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A Rare Case Report of Fibromatosis (Desmoid Type-Staghorn vessel Pattern) - Anterior Chest Wall

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ABSTRACT

We report a case of desmoid fibromatosis of the anterior chest wall. Deep fibromatosis or desmoid tumour is a low-grade, soft tissue lesion that is notable for its local infiltration and most commonly occurs in the abdomen. Deep fibromatosis of the chest wall is a rare condition. A 17 year old male presented with an anterior chest wall swelling since 2 months, not associated with pain, gradually increasing in size with features of extension into skeletal muscle on radiology. On Fine Needle Aspiration Cytology, it was diagnosed as a spindle cell tumour and a wide local excision of the tumour was done.

Key words: Fibromatosis, Soft tissue tumour, Chest wall Swelling, Desmoid Tumour.

INTRODUCTION:

Deep fibromatosis or desmoid tumours are non-encapsulated, locally aggressive soft tissue tumours that could infiltrate the skeletal muscles and are myofibroblastic in origin. They are classified as deep (desmoid type) and superficial/ penile type. These are heterogenous benign tumours also called as aggressive fibromatosis. These rare tumours account for approximately 0.03- 0.1% of all solid tumours and 3.6% of all fibrous tissue neoplasms.[1, 2] Fibromatosis of the chest wall are rare tumors and only represent 10% to 20% of all fibromatosis. [3, 4]

Desmoid type fibromatosis can be further subdivided according to the site- extra abdominal, abdominal (abdominal wall) and intra-abdominal (deep pelvic and mesenteric) variants.

These are benign tumours with infiltrative borders and a propensity for local recurrence. They are characterised by proliferation of fibroblasts and myofibroblast type spindle cells, though the morphologic spectrum of this entity spans beyond the conventional histological pattern which could possibly result in misdiagnosis. These tumours could also erode bone and surrounding blood vessels and nerves.

CASE REPORT

A 17 year old male presented with a swelling on the left side of the anterior chest wall gradually progressive over 2 months. The swelling was not associated with pain. On examination, the swelling was non-tender, hard in consistency with normal skin over the swelling.

On Ultrasonography, 4.8 x 4.5 x 2.2 cm (Volume 25 cc), well defined heteroechoic, predominantly hypoechoic area was noted in the intramuscular plane on the left side of the chest wall with no evidence of vascularity.

FNAC demonstrated a spindle cell lesion with moderate cellularity comprising of spindle shaped cells with oval nuclei admixed with blood and stromal fragments. Subsequently excision of the swelling from the left anterior chest wall was performed for histopathological examination.

Gross examination: Received greyish-brown, soft to firm, non-

encapsulated mass with an ill-defined surface measuring 5 x 4 x 2.5 cm which on cut section showed grey-white homogenous areas.

Microscopic examination: The sections showed a non-encapsulated and ill-circumscribed lesion comprising of fibroblastic and myofibroblastic spindle cells arranged in sheets along with abundant vascular proliferation. Occasional floret type of giant cells and fibrocollagenous stroma was noted. Individual cells appeared elongated, oval with bland nuclei, bearing tapering ends and eosinophilic cytoplasm. Numerous blood vessels lined by flattened endothelial cells were seen. There was no necrosis or mitosis seen. The features were suggestive of desmoid fibromatosis arising from the anterior wall with extension into the skeletal muscle beneath.

The given biopsy also showed areas with thin-walled, branched blood vessels lined by a single layer of flat to ovoid endothelial cells that seemed hyperchromatic compared with the bland fibroblasts and myofibroblasts in the surrounding stroma. These features are suggestive of Staghorn Vessel Pattern of Desmoid- type fibromatosis.

For confirmation of diagnosis, Immunohistochemistry of tissue sections was done for S100, SMA, CD34 and Vimentin. Vimentin was strongly positive, smooth muscle antigen was focally positive, both CD34 and S100 were negative.
Immunohistochemical stains thus confirmed the diagnosis of Deep Fibromatosis (Desmoid type).

GROSS:

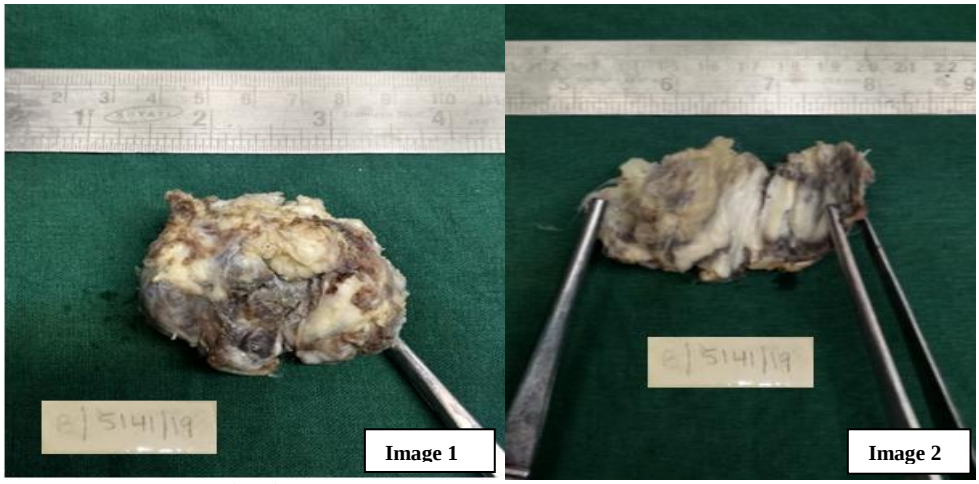


Image 1 and 2 show greyish-brown, soft to firm non-encapsulated tissue mass with irregular borders measuring 5x 3.5 x 2 cm which on cut section revealed grey-white homogenous areas.

FNAC

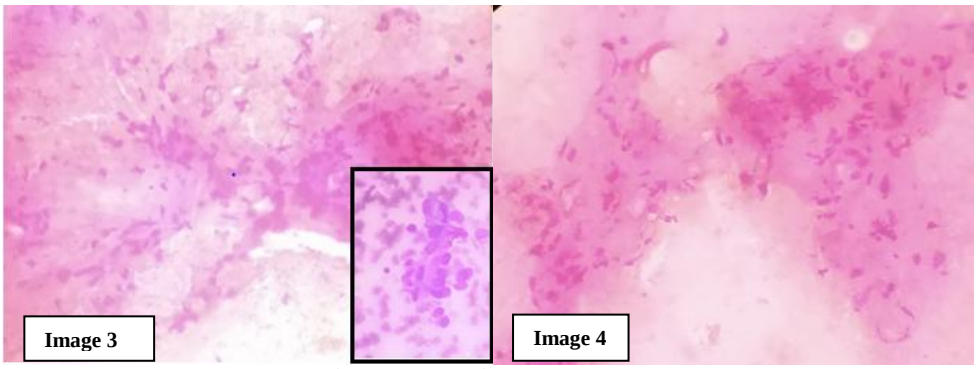
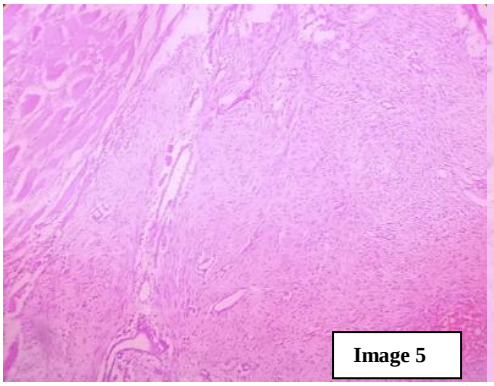


Image 3 and 4 show the conventional pattern of desmoid type fibromatosis on high power: The smears show moderate cellularity comprising of long sweeping fascicles of spindle cells that stretch across uniformly with oval nuclei mixed with blood and stromal fragments. Inset of image 3 shows a cluster of uniform plump spindle cells can be seen.



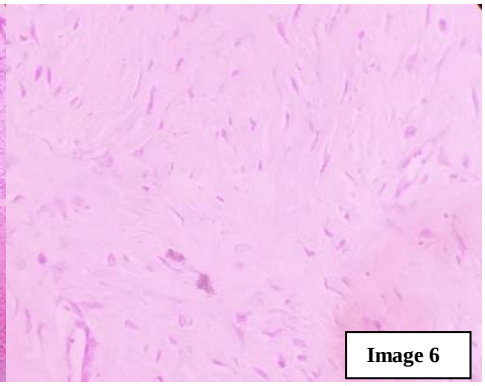
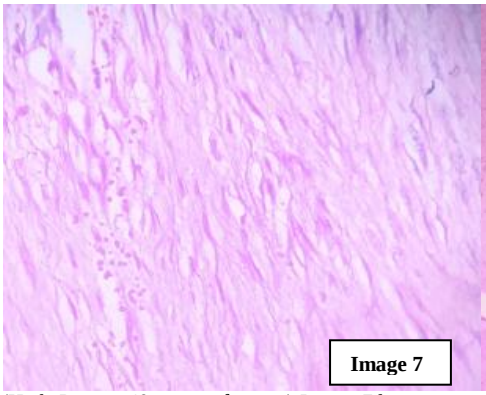


Image 5 and 6(10x magnification) :Image 5 depicts the extension of this tumour into the surrounding muscle fibres where infiltration can be seen. Image 6 shows fibroblast and myofibroblasts bearing pale bland oval to elongated nuclei with tapering ends. No evidence of pleomorphism, mitotic activity, necrosis or haemorrhage.



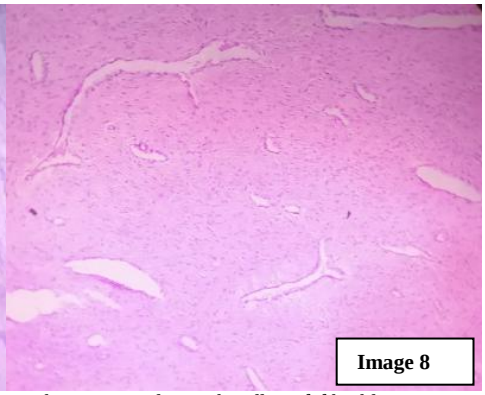


Image 7 & 8(High Power, 40x magnification):Image 7depicts section showing interlacing bundles of fibroblasts separated by variable amounts of collagen. In image 8, staghorn pattern of thin walled, branched blood vessels can be evidently seen.

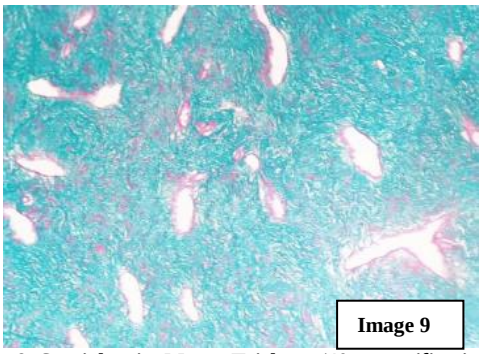


Image 9: Special stain, Mason Trichome(40x magnification).

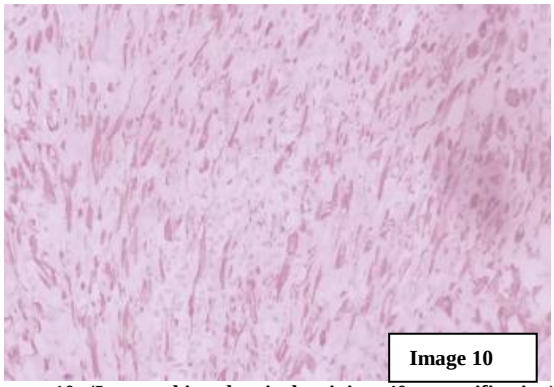


Image 10: (Immunohistochemical staining, 40x magnification): Section shows strong positivity for Vimentin.

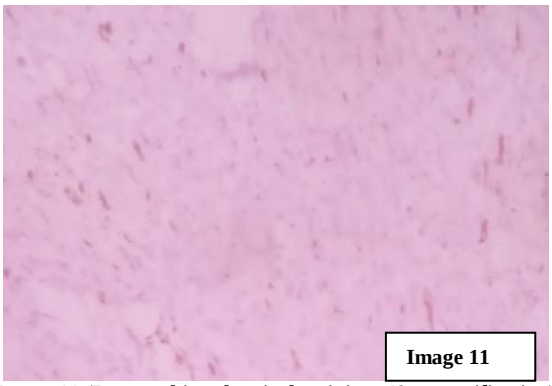


Image 11 (Immunohistochemical staining, 40x magnification): Section shows focal areas of positivity for Smooth Muscle antigen(SMA).

DISCUSSION

Deep Fibromatosis or Desmoid Tumours are locally aggressive tumours of myofibroblastic origin that lack metastatic potential. These should be differentiated from similar appearing, spindled-tumours that have metastatic potential, which is a difficult task. They are characterised by infiltrative pattern and a tendency towards local recurrence.

Fibromatosis can occur sporadically or in association with familial adenomatous polyposis (FAP). Incidence of sporadic type is 2–4 /million and 3.5–32% in FAP. Fibromatosis is classified into superficial (such as Dupuytren's, Ledderhose's and Peyronie's diseases) and deep known as desmoidtumour.[5]

Males and females of all ages are affected by desmoid tumors, but a propensity for fertile women has been noted by many authors.[1, 9] CTNNB1 and APC gene mutations are detected in approximately 89 % of the cases. Factors responsible could be a combination of the above mentioned genetic mutations, high estrogen states and canonical Wnt/ Beta catenin pathway activation due to antecedent trauma.[6]

Majority of the tumours, i.e. 51 to 67%, are intra-abdominal or in the abdominal wall. The most common extraabdominal sites are shoulder girdle, chest wall and inguinal region.

Deep fibromatosis has a range of cytological findings on FNA from low cellularity to moderate cellularity with bland spindled cells having bipolar nuclei and delicate, wispy cytoplasm. Stroma mostly appears metachromatic and focally very dense. It could appear loose with myxoid quality. [7]

Seven morphological patterns are known which include conventional pattern, hyalinised or hypocellular, staghorn vessel, myxoid, keloidal, nodular fasciitis-like and hypercellular. The most common pattern observed is the conventional pattern which consists of long sweeping fascicles composed of uniform and slender fibroblasts or myofibroblasts. Cells show pale stained nuclei with inconspicuous nucleoli without nuclear hyperchromasia or cytological atypia along with thin walled blood vessels and perivascular oedema. The second most common pattern is hyalinised pattern which shows decreased cellular density and increased amount of stromal hyalinization with widely spaced fibroblasts or myofibroblasts in dense eosinophilic matrix and compressed thin walled blood vessels.[8]

Staghorn vessel pattern is characterized by hemipericytoma like thin walled, branched vessels which have a single layer lining of flat-to-ovoid hyperchromatic endothelial cells with bland fibroblasts/ myofibroblasts in the stroma. Myxoid pattern shows spindle cells which are loosely arranged in myxoid matrix and absent thin walled blood vessels.

In keloidal pattern, the key feature is bright eosinophilic glassy collagen fiber bands along with bland spindle cells. Nodular Fasciitis- like pattern shows loosely arranged spindle to stellate-shaped cells and stromal haemorrhage. Hypercellular pattern is the least common pattern consisting of lesional cells arranged in long fascicles with increased cellular density.

Keloidal and staghorn pattern is more prevalent in intra-abdominal lesions compared to abdominal wall and extraabdominal sites. [8]

A retrospective study of 142 patients by Fiore *et al*. suggested that larger tumors and tumors located on the trunk were associated with a higher risk of recurrence. Prevention of recurrence is dependent on factors such as tumor location, ability to obtain negative margins, and adjuvant therapy.[1, 10]

The differential diagnosis of desmoid tumours include benign reactive conditions such as scar and nodular fasciitis, and other mesenchymal neoplasms, such as low-grade fibrosarcoma and malignant nerve sheath tumour.[5]

CONCLUSION

This case was diagnosed as Desmoid Fibromatosis- Staghorn vessel pattern with infiltration into the surrounding skeletal muscles and blood vessels. The clinical history in patients with suspected deep fibromatosis is vital. Due to the infiltrative pattern, the radiological impression for theses tumours could be suggestive of malignancy. The cytomorphology is not very specific resulting in descriptive

diagnosis and hence excision biopsy and immunohistochemical stains confirm the diagnosis. A recommended panel for similar appearing spindle cell tumours must include CD 34, S 100, vimentin and SMA. Other markers like Beta catenin, desmin and c kit work up can also be done. On histopathology the morphological spectrum of desmoid- type fibromatosis is broad and awareness of the various histologic patterns is crucial for accurate diagnosis.

The treatment of choice is wide local excision with clear margins. This is difficult as there could be infiltration into adjacent structures such as muscles, blood vessels and nerves due to aggressive nature of the tumour. Adjuvant therapy should be considered for positive margins. Follow up should be advised to check recurrence.

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