

[12]. Leal RF, Silva PV, Ayrizono Mde L, Fagundes JJ, Amstalden EM, Coy CS: Desmoid tumor in patients with familial adenomatous polyposis. Arq Gastroenterol. 2010, 47:373-8. 10.1590/s0004-28032010000400010

[13]. Wang YC, Wong JU: Complete remission of pancreatic head desmoid tumor treated by COX-2 inhibitor-a case report. World J Surg Oncol. 2016, 14:190. 10.1186/s12957-016-0944-z

[14]. Bhattacharya B, Dilworth HP, Iacobuzio-Donahue C, et al.: Nuclear beta-catenin expression distinguishes deep fibromatosis from other benign and malignant fibroblastic and myofibroblastic lesions. Am J Surg Pathol. 2005, 29:653-9. 10.1097/01.pas.0000157938.95785.da

[15]. The management of desmoid tumours: a joint global consensus-based guideline approach for adult and paediatric patients. Eur J Cancer. 2020, 127:96-107. 10.1016/j.ejca.2019.11.013

[16]. Agresta L, Kim H, Turpin BK, et al.: Pazopanib therapy for desmoid tumors in adolescent and young adult patients. Pediatr Blood Cancer. 2018, 65:e26968. 10.1002/pbc.26968

[17]. Shastri AD Sushruta, Sushruta Samhita, Ayurvedatatva sandipika Hindi Commentry Nidansthan , 11/17-19 Varanasi : Chaukhamba Sanskrit Samsthana ; 2001.