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Coadunation and Immixture-EWSR1: SMA3 Fibroblastic Tumour

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Editorial

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EWSR1::SMAD3 fibroblastic tumour is an extremely exceptional, contemporary, benign mesenchymal neoplasm of fibroblastic origin demonstrating varied morphological patterns, characteristic genetic fusions and localized tumour aggression. Tumefaction is predominantly situated upon extremities and exhibits an intense predilection for acral segments. Characteristic diagnostic features appear as a miniature neoplasm confined to acral cutaneous or subcutaneous tissue demonstrating a zonal configuration with hypo-cellular or acellular, hyalinised centric zone and intersecting fascicles of monomorphic, peripheral spindle shaped cells which appear immune reactive to ERG. EWSR1::SMAD3 genetic fusion is observed which assists cogent tumour classification. Neoplasm manifests a sporadic disease emergence.

Additionally designated EWSR1::SMAD3 positive fibroblastic tumour, neoplasm is characteristically comprised of dual components of peripheral, hyper-cellular foci of spindle shaped fibroblasts commingled with centric acellular foci of hyalinised tumour. Tumefaction expresses a diffuse, intense immune reactivity to ERG, a feature which may be employed for excluding pertinent, challenging differential diagnosis.

The infrequently discerned EWSR1:SMAD3 associated fibroblastic tumour exemplifies a wide range of disease emergence from infancy up to 7th decade. A female predominance is observed with female to male proportion at 4:1 [1,2].

EWSR1::SMAD3 associated fibroblastic tumour predominantly incriminates extremities, especially acral segments.

Of obscure aetiology, EWSR1:: SMAD3 associated fibroblastic tumour is predominantly triggered by cogent genetic fusion. Characteristically, neoplasm demonstrates a repetitive EWSR1::SMAD3 genetic fusion. Besides, exon 7 of EWSR1 gene confined to chromosome 22q12.2 exhibits fusion to exon 5 or exon 6 of SMAD3 gene situated upon chromosome 15q22.33 [1,2]. Tumefaction appears as a miniature, painless, superficial nodule or gradually progressive neoplasm confined to acral sites of upper limb or lower limb as hands or feet. Tumour emerges within dermal cutaneous zones or subcutaneous tissue and exhibits localized tissue infiltration. Tumour reoccurrence frequently ensues due to infiltrative pattern of neoplastic growth. Neoplasm may reappear within several years following initial surgical excision. Malignant metamorphosis remains undocumented [2,3]. Currently, cytological features remain undocumented.

Grossly, a miniature, firm, nodular, greyish white tumefaction is encountered. Generally, tumour magnitude varies from 1 centimetre to 2 centimetres. Upon microscopy, dermal or subcutaneous neoplasm may about the epidermis. Tumefaction manifests a nodular, mildly lobulated or plexiform configuration with an infiltrative pattern of neoplastic evolution. Occasionally, neoplasm appears to engulf circumscribing subcutaneous adipose tissue [2,3]. Characteristic microscopic features manifest as an acellular, hyalinised centric zone encompassed within a hyper-cellular peripheral zone delineating intersecting cellular fascicles of bland, spindle shaped cells. Cellular and nuclear pleomorphism, atypia and hyperchromatic nuclei appear absent. Mitotic figures are negligible. Foci of stippled, dystrophic calcification may ensue. Few neoplasms exhibit distinct zones with centric hyalinization and cellular peripheral zone. Focal stippled calcification within hyalinised areas is documented [2,3].

Tumefaction is comprised of dual components as hyper-cellular foci of spindle shaped fibroblasts gradually blending with foci of acellular hyalinised tissue (Figures 1&2). Spindle shaped fibroblastic cells configure intersecting, well defined, hyper-cellular fascicles. Uniform spindle shaped cells are incorporated with moderate, eosinophilic cytoplasm and uniform, elongated, wavy nuclei. Neoplastic cells appear devoid of cellular or nuclear pelomorphism or atypia. Mitotic activity is negligible. Upon ultrastructural examination, morphological features remain undefined [3,4].

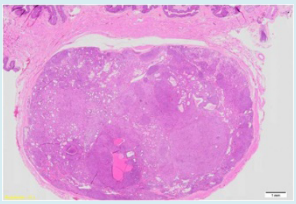

Figure 1: EWSR1::SMAD3 fibroblastic tumour delineating a lobulated neoplasm composed of an acellular, hyalinised centric zone surrounded by peripheral fascicles of spindle shaped cells. Cellular atypia, pleomorphism and mitotic activity is negligible [5,6].

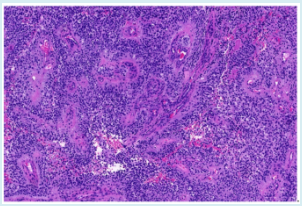

Figure 2: EWSR1::SMAD3 fibroblastic tumours demonstrating a nodular neoplasm composed of hyalinised centric area circumscribed by peripheral fascicles of uniform spindle shaped cells lacking pleomorphism or atypia [7].

EWSR1::SMAD3 associated fibroblastic tumour appears immune reactive to ERG or SATB2. Besides, weak immune reactivity to smooth muscle actin (SMA), keratins or epithelial membrane antigen (EMA) may be encountered. EWSR1::SMAD3 associated fibroblastic tumour appears immune non-reactive to factor XIIIa, muscle specific actin(MSA), desmin, caldesmon, S100 protein, SOX10, CD34, human melanoma black 45(HMB45) antigen, MUC4, pan-TRK, synaptophysin, chromogranin, CD17 or DOG1 [4,5]. EWSR1::SMAD3 associated fibroblastic tumour requires segregation from neoplasms such as cellular fibrous histiocytoma, myofibroma, lipofibromatosis, calcifying aponeurotic fibroma, acral fibromyxoma, dermal nerve sheath myxoma, myxofibrosarcoma, lipofibromatosis-like neural tumour, fibromatosis, monophasic synovial sarcoma or cutaneous variants of myoepithelioma [4,5]. EWSR1:SMAD3 fibroblastic tumour can be appropriately discerned with cogent evaluation of factors such as site of tumour emergence, clinical manifestations, morphological features, immunohistochemistry and molecular characteristics. A hyper-cellular tumefaction confined to distal extremities composed of monomorphic population of uniform, spindle shaped, fibroblastic cells intermingled with centric, acellular hyalinised areas appears indicative of EWSR1::SMAD3 associated fibroblastic tumour. Mitotic activity is minimal. Tumour cells depict a diffuse, intense nuclear immune reactivity to ERG. Instances challenging to discern may be confirmed by detection of typical EWSR1 genetic rearrangements [4,5]. Upon ultrasonography, a subcutaneous, hypoechoic nodule is encountered. Magnetic resonance imaging (MRI) enunciates miniature, well demarcated tumour nodules. T1 weighted magnetic resonance imaging exhibits hypo-intense signal intensity. T2 weighted magnetic resonance imaging exemplifies a hyper-intense signal intensity. EWSR1:SMAD3 fibroblastic tumour may be appropriately alleviated with comprehensive surgical extermination of the neoplasm. Tumour reoccurrence may ensue with inadequate surgical resection and appears contingent to adequacy of resected surgical perimeter [4,5].

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