AUCTORES
Globalize your Research

Journal of Surgical Case Reports and Images

Divya Dahiya *

Open Access

Case Report

Caecal desmoid tumour – A rare tumour at uncommon location and review of literature

Kishore Abuji[1], Anil Naik[1], Tanvi Jain[1], Kim Vaiphei[2], Divya Dahiya[1*]

[1]Department of General Surgery, Post Graduate Institute of Medical Education and Research, Chandigarh 160012, India

[2]Department of Pathology, Post Graduate Institute of Medical Education and Research, Chandigarh 160012, India

***Correspondence Author:** Divya Dahiya, Department of General Surgery, Post Graduate Institute of Medical Education and Research, Chandigarh 160012, India.

Received Date: April 29, 2021; Accepted Date: May 03, 2021; Published Date; July 19, 2021

Citation: Abuji K, Naik A, Jain T, Vaiphei K, Dahiya D. (2021) Caecal desmoid tumour – A rare tumour at uncommon location and review of literature. *Journal of Surgical Case Reports and Images* 5(1); **DOI:**10.31579/2690-1897/075

Abstract:

A 49-year female presented to surgery outpatient clinic with the awareness of a right lower abdomen asymptomatic lump for one week. Contrast enhanced computed tomography of the abdomen demonstrated a 10x11x15cm heterogeneous lesion anterior to the ileocecal junction with the possibility of leiomyosarcoma. Patient was prepared for laparotomy and intraoperative there was a large tumour arising from the medial wall of cecum. Right hemicolectomy was performed, and histopathology came as a desmoid tumour of the cecum. The patient had an uneventful postoperative course and is well on one year follow up.

Key Words: desmoid tumour, caecum, surgery, right hemicolectomy

Introduction:

Desmoid tumour (DT) is a benign tumour, and it arises from musculoaponeurotic structures with an estimated incidence of 0.03% of all malignancies and less than 3% of all soft tissue tumours [1]. Only about 12-18% of DTs are intra-abdominal, and they commonly arise from the mesentery [2]. Although benign it is a slow-growing locally aggressive tumour with no metastatic potential. Herein we report a case of a 49-year female that presented with an asymptomatic abdominal lump mimicking gastrointestinal stromal tumour on imaging and was managed surgically by en bloc excision. The final diagnosis of histopathology came as a caecal DT.

Case presentation:

A 49-year female with no known co-morbidities presented to surgery outpatient clinic with an awareness of lump in the right lower abdomen for one week. The lump was not associated with pain or symptoms suggestive of intestinal obstruction or gastrointestinal bleed. She had normal appetite and no history of weight loss or recent change in bowel habits. She was an averagely built and nourished lady. Her general physical examination was unremarkable. On per abdominal examination, there was a 10x10 cm well-defined firm mass in right iliac fossa with a smooth surface and with limited mobility both in the transverse and longitudinal plane. Colonoscopy and blood investigations, including CEA and CA125 were normal. On Contrast enhanced computed tomography (CECT) of the abdomen, there was a 10x11x15cm homogenous mass lesion with mild enhancement anterior to ICJ (Figure 1).

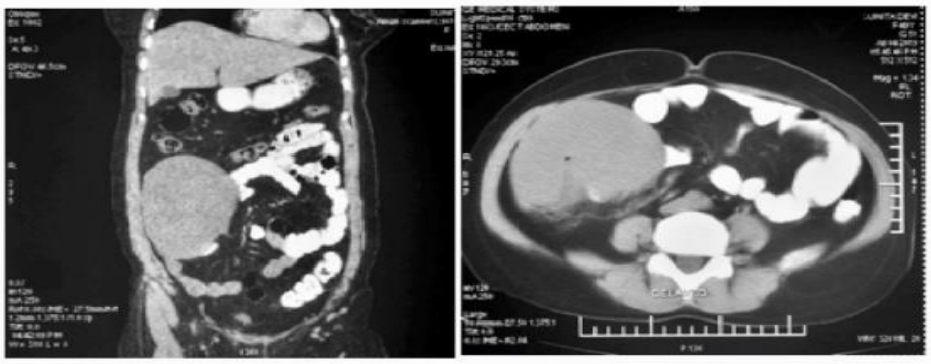

Figure 1: *A) a: Coronal image showing a well-defined homogeneously enhancing soft tissue lesion in the right lumbar region, the caecum is not seen separately from the lesion suggesting the likely origin. b: the ileal loops are displaced towards the midline. B) axial image depicting the lesion with displacement of the ileal loo*

Cecum was not seen distinctly, and the appendix was normal. A provisional diagnosis of a caecal mass likely leiomyosarcoma was made.

Patient was prepared for surgery, and on exploration, there was about 17x10 cm tumour arising from the medial wall of the caecum and adjoining mesentery (Figure 2). Liver, small and large bowel, appendix, peritoneum and ovaries were normal. Right hemicolectomy with ileocolic anastomosis was performed. On gross examination, the caecum on its serosal aspect showed a large encapsulated mass measuring 16.5x12x9cm. There was a solid, homogenous, shiny, mucinous tumour with fibrous streaks with no definite areas of necrosis on cut section. Tumour was strictly restricted to submucosa grossly with no capsular breach. Microscopic examination revealed a tumour from the serosal aspect, composed of spindle cells arranged in interlacing fascicles. The tumour cells had a bland nuclear morphology with wavy to oval nucleoli and moderate eosinophilic cytoplasm, with no mitosis (Figure 3). On immunohistochemistry, beta-catenin positivity and negative staining for c-kit and DOG-1 helped clinching the diagnosis of a desmoid tumour.

The postoperative course was uneventful and patient was discharged on postoperative day 7 in a stable condition. She is well after one year of surgery without any evidence of clinical or radiological recurrence.

Figures:

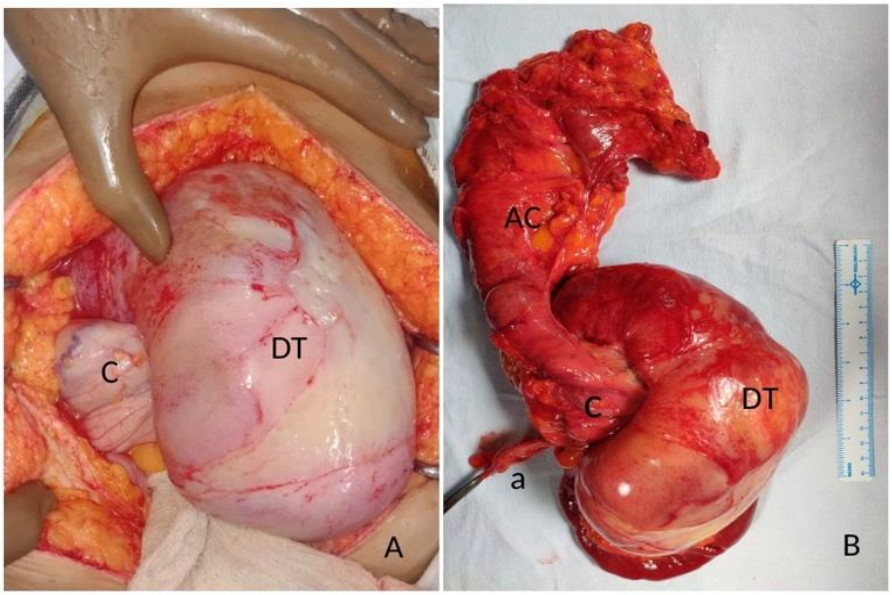

Figure 2: *Intraoperative view:* **A)** *A desmoid tumour (DT) arising from the medial wall of the caecum(C).* **B)** *Right hemicolectomy specimen: DT – encapsulated desmoid tumour,* **C-** *Caecum, AC – ascending colon, a – appendix*

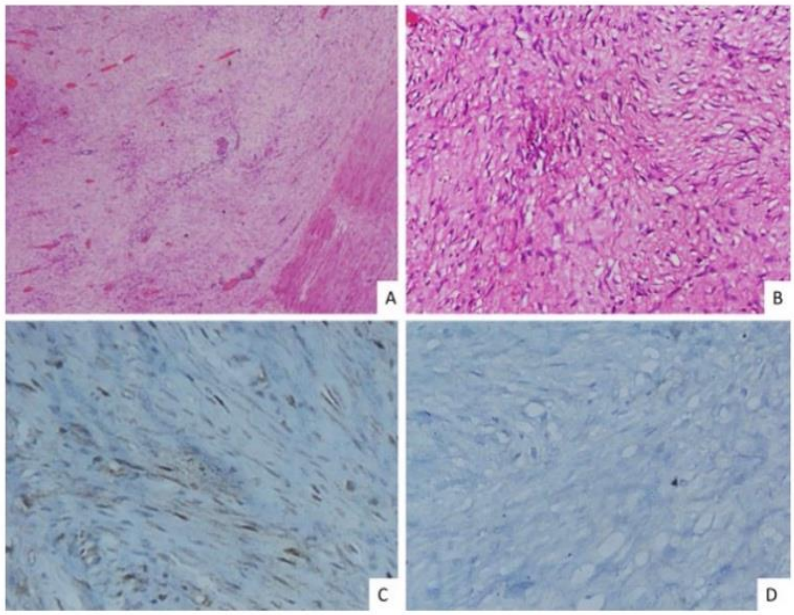

FIGURE 3: *Tumor composed of spindle cells arranged in interlacing fascicles*

(A - hematoxylin and eosin stain, magnification 100x; B - hematoxylin and eosin stain, magnification 400x), Immunohistochemistry reveals tumour cells show nuclear positive (C - immunoperoxidase stain, magnification 400x) and cytoplasmic positivity (D - immunoperoxidase stain, magnification 400x) for beta-catenin

Discussion:

DTs are sporadic (80%), and only 20% are associated with familial adenomatous polyposis (FAP) syndrome or Gardner syndrome [1]. They may be associated with trauma, pregnancy and use of oral contraceptive pills; and can present at any age but more commonly seen between 25-35 years with a slight female preponderance [3]. As intra-abdominal DTs are slow-growing and locally infiltrative; the patient may be asymptomatic, to begin with and diagnosed incidentally on imaging performed otherwise. When they attain a large size or because of inherent infiltrative nature they may cause obstructive or compressive symptoms; and present with vague abdominal complaints, abdominal pain or lump, intestinal obstruction or perforation and bleeding secondary to compromise of blood supply and submucosal ischemia. They may also present with compression of ureter causing hydroureteronephrosis and splenic vein compression leading to portal hypertension [4].

Diagnosis of intra-abdominal DTs is tough due to its rarity and resemblance to other mesenchymal tumours. CECT abdomen aids in defining tumour size, location, invasion of adjacent structures and resectability. Most DTs on CECT scan appear as well defined or poorly defined homogeneous tumour which may be iso or hyperdense compared to muscle [5].

Mesenteric tumours may have strands radiating into adjacent fat or as a whorled appearance of fibrosis. MRI is another modality to diagnose DTs. MRI characteristics of DTs depend upon the histological component of the tumour. A fibrotic and collagenous portion of DT has shown low signal intensity on T2 weighted images and mild enhancement on the delayed phase of post contrast images; cellular stroma and myxoid matrix manifest as hyperintense areas on T2 images. Presence of T1 and T2 hypointense bands are seen within the tumour is reported to be a characteristic MRI finding of DTs (band sign), and is present in about 60-90% of DTs [6].

The role of FDG-PET is not fully defined for diagnosis and treatment, but can be used to estimate the response to chemotherapy [7]. On histopathological examination, these are characterized by proliferative spindle cells in the background of abundant collagen [8].

Mesenteric desmoid has large amount of myxoid stroma compare to intrabdominal DTs. Typical morphological features of desmoid tumour include the presence of proliferative, involutional and residual phase simultaneously with positive staining for nuclear b-catenin, vimentin and smooth muscle actin. They rarely stain positive for cyclooxygenase 2, tyrosine kinase, androgen and estrogen receptors but are always negative for desmin, S-100, CD34 and c-KIT [9].

As most of these tumours occur sporadically, these tumours must be identified and operated owing to their locally aggressive nature and potential complications of perforation, obstruction and abscess formation. Delay in surgery may lead to need for more radical procedures due to extensive infiltration and increased morbidity to the patient. There are no definite guidelines available for the management of DTs. However, asymptomatic patients in the absence of biologic prognostic factors (initial tumour size, growth rate) which are capable of predicting the natural history of the disease, watchful waiting is a reasonable approach to minimise overtreatment and unnecessary morbidity in a subset of patients [10]. Also, in familial cases, patient should be kept under follow up and to be operated only if indicated as in these cases recurrence rate up to 75% has been reported after surgery [11]. Surgery aims at complete resection of the tumour so as to achieve negative margins. Patients with involved resection margins experience recurrence within 15-24 months [12]. Therefore, adjuvant treatment should be given in cases of incomplete resection. When the tumour is locally advanced involving vital structures and surgery is not feasible, options of radiotherapy, chemotherapy and systemic hormone therapy should be considered. Thus, a stepwise multidisciplinary approach with the involvement of medical oncologist may lead to a better outcome. Systemic treatment includes non-steroidal anti-inflammatory drugs (Sulindac& Celecoxib), tamoxifen (suppresses DT growth due to the presence of estrogen receptor on tumour cells), tyrosine kinase inhibitors (imatinib, sunitinib, sorafenib), and chemotherapy(doxorubicin and methotrexate with vinca alkaloids) which can be used as adjuvant or neoadjuvant treatment with or without radiotherapy [13, 14].

The patient should be followed up regularly to detect early recurrence or relapse even after achieving R-0 resection. NCCN guidelines recommend 3-6 monthly follow up for 2-3 years and annually thereafter [15].

Conclusion:

DTs are rare and locally aggressive with a variable clinical presentation. Therefore, a high level of clinical suspicion is required for its diagnosis. Resection with negative margin is the gold standard treatment for sporadic tumours. However, radiation and systemic treatment may be required in cases of residual tumour or recurrence.

References:

1. Li Destri G, Ferraro MJ, Calabrini M, Pennisi M, Magro G. (2014). Desmoid-type fibromatosis of the mesentery: report of a sporadic case with emphasis on differential diagnostic problems. *Case Rep Med;* 850180
2. Georgiades C, Vallianou N, Argyrakos T, Aristodimou A, Kolovelonis G. (2012). An unusual case of desmoid tumour presenting as haemorrhagic shock. *Ann R Coll Surg Engl*; 94:81-82
3. Eastley N, Hennig I, Esler C, Ashford R. (2015). Nationwide trends in the current management of desmoid (aggressive) fibromatosis. *Clin Oncol* (R Coll Radiol);27:362-368
4. Peled Z, Linder R, Gilshtein H, Kakiashvili E, Kluger Y. (2012). Cecal fibromatosis (desmoid tumour) mimicking periappendicular abscess: a case report. *Case Rep Oncol*;5:511-514
5. Xhaja X, Church J. (2013). Small bowel obstruction in patients with familial adenomatous polyposis related desmoid disease. *Colorectal Dis*;15:1489-1492
6. Dinauer PA, Brixey CJ, Moncur JT, Fanburg-Smith JC, Murphey MD. (2007). Pathologic and MR imaging features of benign fibrous soft-tissue tumours in adults. *Radiographics*; 27:173-187
7. Basu S, Nair N, Banavali S. (2007). Uptake characteristics of fluorodeoxyglucose (FDG) in deep fibromatosis and abdominal desmoids: potential clinical role of FDG-PET in the management. *Br J Radiol*;80:750-756.
8. Misemer BS, Skubitz AP, Carlos Manivel J, Schmechel SC, Cheng EY, et al. (2014). Expression of FAP, ADAM12, WISP1, and SOX11 is heterogeneous in aggressive fibromatosis and spatially relates to the histologic features of tumour activity. *Cancer Med*;3:81-90.

9. Kotiligam D, Lazar A, Pollock R, Lev D. (2008). Desmoid tumour: a disease opportune for molecular insights. *Histol Histopathol*; 23:117-126
10. Kasper B, Baumgarten C, Bonvalot S, Haas R, Haller F, et al. (2015). Management of sporadic desmoid-type fibromatosis: a European consensus approach based on patients' and professionals' expertise – a sarcoma patients EuroNet and European Organisation for Research and Treatment of Cancer/Soft Tissue and Bone Sarcoma Group initiative. *Eur J Cancer*;51:127-136.
11. Williams AD, Heightchew K, Siripirapu V. (2016). Diagnostic and therapeutic dilemmas in intra-abdominal desmoid tumours: a case report and literature review. *Int J Surg Case Rep*;26:150-153
12. Lev D, Kotilingam D, Wei C, Ballo MT, Zagars GK, et al. (2007). Optimizing treatment of desmoid tumours. *J Clin Oncol*;25:1785-1791
13. Kasper B, Ströbel P, Hohenberger P. (2011). Desmoid tumours: clinical features and treatment options for advanced disease. *Oncol;*16:682
14. Yao X, Corbett T, Gupta A, Kandel R, Verma S, et al. (2014). A systematic review of active treatment options in patients with desmoid tumours. *Curr Oncol*;21:613
15. Margaret VM, John M, Kane RS, Marilyn M, Edwin C, et al. (2019). Plymouth Meeting, PA (USA): National Comprehensive Cancer Network.

DOI: 10.31579/2690-1897/075