Desmoid Tumour - A Rare Entity

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INTRODUCTION

Desmoid tumours are myofibroblastic tumours that are one of the rarest soft tissue tumours to occur. The tumours are more common in the proximal region of upper extremities, abdominal wall and mesentery. The tumours are non-cancerous and therefore have no metastatic potential but can invade the surrounding structures locally. The recurrence rate is high; so complete surgical excision is mandatory. We report a case of 65-year-old man with a complaint of painless abdominal mass on the side of abdomen. The lump was managed with radical excision and was diagnosed histopathologically as desmoid tumour.

In 1838 muller coined the term desmoids which means tendon-like in Greek. Desmoid tumours are rare myofibroblastic tumours that arise from aponeurosis of muscles. The tumours are well differentiated, benign in nature with almost no metastatic potential but can be locally aggressive. It accounts for 3 % of all soft tissue neoplasm. The tumours are twice as common in females as males. Desmoid tumours are more common in the reproductive age group indicating that hormones like oestrogen has an important role in genesis of tumour. The age group commonly affected is between 25 - 35 years. Desmoid tumour of abdomen is commonly accompanied by familial adenomatous polyposis.[1]

PRESENTATION OF CASE

Here we report a case of 65-year-old man who came to the surgery department with complaints of pain in abdomen and constipation for 5 days. He had no history of any trauma or surgery. The family history was not significant. On physical examination, a lump of 10 x 6 cm was palpated in lower abdomen. The lump was firm, non-tender with diffuse border. The lower border was not palpable.

Relevant blood investigations were done and were in the normal range. X-ray findings showed features suggestive of subacute intestinal obstruction.

MRI Findings

Revealed a large mass arising from the anterior lower abdominal wall muscles. Surrounding organs appear normal. No enlarged lymph nodes were detected. No ascites. Features were suggestive of desmoid tumour.

Operative Procedure

Radical excision of the tumour.

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Gross Examination

We received a globular mass with attached intestinal loop. The mass is irregular, haemorrhagic and brown in colour measuring 12 x 11.5 x 7 cm.

Microscopy

Section of cell blocks stained with haematoxylin and eosin show bundles of eosinophilic spindle cells. These cells are monomorphous, with regular nuclei and pale cytoplasm. Focal areas of haemorrhages are seen. Some areas also show congested capillaries.

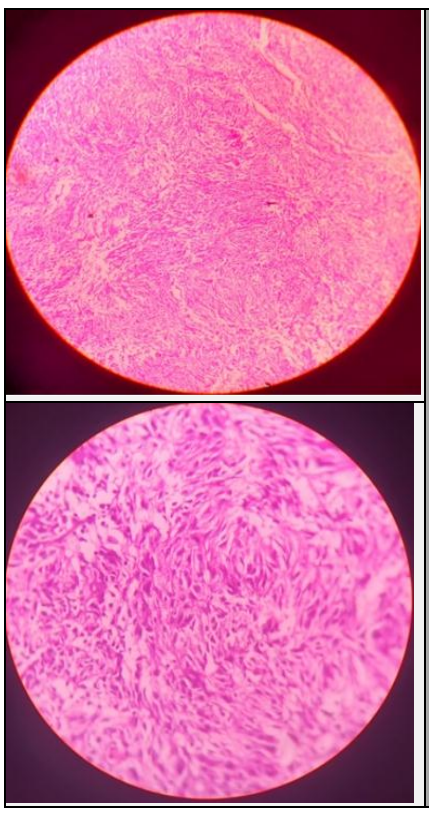

Figure 1. Desmoid Tumour (10x) Desmoid Tumour (40x)

DISCUSSION

Desmoid tumours are neoplasm of fibrous tissue arising from the fascia and aponeurosis of muscular structures anywhere in the body. Desmoid tumours are benign deep with a locally invasive behavior.[2] Nearly, 3.7 is the incidence per one million persons and has a female preponderance. The other factors responsible in development of the desmoids tumour are: familial adenomatous polyposis (FAP), hormonal therapy and infrequently with surgical trauma (1 in 4 cases).[3,4] Oestrogen may help in the tumorigenesis as it is more common in females of reproductive age group.[5] The most common cause of desmoid tumour is as a manifestation of FAP outside the colon. Broadly desmoid tumours can be classified into intrabdominal and extra-abdominal. The most common site affected extra-abdominally are the trunk and shoulder girdle, while intraabdominally mesentery and aponeurosis of anterior abdominal wall mostly the one arising from rectus abdominis and internal oblique are more common.

Desmoid tumour of abdomen is a firm mass that is commonly located in the upper anterolateral area. The margins of the mass are not well-defined with an indistinct capsule.[6] The treatment is complete surgical excision by local wide excision followed by reconstruction of the defect of extra-abdominal desmoid tumour. This was the treatment carried out in the reported case with no local recurrence for over a 6-month duration. Other treatment is chemotherapy, antioestrogen, radiotherapy that are used when the tumour is inoperable, local recurrences and in lesion which is not completely excised[1,2,3,4,7] Recurrence of tumour is reported to range from 19 % to 77 % of cases. The recurrence depends on the size of tumour and on the complete removal of tumour by surgical excision.[8]

Malignant conditions like fibrosarcoma, rhabdomyosarcoma, synoviosarcoma, liposarcoma, fibrous histiocytoma, lymphoma and metastases give intense enhancement. Benign conditions like neurofibroma, neuroma and leiomyomas don't show enhancement. Hematomas can be present at following location rectus sheath, chest wall, mesentery, retroperitoneum, and space of Retzius should be correlated with clinical history and will not show enhancement. Ultrasound evaluation of uterine leiomyoma in perimenopausal females was done by Daga et al.[9] Sharma et al. conducted a study on Gallium-68 DOTA-NOC positron emission tomography / computed tomography (PET / CT) as an alternate predictor of disease activity in sarcoidosis.[10] Different radio diagnostic procedures were used in swellings of abdominal region and adjacent structures including CT and MRI by Swarnkar et al.,[11,12] Talwar et al.,[13] Samad et al.,[14] Lamture et al.,[15,16] and Jindal et al.[17]

Desmoid tumour has to be differentiated from other anterior abdominal neoplasms like fibrosarcoma, rhabdomyosarcoma, neurofibroma, liposarcoma, lymphoma, leiomyosarcoma and other benign fibrous tumours and primitive neuroectodermal tumors.[7] As recurrence is not uncommon, a regular follow up is mandatory. The recommendation of National Comprehensive Cancer Network's Soft Tissue Sarcoma guideline is to keep a regular follow up of the patients after surgery. A thorough history and physical examination should be done followed by imaging investigation like CT scan or MRI in every 3 - 6 months for a period of 2 - 3 years followed by once every year.

CONCLUSIONS

Though it is a rare entity, desmoid tumours should be considered as a primary diagnosis when a patient complains of a painless abdominal mass on the anterior aspect of abdominal wall, especially in females of reproductive age group. The best treatment option is wide surgical resection with free margin. For better results, the abdominal defect after wide excision is reconstructed by utilization of prosthetics mesh.

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