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Case Report

Desmoplastic Fibroblastoma (Collagenous Fibroma) of Thigh- A Rare Case Report

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Abstract: Desmoplastic fibroblastoma (collagenous fibroma) is a rare benign fibrous tumour thought to arise from subcutaneous tissue or skeletal muscle. We herein report a case of elderly female presented with a slow growing non tender mass on anterior aspect of right thigh. Radiological investigation showed a mass in intermuscular plane. Lumpectomy was performed, histopathologic examination of the subsequent specimen confirmed the diagnosis of desmoplastic fibroblastoma.
Keywords: Desmoplastic fibroblastoma, collagenous fibroma, benign tumour, histopathologic examination

INTRODUCTION

Desmoplastic fibroblastoma, also known as collagenous fibroma (CF) is a benign fibroblastic/ myofibroblastic tumour, typically occurs in the subcutaneous tissue or skeletal muscle in adults [1]. Evans was the first to introduce the term desmoplatic fibroblastoma, in 1995, a unique fibrous soft tissue tumour comprising spindle-shaped to stellate fibroblastic cells sparsely distributed in a dense fibrous background [2, 3]. It presents as a painless, firm, well-circumscribed mass of long standing-duration and behaves in a benign fashion. There is no evidence of bone involvement except for a rare case with surface erosion [4].

Case report

A 60-year-old previously healthy female presented with 1-year history of slow growing lump with dull aching pain at lower part of right thigh. She had no history of trauma. Physical examination revealed a mobile, irregularly round, firm palpable mass deeply seated in the quadriceps muscle on anterior aspect of lower thigh. Mass was not associated with tenderness or pathological change in the overlying skin.

A plain radiograph of knee joint showed features of osteoarthritis but no evidence of soft tissue calcification was seen. Ultrasonography showed an avascular homogenous hyperechoic soft tissue mass measuring 5x4cm located above the knee joint, close to joint capsule in subfascial plane below the rectus femoris muscle. Ultrasonography guided FNA was performed but yielded scanty material composed of occasional fragments of fibrocollagenous stromal material and was suspicious of benign spindle cell lesion. Excision biopsy was done and sent for histopathological examination.

A globular, circumscribed, unencapsulated grey white soft tissue mass m/s 4.5x3.5x2.5cm was received covered with fibrofatty tissue. Cut section was pearly grey in color, lobulated with firm cartilage like consistency (Figure 1). Section studied with hematoxylin & eosin stain showed well circumscribed lesional tissue rimmed by fatty tissue and skeletal muscle fibres. Lesional tissue composed of benign-appearing spindle-shaped and stellate tumour cells with bland nuclei dispersed in predominantly fibrous, hyalinized collagenous stroma and focal myxocollagenous stroma (Figure 2). Tumour is intersected by fibrous septa and interspersed with inconspicuous thin walled blood vessels. No evidence of necrosis, nuclear atypia or abnormal mitosis. Final diagnosis of desmoplastic fibroblastoma (collagenous fibroma) was rendered.

Fig. 1: Cut section of mass showing pearly grey lobulated appearance

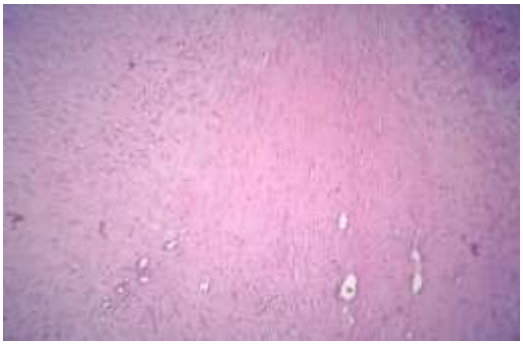

Fig. 2: Tumour demonstrating low cellularity composed of bland spindle, stellate cells dispersed in fibrous and myxocollagenous stroma

DISCUSSION

Soft tissue tumours are relatively common, but frequently represent a diagnostic problem for pathologists due to the similarity in histological aspects and an overlap in the staining profile among cells with fibroblastic and myofibroblastic differentiation. In addition, collagen-rich variants of nonfibroblastic or myofibroblastic neoplasms may demonstrate a similar profile that masks their true nature [5].

Collagenous fibroma is a rare benign tumour with less than, 100 cases reported in the English literature [6].Clinically, collagenous fibroma presents as firm, well circumscribed, subcutaneous or intramuscular, painless mass of long duration [7]. These tumours show predilection for male patients (5:1) and have a peak incidence in the fifth and sixth decades of life. The lesion has been reported in the arm (24%), shoulder (19%), posterior neck or upper back (14%), abdominal wall and hip joints (6%) [5, 8, 9]. Most tumours were located in the subcutaneous tissue or in skeletal muscle [9-11].The tumours range in size from 1 cm to 20 cm [7]. Collagenous fibroma typically infiltrates subcutaneous fat and skeletal muscle, and this has been observed in up to 51% of cases [8, 9].

Grossly, the tumour can be oval, fusiform or disc shaped with homogenous pearl gray colour on cut section [7, 8].

Microscopically, the lesion is usually nodular and partially surrounded by a fibrous pseudocapsule. It is hypocellular and composed by fibroblast/myofibroblast cells dispersed in a collagen rich stroma. These cells vary from spindle to stellate-shaped, often binucleated with large elongated to oval nuclei. The nuclei exhibit delicate chromatin, and small eosinophilic nucleoli may be present. Inflammatory cells, mitotic figures, areas of necrosis, metaplastic bone formation and calcification are not seen [2]. Adjacent skeletal muscle and fat tissue involvement is seen in 51% of the cases [7, 8].

The differential diagnosis of collagenous fibroma include neurofibroma, fibromatosis, nodular fasciitis, fibroma of the tendon sheath, solitary fibrous tumour, perineurioma, sclerotic fibroma of the skin, calcifying fibrous pseudotumour and myxoma [7].

CF diagnosis is practically morphologic, since immunohistochemistry investigation is not very elucidative. The cells are negative for S100 protein and strongly positive for vimentin and variable positivity for factor XIIIa and SMA [2].

IHC aids when doubt exists. S100 protein, CD34 and EMA are helpful in excluding neurofibroma, solitary fibrous tumour and perineurioma respectively [7].

The origin of CF is still unclear. It has been suggested that CF might originate from a chromosomal rearrangement (11q12) similar to the fibroma of the tendon sheath [2, 12]. However, it is still unclear if this lesion is a reactive process or a true neoplasm [2].

Treatment of desmoplastic fibroblastoma is surgical excision with no reported incidence of local recurrence or metastases [13].

Our case was a patient of 60 years, presenting with a slow growing mass in the thigh for duration of 1 year. The history, clinical, gross and microscopic findings of our case is in concordance with the literature. The discordant features include the presence of dull aching pain that can be attributed to the size of the tumour, the patient being a 60 year old lady and the tumour in the subfascial plane near the knee joint capsule.

CONCLUSION

Desmoplastic fibroblastoma (collagenous fibroma) is a rare benign tumour of fibroblastic differentiation. It can pose diagnostic dilemma for pathologists due to its histological similarity with other soft tissue tumours of fibroblastic/myofibroblastic differentiation. Careful microscopic evaluation helps in distinguishing it from other locally aggressive tumour like desmoid tumour.

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