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RECCURENT DESMOID TUMOUR OF THIGH – A RARE CASE REPORT

General Surgery

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ABSTRACT

Extra abdominal desmoid tumour is a rare entity. It is a locally aggressive tumour despite being histologically benign . To avoid local reccurence it is important to preoperatively detect the exact localization, extension, infiltration and dissemination of this tumour. We report a case of reccurent desmoid tumour of thigh which arose in the antero-lateral aspect of left thigh in a 46 years lady. This case is being presented as it's occurrence in thigh is very rare. The aim of this article is to assess the investigation of choice, modality of treatment, and the importance of adjuvant radiotherapy in recurrent desmoid tumour.

KEYWORDS

Desmoid tumour of thigh, Wide surgical excision.

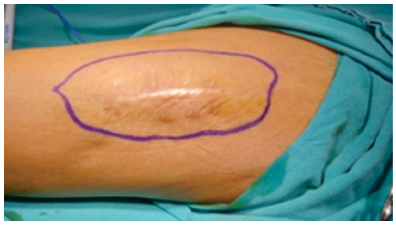
INTRODUCTION:

Desmoid tumors are rare neoplasms of uncertain etiology arising from fascial or deep musculo-aponeurotic structures. They are slightly more predominant in females and tend to occur during the third and fourth decades of life [1] . Etiological factors are trauma , history of local surgery, genetic factors like inherited mutation in APC gene (adenomatous polyposis coli) and high estrogenic states, including pregnancy [2]. Desmoid tumours are characterized by a benign histological appearance and no metastatic potential, and are locally aggressive tumors with a high rate of recurrence [3] . They may be localized in the abdominal wall, the bowel, and the mesentery (associated with familial adenomatous polyposis) or in extraabdominal sites, such as the trunk and the extremities. The incidence of desmoid tumor range from 2 to 4 per million [4]. They constitute 3% of all soft tissue tumours and 0.03% of all neoplasms.

CASE REPORT :

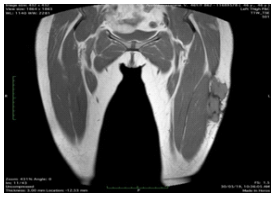
A 46 years lady was admitted in our hospital for a swelling on anterolateral aspect of the left thigh, gradually increasing in size since 3 years. She had a history of tumor excision in the same region, performed in another hospital 5 years back. General examination was unremarkable . The local physical examination showed a tumoral mass located on the anterolateral aspect of the left thigh, under an old 7 centimeters long scar. The tumor was 12x2cm , of firm consistency, painful, relatively fixed and seemed to infiltrate the subcutaneous tissue of the described area . Systemic examination was normal. There were no clinical features of metastasis .

1.Preoperative Image Of Left Thigh

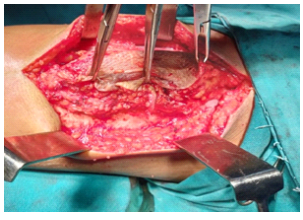


No motor, sensitivity or vascular alterations of the left lower limb were found. Routine investigations were normal. USG of left thigh showed a heterogenous hypoechoic lesion in subcutaneous plane of size 11.9x2.1cm and there was no evidence of intralesional calcifications, with minimum vascularity noted on Doppler study. MRI of left thigh revealed a multilobulated intensely enhancing soft tissue mass lesion of size 9.3x4.0cm involving subcutaneous fat plane without any infiltration into adjacent muscle planes , suggestive of desmoid tumour or any benign or malignant mesenchymal tumour. FNAC was suggestive of benign spindle cell neoplasm. Under spinal anesthesia a wide tumor excision was performed with macroscopic tumor free margins .The superficial part of the tumour was adherent to the skin at few places with puckering, and the deeper part of the tumour was adherent to the fascia covering the vastrus lateralis muscle.

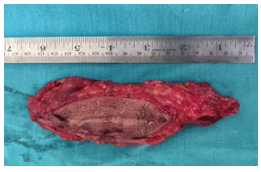
2. MRI Of Left Thigh (Coronal Section)



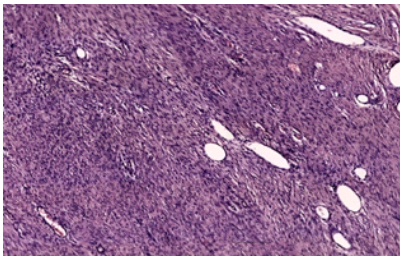
3. Intraoperative Image



4. Specimen



5. Histopathology



The postoperative period was uneventful, wound was healthy and the patient was discharged on the 12th day after suture removal. Histopathological examination of the specimen revealed skin with tumour in subcutaneous plane, consisting of fascicles of spindle shaped cells which are arranged in interlacing bundles. The cells are spindle shaped with spindle shaped nucleus and distinct nucleolus. Few cells are showing fibroblastic and few cells are showing myofibroplastic appearance.Mitotic activity is seen and is low[1/10HPF]. Focal areas are showing lymphocytic collections. The posterior margin also shows tumour, and features are suggestive of desmoid type fibromatosis. The patient had adjuvant radiotherapy of 2Gy per day for 5days in a week, for 6 weeks i.e a total of 60Gy in 30 fractions. The patient has completed the full course of radiation and is doing well, four months post surgery.

DISCUSSION :

Desmoid tumours also known as aggressive fibromatoses, are unique mesenchymal neoplasms that are often considered to be locally malignant but non metastasizing neoplasms. Specifically, these tumours are aggressive fibroblastic proliferation of well-circumscribed, locally invasive, and differentiated fibrous tissue.

Although USG shows the extent of the tumour and its relation to neurovascular structures , magnetic resonance imaging is the modality of choice for the diagnosis and the evaluation of the tumor extent and the progression of the desmoid tumour before and after treatment. It may also be helpful in differentiating tumour progression from postsurgical fibrosis [5]. To avoid local recurrence it is important to preoperatively detect the exact localization and extension of infiltrating lesion in this tumour.

Surgery is the treatment of choice for extra abdominal desmoid tumors. For primary treatment, wide excision with free tumor marginal resection is the gold standard. Reccurent desmoid tumour poses a challenge for the treatment options. Reexcision for treating the recurrent disease is preferred by most authors, resulting in a cure rate similar to that of the primary surgical resection . However, local control remains difficult. The recurrence rates after wide local excision is reported to be more than 40%, related to age, extra-abdominal localization of the tumor and section margins[6]. Desmoid tumours have a high rate of reccurence following even complete surgical removal. Recent meta analysis of 1,295 patients demonstrated that risk of recurrence with positive surgical margins was almost twofold higher [7]. More recently, Janssen et al. performed a meta analysis on the influence of surgical margin and adjuvant radiotherapy on local reccurence and found that radiotherapy reduces the risk of reccurence in patients with microscopically positive resection margins.The association of adjuvant radiotherapy was even stronger for resection of reccurent tumours with positive margins[7]. Radiation therapy with doses of 50–60 Gy is a viable alternative to surgery and a useful adjunct to incomplete resection of primary extra-abdominal desmoid tumors for the control of residual disease [8]. NSAIDS (sulindac or celecoxib), hormonal or biological agents (tamoxifen,toremifene or low dose interferon), chemotherapy(methotrexate , vinblastine and doxorubicin based regimens), Tyrosine kinase inhibitors(imatinib and sorafenib) are options for systemic therapy for patients with advanced or unresectable desmoid tumours [9]. The predisposition of aggressive desmoid tumours to locally recur is related to its infiltrative nature, the lack of pseudo capsule and possibility of diffusion along muscle fibres and fascial planes which makes it difficult for the surgeon to grossly identify the true extent of disease [10] .

CONCLUSION :

Desmoid tumour of thigh is a rare entity . Reccurence can be prevented with an aggressive multidisciplinary approach. MRI is the preferred investigative modality . A wide surgical resection with adjuvant radiation therapy remains the treatment option for the local control of the disease.

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