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Editorial

Rugged and Ropy-Nodular Fasciitis Salivary Gland

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Nodular fasciitis configures as a benign, self limiting neoplasm of fibroblastic or myofibroblastic derivation. Tumefaction delineates comprehensive and repetitive fusion genes wherein the MYH9::USP6 genetic fusion is commonly observed although diverse partner genes are enunciated with USP6 gene.

Additionally designated as cranial fasciitis, nodular fasciitis may concur with entities as fibroma of tendon sheath, myositis ossificans, cranial fasciitis or aneurysmal bone cyst.

The nomenclature of pseudosarcomatous fasciitis is considered as obsolete.

Tumefaction expounds significant age range of disease emergence and an extensive anatomic distribution.

Nodular fasciitis emerges as a common mesenchymal neoplasm preponderantly implicating young adults although no age of disease emergence is exempt. A definitive gender predilection is absent [1,2].

Nodular fasciitis is commonly confined to sites such as upper and lower extremities, head and neck or trunk although no site of disease emergence is exempt. Notwithstanding, lesions confined to intravascular, intra-articular or intra-parotid sites may be discerned, in addition to cranium or placenta. Frequently, neoplasm originates from the subcutis although may arise within the dermis, fascia or skeletal muscle [1,2].

Of obscure aetiology and sporadic occurrence, neoplasm was pre-emptively considered to be of traumatic origin [2,3].

Comprehensive instances of nodular fasciitis are pervaded with fusions genes wherein MYH9::USP6 is a commonly detected genetic fusion product. Besides, several and diverse genes may partner with chromosomal rearrangements within USP6 gene [2,3].

Clinically, an asymptomatic, tender or minimally painful, miniature neoplasm < 3 centimetre diameter is encountered. However, tumefaction may be enlarged. Neoplasm is rapidly progressive, a feature which clinically indicates a malignant neoplasm.

Exceptionally, a malignant tumefaction, as detected upon clinical or morphological grounds may be exemplified [2,3].

Grossly, neoplasm is solitary with soft to firm consistency and grey to whitish, tan or light pink hue. Tumour periphery may be circumscribed or infiltrative. Characteristically, tumour magnitude appears < 3 centimetres. However, tumour diameter of up to 7 centimetres may be exceptionally encountered. Cut surface is glistening [3,4].

Upon microscopy, a variably cellular neoplasm demonstrates spindle shaped or stellate cells enmeshed within a myxoid to collagenous extracellular matrix. Tumour cells are impregnated with uniform, bland, elliptical nuclei. Tumour parenchyma frequently depicts disseminated chronic inflammatory cells as lymphocytes, histiocytes or osteoclast-like giant cells [3,4].

Neoplasm configures a loose, fascicular or storiform pattern, designated as 'tissue culture-like' or 'feathery' pattern of neoplastic evolution. Lesions of extended duration may be significantly collagenous. Foci of cystic degeneration may be discerned. Mitotic figures are significant although atypical mitotic figures appear absent [4,5].

Exceptionally, certain neoplasms delineate malignant morphological features as cellular or nuclear pleomorphism or atypical mitotic figures. Aforesaid variants may be contemplated as nodular fasciitis with molecular concurrence of genetic rearrangements of USP6 gene [4,5]. Ultrastructural examination depicts morphological features characteristic of fibroblasts. Besides, peripheral, longitudinal, myofilaments and hemidesmosome-like structures may be discerned [4,5].

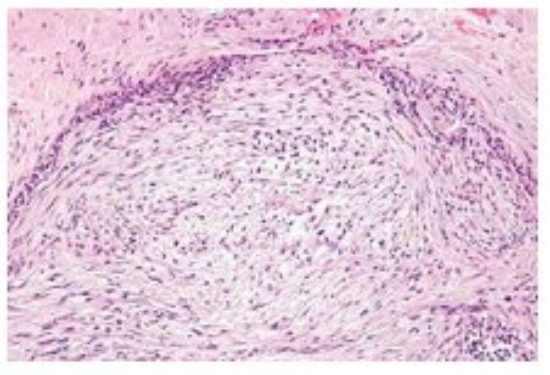

Figure 1: Nodular fasciitis demonstrating fascicles of spindle shaped cells with uniform, elliptical nuclei configuring a storiform pattern. Surrounding stroma is myxoid and infiltrated by chronic inflammatory cells. Focal mitotic figures are seen [11].

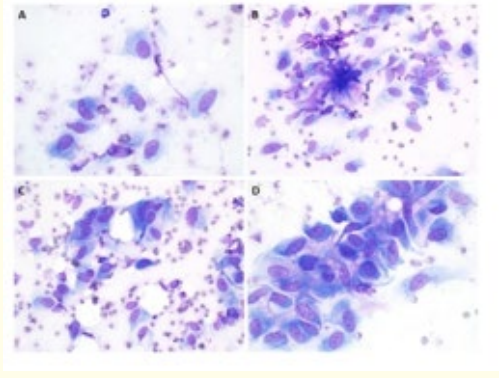

Figure 2: Nodular fasciitis delineating aggregates of spindle shaped cells impregnated with bland, elliptical nuclei. Foci of myxoid stromal cells appear intermingled with spindle cellular component. Red cell extravasation is encountered [12].

Benign Epithelial Tumours	Malignant Epithelial Tumours
Pleomorphic adenoma	Mucoepidermoid carcinoma
Basal cell adenoma	Adenoid cystic carcinoma
Warthin tumour	Acinic cell carcinoma
Oncocytoma	Secretory carcinoma
Salivary gland myoepithelioma	Micro-secretory adenocarcinoma
Canalicular adenoma	Polymorphous adenocarcinoma
Cystadenoma of salivary gland	Hyalinising clear cell carcinoma
Ductal papilloma	Basal cell adenocarcinoma
Sialadenoma papilliferum	Intra-ductal carcinoma
Lymphadenoma	Salivary duct carcinoma
Sebaceous adenoma	Myoepithelial carcinoma
Intercalated duct adenoma and hyperplasia	Epithelial-myoepithelial carcinoma
Striated duct adenoma	Mucinous adenocarcinoma
Sclerosing polycystic adenoma	Sclerosing micro-cystic adenocarcinoma
Keratocystoma	Carcinoma ex pleomorphic adenoma
Sialolipoma(mesenchymal tumour of salivary gland)	Carcinosarcoma of salivary glands
	Sebaceous adenocarcinoma
	Lympho-epithelial carcinoma
	Squamous cell carcinoma
	Sialoblastoma
	Salivary carcinoma (NOS) and emerging entities

Table 1: WHO classification of salivary gland tumors(5th edition) [6].

Nodular fasciitis of the salivary gland appears immune reactive to smooth muscle actin (SMA), muscle specific actin (MSA) and calponin. Tumour cells appear immune non reactive to desmin, H-caldesmon, S100 protein, SOX10, CD34, ERG, epithelial membrane antigen (EMA) or keratin [6,7].

Nodular fasciitis of salivary gland requires segregation from neoplasms as dermatofibrosarcoma protuberans, desmoid type fibromatosis or fibrous histicytoma [7,8].

The distinctive neoplasm demonstrates characteristic morphology contingent to haematoxylin and eosin stain of formalin

fixed paraffin embedded sections and is amenable to appropriate classification. Upon immunohistochemistry, tumour cells expound myofibroblastic differentiation [7,8].

Nodular fasciitis of salivary gland predominantly depicts MYH9::USP6 fusion genes within constituent cells. Besides, genomic rearrangements within USP6 gene may be ascertained by fluorescent *in situ* hybridization (FISH), polymerase chain reaction (PCR) or next generation sequencing (NGS) methodologies [8,9].

Upon radiography, non specific imaging features are discerned.

Upon magnetic resonance imaging (MRI), manifestations are nonspecific and cogent segregation from sarcoma may be challenging [8,9].

Nodular fasciitis of salivary gland may be appropriately alleviated by simple surgical extermination of the neoplasm. Occasionally, the lesion retrogresses spontaneously or following surgical tissue sampling. Tumour reoccurrence may exceptionally ensue with inadequate surgical eradication [9,10].

The pre-eminently benign nodular fasciitis is accompanied by superior prognostic outcomes [9,10].

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11. Image 1 Courtesy: Wikipedia.
12. Image 2 Courtesy: American Cancer Society.