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Editorial

Ostensible and Wiry-Superficial CD34+ Fibroblastic Tumour

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Superficial CD34+ fibroblastic tumour is a distinctive, low grade tumefaction arising within superficial sites as diverse cutaneous surfaces or subcutaneous tissue. Superficial CD34+ fibroblastic tumour is additionally designated as PRDM10 rearranged soft tissue tumour. Characteristically, fascicular proliferation of spindle shaped cells pervaded with abundant, eosinophilic, granular to glassy cytoplasm is encountered. Although nuclear pleomorphism may be significant, mitotic activity is minimal. Tumour cells comprehensively appear immune reactive to CD34. Besides, focal immune reactivity to keratin (AE1/AE3) is encountered within ~ 70% neoplasms.

Of obscure aetiology, majority of neoplasms appear within middle aged adults with an age range of disease emergence between 20 years to 76 years and median age of disease occurrence at 38 years. A mild male preponderance is observed [1,2].

Superficial CD34+ fibroblastic tumour may delineate genomic rearrangements within PRDM10 gene or chromosomal translocation t(2;5)(q31;q31) [1,2].

Neoplasm commonly incriminates lower limbs, especially thighs although no site of disease emergence is exempt [1,2].

Characteristically, neoplasm represents as a gradually progressive, enlarging, firm, painless tumefaction confined to superficial soft tissues or various cutaneous zones [1,2].

Upon gross examination, a firm, yellow to tan soft tissue neoplasm of magnitude varying from one centimetre to 10 centimetres is observed. Cut surface is variably gelatinous [1,2].

Upon frozen section, a low grade neoplasm comprised of spindle shaped cells is encountered. Constituent spindle shaped cells delineate cellular and nuclear pleomorphism. Mitotic activity is minimal [1,2].

Cytological assessment demonstrates a cellular specimen predominantly comprised of clusters and singularly dispersed spindle shaped cells intermingled with fragments of collagen rich stroma. Innumerable spindle shaped cells appear permeated with tapered nuclei and elongated cytoplasmic processes. Spindle shaped cellular component is commingled with a smattering of enlarged, pleomorphic cells imbued with enlarged, plump or bizarre nuclei with irregular nuclear contour [1,2].

Upon microscopy, tumefaction is confined to dermis or subcutaneous adipose tissue. The well circumscribed neoplasm is constituted of fascicles or sheets of spindle shaped tumour cells [2,3].

Neoplastic spindle shaped cells or epithelioid cells are pervaded with abundant, eosinophilic, granular or glassy cytoplasm, pleomorphic, hyperchromatic nuclei, intra-nuclear cytoplasmic pseudo-inclusions and variably prominent nucleoli. A population of tumour cells demonstrate xanthomatous alterations [2,3].

Mitotic activity is minimal. Tumour necrosis is uncommon.

Neoplasm is infiltrated by an admixture of chronic inflammatory cells as small lymphocytes, plasma cells, mast cells and innumerable eosinophils.

Arborizing capillaries and vascular articulations are frequently admixed within the neoplastic component [2,3].

Upon ultrastructural examination, spindle shaped cells or elliptical cells appear devoid of intercellular junctions and

are permeated with irregular or convoluted nuclei, abundant euchromatin and prominent nucleoli. Besides, abundant rough endoplasmic reticulum, mitochondria, lysosomes, ribosomal aggregates and aggregated lipid globules may be discerned [2,3].

Superficial CD34+ fibroblastic tumour appears immune reactive to CD34 and INI1/SMARCB1. Focal reactivity to cytokeratin and nuclear reactivity to PRDM10 is encountered [4,5].

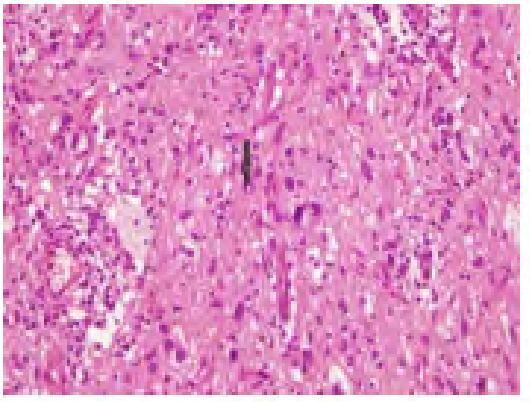

Figure 1: Superficial CD34+ fibroblastic tumour delineating fascicles of spindle shaped cells with abundant, eosinophilic cytoplasm and pleomorphic hyperchromatic nuclei intermingled with arborizing capillaries and an inflammatory infiltrate of mast cells, lymphocytes, plasma cells and numerous eosinophils [6].

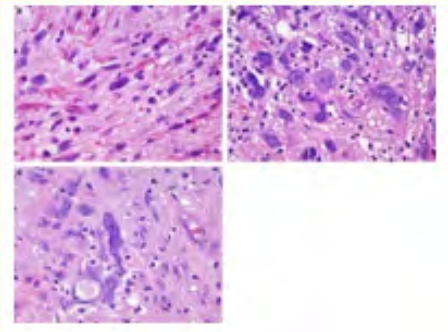

Figure 2: Superficial CD34+ fibroblastic tumour enunciating sheets of spindle shaped cells with abundant, eosinophilic cytoplasm and hyperchromatic, pleomorphic nuclei intermingled with arborizing vascular articulations and an inflammatory exudate comprised of mast cells, lymphocytes, plasma cells and innumerable eosinophils [7].

Superficial CD34+ fibroblastic tumour appears immune non reactive to desmin, α smooth muscle actin, S100 protein, ERG, FLI1, CD99, anaplastic lymphoma kinase 1(ALK1), BCL2 and p53.

Upon fluorescent in situ hybridization(FISH), neoplasm is devoid of TGFBR3 or MGEA5 genetic loci [4,5].

Superficial CD34+ fibroblastic tumour requires segregation from neoplasms such as myxoinflammatory fibroblastic sarcoma, myxofibrosarcoma, dermatofibrosarcoma protuberans, undifferentiated pleomorphic sarcoma, epithelioid sarcoma, pleomorphic hyalinising angietactic tumour, malignant granular cell tumour, solitary fibrous tumour or low grade fibromyxoid sarcoma [4,5].

Superficial CD34+ fibroblastic tumour can be optimally discerned with cogent tissue sampling followed by precise morphological assessment. Computerized tomography (CT) or magnetic resonance imaging (MRI) depicts a well circumscribed tumefaction devoid of calcification. Tumour appears preponderantly confined to subcutaneous adipose tissue.

Upon 2-(^{18}F) fluoro-2-deoxy-D-glucose (^{18}F-FDG) positron emission tomography, neoplasm depicts an abnormal contrast uptake.

Superficial CD34+ fibroblastic tumour can be appropriately treated with comprehensive surgical excision of the neoplasm [4,5].

Tumefaction represents with indolent clinical behaviour. Majority of instances appear to lack localized disease reoccurrence. However, regional lymph node metastases following inadequate surgical extermination of the primary neoplasm is documented [4,5].

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6. Image 1 Courtesy: Wiley online library.

7. Image 2 Courtesy: Nature.com.