

Extra-abdominal desmoid type fibromatosis in the infra clavicular region (pectoralis major muscle): a rare case report

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Abstract: Desmoid tumours are rare mesenchymal tumor, accounting for 0.03% of all neoplasms. The deep fibromatoses are classified into extra-abdominal, abdominal, and intra-abdominal. Extra-abdominal fibromatosis is a rare entity of desmoids tumor. Muscles of the shoulder, pelvic girdles and the thigh are involved most commonly. Here we report a case of extra-abdominal desmoid tumor in infra clavicular region in young female patient.

Keywords: Desmoid tumor, infra clavicular fibromatosis, pectoralis major muscle, extra-abdominal.

INTRODUCTION

Desmoid tumors (aggressive fibromatosis) are an extremely rare entity of mesenchymal tumor. Although these tumors are slow growing and histologically benign, but at various anatomic sites show local invasion. Desmoid tumors are most commonly intra abdominal and arise from the aponeurosis of the rectus abdominis muscle. The extra-abdominal form of desmoid tumor is very rare and most common sites are shoulder, chest wall, back, thigh, and head/neck [1, 2].

Because of their resemblance to vascular neoplasms, diagnosis of desmoid tumors poses a clinically challenging problem. Generally, histopathological examination is required for definitive diagnosis. Surgery, radiotherapy, or both are regarded as the standard treatment options for desmoid tumors [3].

CASE REPORT

A 17 years old girl attended orthopedic outpatient department with complaints of swelling of size 8x5 cms in left infra clavicular region for two month duration.(fig.1) Swelling was painless and gradually progressing in size. Swelling was palpable separately from left breast tissue and was adherent to deeper tissue. There was no history of preceding trauma. She was prescribed antibiotic and analgesics for a short period of time.

Basic hematological and biochemical tests including pulmonary functions tests were within normal limits. Chest X-ray finding were in normal limit except scoliosis. USG of chest swelling revealed a hypo echoic mass with underlying irregularity of bone. CECT advised for better characterization of the lesion. CECT thorax showed poorly enhanced soft tissue mass lesion posterior to left pectoralis major muscle. Lesion insinuate into adjacent intercostals space. Underlying lung field appeared normal. Focal destructive lesions are seen involving various vertebral bodies. No mediastinal lymphadenopathy was noticed.

Repeated FNAC of the chest swelling yielded only blood. Biopsy of the swelling showed muscle bundles, fibrocollagenous and fibro adipose tissue revealing infiltration by mononuclear cells with few groups and fascicles of oval to spindle shaped cells with patent vessel (Fig 2& 3). These cells were separated by abundant collagen zed stroma. (Fig 4) Periphery of the lesion showed advancing edge of the lesion. In this particular field, the tumor is not encapsulated and is surrounded by intense inflammatory reaction with infiltration into adjacent fat.(Fig.5) VG stained section revealing dense collagenous stroma.(Fig.6) On immuno histo chemistry, these tumor cells were negative for desmin, S-100 and positive for vimentin and SMA. On these histological and immuno histo chemistry finding, a possibility of mesenchymal tumor extra-abdominal desmoid tumor was suggested. The patient was under observation.



Fig. 1: Clinical photograph of infra clavicular swelling of size 8x5 cms

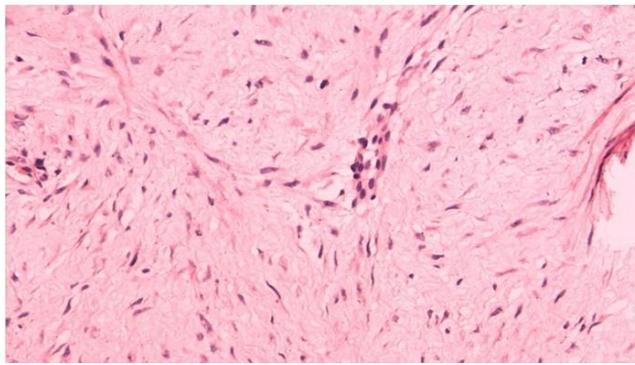


Fig. 2: H&E section showed few groups and fascicles of oval to spindle shaped cells

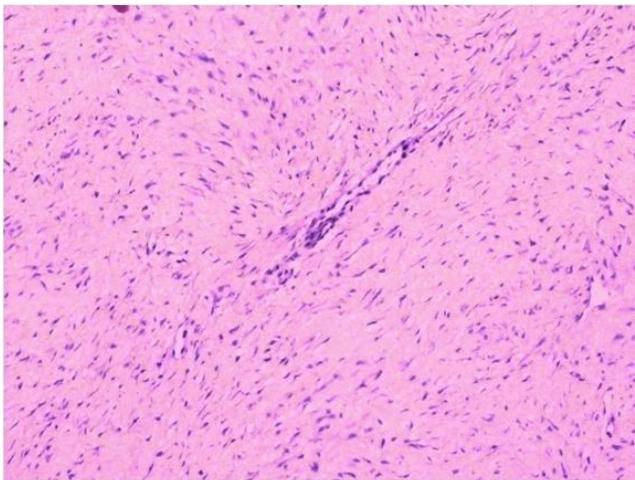


Fig. 3: H&E section showed few groups and fascicles of oval to spindle shaped cells with patent vessel

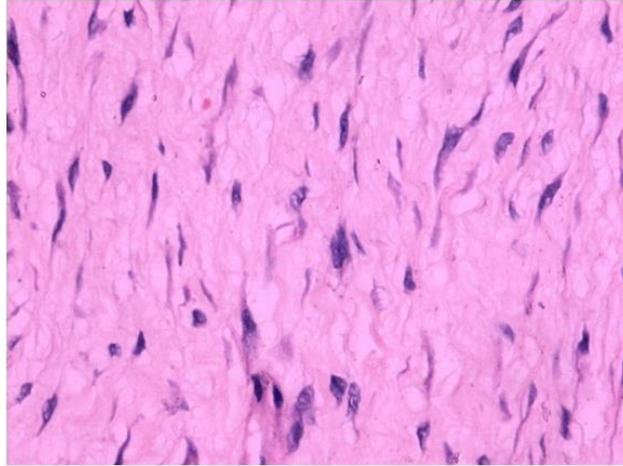


Fig. 4: H&E section showed spindle cells were separated by abundant collagenized stroma

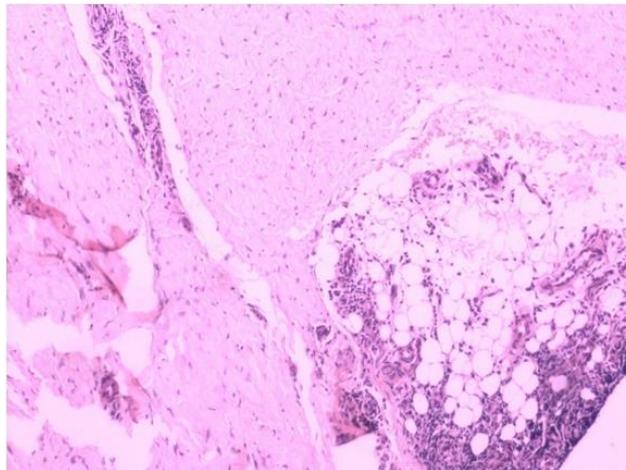


Fig. 5: Periphery of the lesion was surrounded by intense inflammatory reaction with infiltration into adjacent fat.

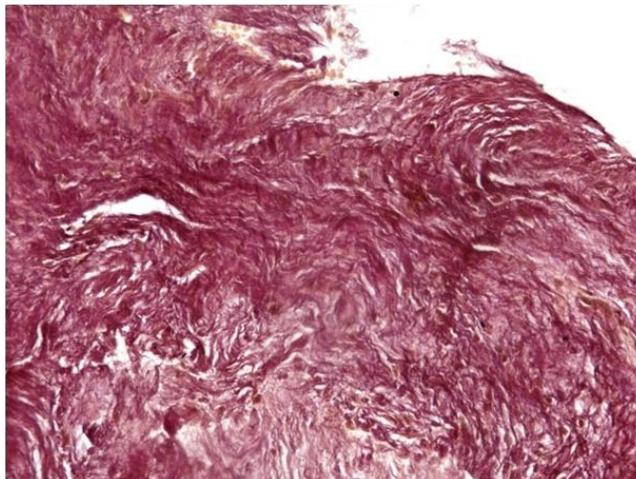


Fig. 6: VG stained section revealing dense collagenous stroma.

DISCUSSION

Desmoid tumours are rare mesenchymal tumor, accounting for 0.03% of all neoplasms. Estimated incidence rate of desmoid tumor is 24 per

million per year. The term “desmoid” describe the hard, tendon like appearance of the tumor. Masson and Soule believed the term “desmoid tumor” applied for fibrous tissue proliferation with locally infiltrative growth that

may appear in various locations and does not metastasize.4 Extra-abdominal desmoid tumor first recognized by Nichols in 1923 [5].

Fibromatoses are a group of benign fibrous tissue proliferations which show infiltrative growth pattern and a tendency to recur locally without metastasis. The two major class are superficial (fascial) and deep (musculo aponeurotic). The deep fibromatoses are classified into extra-abdominal, abdominal, and intra-abdominal. Extra-abdominal desmoid fibromatosis arises from various soft tissues including the connective tissue of muscles and the overlying fascia or aponeurosis. Muscles of the shoulder, pelvic girdles and the thigh of adolescents and young adults are involved most commonly [6].

Extra-abdominal fibromatosis is a rare entity of desmoids tumor. Etiopathogenesis of extra-abdominal desmoids type fibromatosis is not well known, but genetics, hormonal status, and prior trauma have been associated in most of cases. Considering their rarity, desmoid tumors are often misdiagnosed preoperatively. The differential diagnosis of desmoids tumor includes vascular and soft tissue tumors such as fibrosarcomas or neurofibromas [5, 7].

Desmoid tumor most commonly occurs between puberty and age 40 years. Women are more affected than men. The tumors are more aggressive in younger patients as compared to older. . Multicentric origins of these tumors are also identified in 10-15% of cases [8, 9].

Grossly extra-abdominal desmoids tumors are usually non encapsulated gray white tissue in close proximity of musculature and overlying fascia or aponeurosis. Cut sections are firm and glistening white and resemble scar tissue. Microscopically these tumors are composed of alternating bundles of mono morphic elongated, spindle shaped fibroblast and myo fibroblast which are embedded in collagenous stroma. Mitotic activity and overall cellularity are low. Necrosis and hemorrhage are absent. Immuno histo chemically, the cells are usually actin positive and CD34 and S100 negative. Masson's trichrome stain highlight the collagen fibers interlaced between these tumor cells [10].

Surgery and radiation therapy, either alone or in combination, are treatment of choice for extra-abdominal desmoids. Surgery also provides excellent local control for recurrent desmoids tumor [9]. In our case, surgical excision was done and clinical follow up was advised.

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