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The Sinewy Scruff-Nuchal-Type Fibroma

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Abstract

Nuchal-type fibroma is an uncommon, benign, tumour-like, subcutaneous nodule comprised of bundles of thick collagen fibres confined to dermis and subcutaneous tissue of posterior neck. The condition commonly appears within third to fifth decade and demonstrates a male preponderance. Generally, an asymptomatic, superficial, gradually progressive, solitary or multiple, protrusive, firm to hard, solid, inadequately circumscribed tumefaction is denominated. The pauci-cellular neoplasm is composed of collagen fibres, mature adipose tissue and occasional skeletal muscle fibres with infiltration into surrounding cutaneous adnexal articulations.

Keywords

Squamous Cell Carcinoma; Scalp; Surgery

Preface

Nuchal-type fibroma is an uncommon, benign, tumour-like clinical entity accompanied by a distinct, subcutaneous and dermal proliferation of fibrous tissue. Accumulation of bundles of thick collagen fibres within the posterior neck is a characteristic feature of nuchal-type fibroma. The neoplasm is additionally nomenclated as collagenosis nuchae, nuchal fibrocartilaginous pseudo-tumour or nuchal fibroma. Nuchal-type fibroma was initially described by Enzinger

and Weiss in 1988 [1]. Nuchal-type fibroma commonly arises as a solitary, firm to hard, poorly circumscribed, subcutaneous nodule confined to posterior neck. The neoplasm is an exceptional, benign, fibrous, tumour-like proliferation characteristically displaying dense, hypo-cellular bundles of collagen intermingled with sparse fibroblasts, mature adipose tissue, enmeshed cutaneous adnexal structures and peripheral nerve fibres. Entrapment of cutaneous adnexal structures, mature adipose tissue and peripheral nerve fibres within collagen bundles are denominated as categorical histologic findings. The neoplasm reoccurs when subjected to inadequate surgical extirpation. Extra-nuchal neoplasms are morphologically indistinguishable from lesions arising from nuchal region. Thus, Michal et al., in 1999 proposed the terminology of “nuchal-type fibroma” which comprehensively denominates neoplasms which histologically simulate nuchal-type fibroma, irrespective of site of origin [2].

Disease Characteristics

Nuchal-type fibroma predominantly emerges within cutaneous and subcutaneous tissue of nuchal region defined by nape or posterior neck, thus, engendering the terminology of “nuchal-type fibroma”. The principally benign neoplasm arises predominantly within the dermis and subcutaneous tissue of posterior neck (70%), upper back or associated domains. Approximately, one third (33%) of the nodules may appear within extra-nuchal sites such as upper and lower back, buttocks, extremities, lumbosacral, scapular or facial region, shoulder, axilla, forearm, torso, anterior neck, knee, ankle or scalp. An estimated 50% neoplasms are associated with diabetes mellitus. Gardner’s syndrome or diabetes scleredema and aforementioned conditions demonstrate the occurrence of morphologically similar lesions [3,4]. Extra-nuchal-type fibroma is predominantly disseminated within the shoulder, face, inter-scapular or paraspinous region. Majority of neoplasms are solitary although multiple lesions on upper back or posterior neck are observed [3]. Nuchal-type fibroma typically arises between third to fifth decades with a mean age of 40 years although tumefaction can appear within 3 years to 74 years [4]. The condition demonstrates a male preponderance, a majority (> 80%) instances appear in males with a male to female ratio of 4:1. Nevertheless, male predominance is not observed in Gardner’s syndrome-associated fibromas [4]. Aetiology of nuchal-type fibroma remains obscure and warrants extensive elucidation. Nuchal-type fibroma can concur with trauma, diabetes mellitus or Gardner’s syndrome. Nuchal-type fibroma and Gardner’s-associated fibroma appear identical on histology and a subset of nuchal-type fibromas associated with Gardner’s syndrome can occur at multiple, diverse or uncommon locations. Gardner’s syndrome-associated fibromas may appear within children, a condition where emergence of fibroma is indicative of a sentinel event and mandates further genetic analysis. Gardner’s syndrome-associated fibromas are predominantly extra-nuchal in location and morphologically identical to nuchal-type fibroma [3,4].

Clinical Elucidation

Nuchal-type fibroma characteristically manifests an asymptomatic, superficial, gradually progressive, solitary or multiple, protrusive, firm to hard, solid, inadequately circumscribed tumefaction confined to the dermis or subcutaneous tissue. Tumour magnitude is variable and mean tumour diameter is around 3.5 centimetres although enlarged neoplasms of nearly 20 centimetres are denominated. The enlarged, palpable tumour can be visible beneath an unremarkable, superimposed cutaneous surface [3,4]. On account of the rarity of nuchal-type fibroma, proportionate tumour mortality remains unascertained [4].

Histological Elucidation

The neoplasm delineates a poorly circumscribed, un-encapsulated, firm to hard, greyish-white subcutaneous nodule generally beneath < 8 centimetre magnitude, with a minimal tumour diameter of ≤ 3 centimetres. Extra-nuchal lesions appearing at diverse sites are morphologically similar to nuchal-type fibromas [5,6]. The pauci-cellular, subcutaneous tumefaction is composed of collagen fibres, mature adipose tissue and occasional skeletal muscle fibres. Neoplasm infiltrates dermal tissue surrounding cutaneous adnexal articulations. Sparse, fibroblast-like cells are disseminated within dense, irregularly aligned fascicles of collagen admixed with scattered, fine elastic fibres. A scant infiltrate of chronic inflammatory cells, predominantly lymphocytes, is exemplified. Nerve bundles are quantifiably enhanced and enmeshed within fibrous tissue along with zones of mature adipose tissue cells [5,6]. Typically, the hypo-cellular neoplasm displays a haphazard dissemination of dense collagen fascicles admixed with entrapped, mature adipose tissue, paucity of elastin and enmeshed miniature, peripheral nerve fibres [5]. As the pauci-cellular nuchal-type fibroma is characterized by bundles of collagen intermixed with mature adipocytes and enhanced quantities of miniature nerve fibres, significant foci of mucoid tissue degeneration may ensue within the neoplasm [6]. The un-encapsulated, hypo-cellular neoplasm is constituted by thickened collagen fibres intermixed with delicate elastic fibres. Entrapped mature adipose tissue and enmeshed peripheral nerves, akin to a traumatic neuroma, can be discerned. Tumour tissue can infiltrate abutting skeletal muscle and adipose tissue. Scattered lymphocytes can imbue the tumefaction [5,6]. Majority of nuchal-type fibromas are constituted by haphazard dissemination of thick, dense collagen fibres. In addition, entrapped zones of mature adipose tissue and skeletal muscle are observed. Certain lesions may exhibit enlarged, peripheral nerves fibres with accompanying perineurial fibrosis [5,6].

Immune Histochemical Elucidation

Nuchal-type fibroma is immune reactive to vimentin, CD34 and CD99. Entrapped neural element is immune reactive to S100 protein. Spindle-shaped cells are immune reactive to CD34 and CD99. The neoplasm is immune non-reactive to Smooth Muscle Actin (SMA) and desmin [3,4].

Differential Diagnosis

Nuchal-type fibroma can be misinterpreted on account of an indolent clinical course and histological simulation to adjunctive, benign fibrous tissue neoplasms. Nuchal-type fibroma requires a segregation from

- Elastofibroma is a neoplasm predominantly situated within the subscapular region which is frequently bilateral and displays a prominence of aberrant elastic fibres
- Fibro-lipoma is a circumscribed neoplasm and appears in diverse locales
- Fibromatosis is a tumefaction arising in deep-seated soft tissue, in contrast to the nuchal region. The extensively cellular neoplasm displays broad fascicles of mature fibroblasts
- Gardner's syndrome- associated fibroma depicts histological features simulating a nuchal-type fibroma although lesions are principally extra nuchal. The conditions may be analogous or configure a similar entity
- Solitary fibrous tumour is extensively cellular, demonstrates a characteristic "pattern- less" tumour configuration and staghorn type of vascular articulations [3,4]

Nuchal-type fibroma requires a segregation from fibrous tissue tumours and tumour-like conditions such as desmoid-type fibromatosis, circumscribed storiform collagenoma, connective tissue nevus, collagenous fibroma, fibroma of tendon sheath, scleredema or scar tissue. In contrast to aforesaid neoplasms, nuchal-type fibroma is significantly paucicellular and depicts poor circumscription with entrapped, mature adipose tissue [3,4]. Nuchal-type fibroma requires a demarcation from histologically identical, benign fibrous tissue tumours such as dermatofibrosarcoma protuberans, giant cell fibroblastoma, mammary and extramammary myofibroblastoma or spindle cell and pleomorphic lipoma (Fig. 1-7) [3,4].

Figure 1: Nuchal-type fibroma delineating bundles of collagen admixed with nerve bundles, scarce fibroblasts and lymphocytes and few elastic fibres.

Figure 2: Nuchal-type fibroma demonstrating collagen fascicles admixed with mature adipose tissue, elastic fibres and innumerable nerve fibres.

Figure 3: Nuchal-type fibroma composed of fascicles of collagen, mature adipose tissue cells, elastic fibres, fibroblasts and few lymphocytes.

Figure 4: Nuchal-type fibroma comprised of collagen bundles, mature adipose tissue, numerous lymphocytes, miniature nerve bundles and minimal fibroblasts.

Figure 5: Nuchal-type fibroma delineating collagen bundles admixed with a prominent component of mature adipose tissue cells, fibroblasts, miniature nerve bundles and disseminated lymphocytes.

Figure 6: Nuchal-type fibroma depicting aggregates of collagen intermixed with miniature nerve bundles, few lymphocytes, fibroblasts and elastic fibres.

Figure 7: Nuchal-type fibroma where the spindle-shaped cellular component is immune reactive to CD34.

Investigative Assay

Intra operative frozen section is beneficial and can be adopted for cogent tumour categorization. Accurate intraoperative tumour discernment is crucial. Although asymptomatic, nuchal-type fibroma emerging as a subcutaneous nodule can be precisely ascertained with cogent imaging techniques [7,8]. On ultrasonography, the lesion can be misinterpreted as a lipoma. Magnetic Resonance Imaging (MRI) is a preferred imaging modality for discerning nuchal-type fibroma. The modality demonstrates a superior resolution of soft tissues with multi-planar efficacy. Nuchal-type fibroma is hyper-intense upon T1 weighted imaging and T2 weighted imaging. Magnetic Resonance Imaging (MRI) delineates a tumefaction with mixed signal intensity upon T1 weighted imaging and T2 weighted imaging. Signal intensity can be enhanced, variable and non-unified. Enhanced signal intensity is associated with a strip- like pattern upon T2 weighted imaging and zones of elevated signal intensity are significantly enlarged, in contrast to T1 weighted imaging [7,8]. Upon adoption of Magnetic Resonance Imaging (MRI) as a preferred diagnostic modality, nuchal-type fibroma displays a tumefaction with a minimal, mild or mixed signal intensity upon T1 weighted and T2 weighted imaging. Also, enhanced signal intensity within a nuchal- type fibroma is indicative of mucoid degeneration [7,8].

Therapeutic Options

Surgical extirpation of the neoplasm with a wide, tumour-free perimeter is a preferred treatment strategy. Marginal surgical excision is optimal along with removal of a significant segment of surrounding perimeter of normal tissue [(7,8)]. Comprehensive surgical excision with extermination of circumscribing segment of healthy tissue is suitable for treating nuchal-type fibroma. As the tumour is poorly demarcated, adequate tumour localization and definition of tumour periphery is essential [7]. However, as the neoplasm is un-encapsulated, comprehensive surgical resection can be challenging and may contribute to a propensity for localized tumour reoccurrence. Alternatively, tumour reoccurrence may arise due to persistence of factors which initiate the incurrence of nuchal-type fibroma such as repetitive trauma. Tumour relapse is observed with incomplete surgical excision wherein a singular neoplasm can reoccur several times. Proportion of tumour reappearance is enhanced with inadequate tumour extermination. The neoplasm is devoid of metastasis to distant sites [7,8].

Conclusion

Nuchal-type fibroma is immune reactive to vimentin, CD34 and CD99 and immune non-reactive to Smooth Muscle Actin (SMA) and desmin. Nuchal-type fibroma requires a segregation from elastofibroma, fibro-lipoma, fibromatosis, Gardner's syndrome- associated fibroma, solitary fibrous tumour, desmoid- type fibromatosis, circumscribed storiform collagenoma, connective tissue nevus, collagenous fibroma, fibroma of tendon sheath, scleredema, scar tissue or histologically identical, benign fibrous tissue tumours such as

dermatofibrosarcoma protuberans, giant cell fibroblastoma, mammary and extra- mammary myofibroblastoma or spindle cell and pleomorphic lipoma. Magnetic Resonance Imaging (MRI) is a preferential imaging modality for discerning nuchal-type fibroma and the neoplasm appears hyper-intense upon T1 weighted imaging and T2 weighted imaging. Surgical extirpation of the neoplasm with a wide, tumour-free perimeter is a preferred treatment strategy.

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