DESMOPLASTIC FIBROBLASTOMA (COLLAGENOUS FIBROMA): REPORT OF A RARE CASE IN A FEMALE

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INTRODUCTION: Desmoplastic fibroma is a rare benign subcutaneous tumour that most commonly affects men. The term 'Desmoplastic fibroblastoma' was first coined by Evans in 1995 and was renamed "Collagenous fibroma" by Nielsen in 1996.[1] A unique fibrous soft tissue tumor comprising of spindle to stellate-shaped fibroblastic cells sparsely distributed in dense fibrous background.[2]

Here with we are reporting a case of Desmoplastic fibroma in the nape of the neck in a middle aged female.

CASE REPORT: A 40 year old female presented with history of painless swelling in the nape of the neck since 1 year. She had no history of trauma. Physical examination revealed a mobile, firm, nodular palpable mass. Excision biopsy was done under local anesthesia and sent for histopathological examination.

Gross appearance: Specimen consists of single firm globular pearly white mass of tissue measuring 4 x 3 x 1.5 cm. Cut section revealed diffuse, firm, pearly white tissue. (Fig. 1)

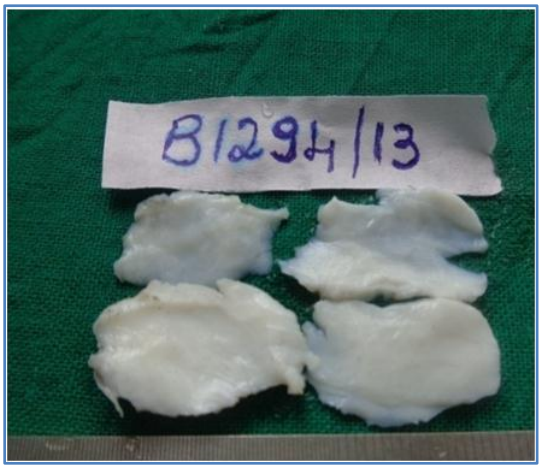

Fig. 1: Gross appearance Desmoplastic Fibroblastoma

Microscopic appearance: Section studied showed a benign neoplastic lesion comprising of hypocellular dense collagenous tissue containing widely spaced bland spindle to stellate shaped cells. Inflammatory cells, mitotic figures, areas of necrosis, metaplastic bone formation and calcification are absent. (Fig. 2 & 3).

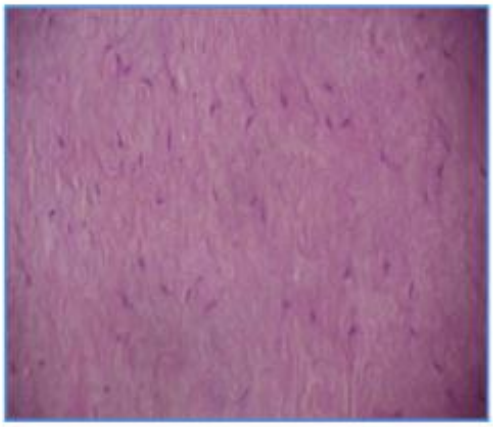

Fig. 2: Microphotograph of Desmoplastic fibroblastoma, H&E, X10

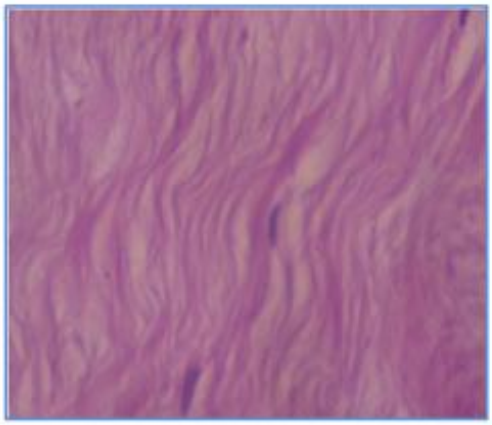

Fig. 3: Microphotograph of Desmoplastic fibroblastoma, H&E, X40

Special stains were done to demonstrate the collagenous nature of tumour. Van Gieson Stain showed collagen staining red and nuclei staining black (Fig. 4). Masson Trichrome stain showed collagen staining blue and nuclei staining black. (Fig. 5).

Fig. 4: van Gieson Stain-collagen: red, nucleus: black, X40

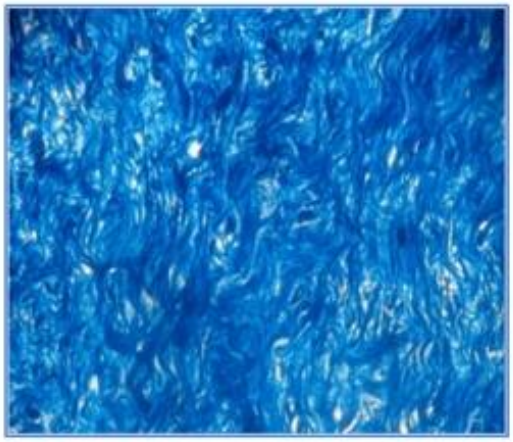

Fig. 5: Masson Trichrome stain- collagen: blue, nucleus: black, X40:: Stain

DISCUSSION: Desmoplastic fibroma is a rare benign tumour with less than 100 cases reported in the English literature. These tumours show predilection for male patients (5:1) and have peak incidence in fifth and sixth decades of life.[2] The lesion has been reported in the arm, shoulder,

nape of the neck, upper part of the back, abdominal wall and hip joints. It is a recently defined benign fibroblastic tumour.[3] The present case is a female and the lesion was present in the nape of neck.

Diagnosis of Desmoplastic fibroblastoma is purely based on morphologic features since immunohistochemistry is not very elucidative. In the present case, histological appearance was typical of Desmoplastic fibroblastoma and special stains demonstrated collagenous nature of the tumour.

Most patients have been treated by conservative simple excision and neither local recurrence nor metastasis has been reported. The present case was treated with surgical excision and there is no recurrence of the lesion even after 1 year of follow-up.

Differential diagnosis[4]: Differential diagnosis for Desmoplastic Fibroblastoma include Desmoid fibromatosis, Fibroma of tendon sheath, Calcifying fibrous tumor, Hyalinized variant of Nodular fasciitis and Low grade fibromyxoid sarcoma.

Desmoid fibromatosis shows Mild to moderate cellularity, moderate vascularity and extensive infiltration of muscle by spindle cells which are usually arranged in fascicular pattern. Desmoplastic Fibroblastoma is paucicellular, hypovascular and may show focal muscle infiltration by spindle and stellate cells which are randomly arranged.

Fibroma of Tendon Sheath occurs most commonly on hands or wrist, is multilobular and is moderately vascular where as Desmoplastic Fibroblastoma shows wide distribution, occurs as a single nodule and is hypovascular.

Calcifying Fibrous Psuedotumor shows small spindled cells, chronic inflammation and **c**alcification. In Desmoplastic Fibroblastoma, the cells are large, stellate shaped and do not show any inflammation or calcification.

In Hyalinized variant of Nodular fasciitis, cellularity is moderate areas, mitotic figures may be numerous and cystic mucin pools are seen. Desmoplastic Fibroblastoma is paucicellular, mitotic figures are absent or rare and mucin pools are not seen.

Low grade fibromyxoid sarcoma is moderately cellular, shows whorled pattern and myxoid areas. Desmoplastic Fibroblastoma is paucicellular, do not show whirling and myxoid areas are absent.

CONCLUSION: Desmoplastic fibroblastoma is a rare benign tumour of fibroblastic differentiation. Careful microscopic evaluation helps in distinguishing it from other soft tissue tumours of fibroblastic\myofibroblastic differentiation including the more aggressive desmoid tumour and low grade fibromyxoid sarcoma. Special stains help to demonstrate collagenous nature of the tumour.

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