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Case Report

Surgery Section

Mesenteric Fibromatosis (Desmoid Tumour) - A Rare Case Report

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ABSTRACT

Mesenteric Fibromatosis (MF) is a proliferative fibroblastic lesion of small intestinal mesentery. It constitutes 8% of all desmoid tumours, which represent 0.03% of all neoplasm. It is histologically benign but may invade locally and recur after excision. It occurs sporadically or in association with Familial adenomatous polyposis (FAP) mutation as a component of Gardner's syndrome. The presenting features of MF are asymptomatic abdominal mass, abdominal discomfort or pain, bowel or ureteral obstruction, intestinal perforation, fistula, functional impairment of ileoanal anastomosis following colectomy in FAP cases. A 29-year-old male presented with a swelling on the right side of the umbilicus for six months and dull aching pain for two months. Fine needle aspiration cytology, ultrasonography, contrast enhanced computerized tomography findings were inconclusive. After Exploratory laparotomy, a mass approx 6x5x4 cm in ileal mesentery was identified and excised along with 20cm of ileum. End to end anastomosis was done and specimen was sent for histopathology which confirmed the diagnosis of MF. Considering the rarity of this tumour and difficulties in diagnostic and therapeutic ambit, we believe it is justified to describe this case which came to our observation.

Keywords: Desmoid tumour, Gardner's syndrome, Mesenteric fibromatosis

CASE REPORT

A 29-year-old male presented with complaints of swelling in the right side of umbilicus for six months associated with dull ache for two months. It was gradually increasing in size and there was no other symptom. There was no significant past or family history.

A globular intraperitoneal lump approx 5x4 cm was found in umbilical region which was firm, mobile, and mildly tender with smooth surface and margin, no fixity but not moving with respiration. Ultrasonography (USG) of whole abdomen showed it as intra abdominal abscess or cyst. Fine needle aspiration cytology revealed a few monolayered clusters of colonic mucosal cells, some muscle fibres and mucin strings in the background of blood with no atypical or malignant cells. Contrast enhanced computerized tomography Revealed an oval hypodense (+25 HU) and mildly enhancing (+46 HU) focal lesion in intraperitoneal cavity with retroperitoneal extension suggestive of leiomyoma or GastroIntestinal Stromal Tumour. Colonoscopy and endoscopy showed no polyps.

Exploratory laparotomy revealed a mass 6x5x4 cm [Table/Fig-1] in ileal mesentery. It was excised along with 20cm of ileum and end to end anastomosis in two layers.

Histopathologically [Table/Fig-2], a lesion in the subepithelium composed of cells with blunt ended nuclei arranged in sheets, fascicles, whorl and haphazard patterns was found. Hyalinised capillaries were interspersed with focal lymphocytic infiltration and cells with moderate nuclearr atypia in peripheral areas characterized MF. Postoperative days were uneventful and discharged on 10th day. Adjuvant radiotherapy (50 Gy) was given to our patient in the post operative period. Follow up for three years which included USG abdomen and colonoscopy didn't reveal any new pathological lesions.

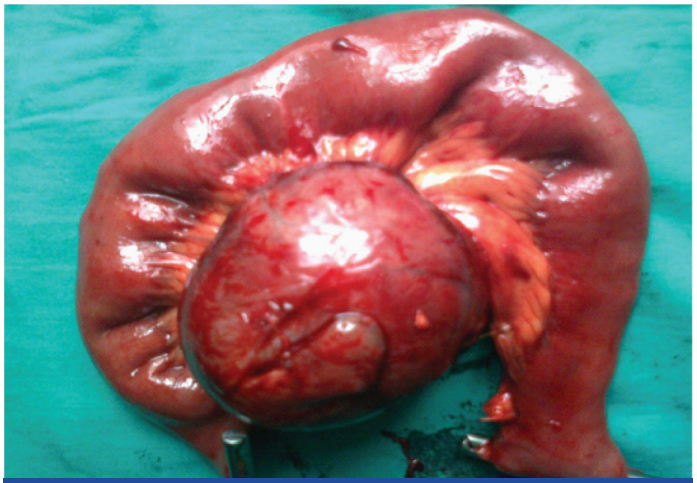
DISCUSSION

The term 'desmoid' was coined by Muller in 1838 and was derived from Greek word 'desmos' meaning band or tendon [1]. These are rare tumours, arising from musculo-aponeurotic elements accounting for about 0.03% of all neoplasm and 3.5% of all fibrous tissue neoplasm [2]. It is commonly seen in reproductive years of women, often during and after pregnancy [1,3]. Desmoid tumours (DT) are histologically benign, but they may show local recurrence after excision [3].

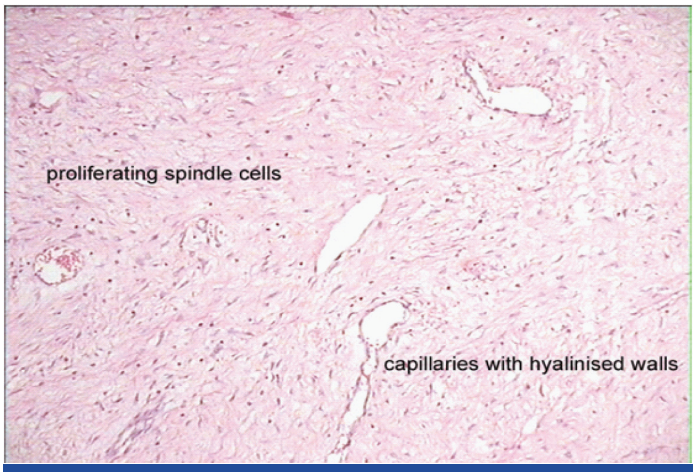
They are classified into three types: abdominal, extra abdominal and intra abdominal [1]. Intra abdominal desmoid or fibromatosis is rare, locally aggressive mesenchymal tumour that usually develops in the mesentery or retro-peritoneum [4]. In our case it was present in the ileal mesentry. It occurs sporadically or in association with 10% to 18% of familial adenomatous polyposis mutation (Gardner's syndrome) [2,4]. In the present case, aetiology is most likely to be sporadic. Extra abdominally these are found in the limbs, pelvis and shoulder girdle [3]. MF represents 8% of all DT, and is usually seen in ileal mesentery and occasionally in mesocolon or gastro-hepatic ligament [5].

Aetiology of DT includes trisomy of chromosome 8 or 20 (sporadic type) [3], trauma, previous abdominal surgery, hormonal stimulation and sometimes de novo [4].

Tumour may present as asymptomatic or painful abdominal mass, bowel or ureteral obstruction, mucosal ischaemia, intestinal perforation, fistula, pyrexia of unknown origin or functional impairment



[Table/Fig-1]: Gross specimen of the MF in ileal mesentry



[Table/Fig-2]: Microscopy showing spindle cells and hyalinised capillaries interspersed with focal lymphocytic infiltration

of ileoanal anastomosis following colectomy in FAP cases [3-5]. In our case patient presented with dull aching pain associated lump in the right side of abdomen without any pressure or obstructive symptoms.

CT scan and MRI show its resectibility and adhesion to neighbouring structures. Histological examination is the only means for confirmation [3]. The imaging studies in the work up of our case were inconclusive. Differential diagnoses include GIST, lymphoma, carcinoid tumour, fibrosarcoma or inflammatory fibroid polyp [1]. In our case, CECT abdomen revealed hypodense (+25HU) and mildly enhancing (+46HU) focal lesion in intraperitoneal cavity with retroperitoneal extension with a possibility of leiomyoma or GIST.

Treatment modalities include wide excision, antiestrogen, radiotherapy and chemotherapy though efficacy is unpredictable [1]. Fotiadis C et al., [6], opined that DT in the abdominal wall is easier to remove and recurrence rates are lower when compared to mesenteric or retroperitoneal DT. Despite radical surgery and adjuvant radiotherapy 25%-50% recur [1]. Neo adjuvant radiotherapy may be tried [3]. Kriz J et al., [7], in their study on role of radiotherapy in DT have shown its usefulness with a local control rate of 79% with long term follow up and have also proposed it as an alternative to mutilating surgery. Adjuvant radiotherapy (50 Gy) was given to our patient in the postoperative period. Pinheiro LV et al., [8] have suggested imaging techniques such as CT scan and/or ultrasound in the surveillance for its early detection of recurrence. In the present case we followed up with USG and colonoscopy for three years.

CONCLUSION

Desmoid tumour, though rare must be considered as differential diagnosis in the middle age. Multidisciplinary approach is needed with long term follow up for detecting tumour recurrence. Radiotherapy needs to be considered as a therapeutic modality.

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