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Case Report

Understanding Nuchal Type Fibroma: A Rare Case Report with Comprehensive Discussion

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Abstract: ***Introduction and Aim***: Nuchal-type fibroma (NTF) is a rare benign fibrous proliferation primarily originating from the dorsocervical neck region. Enzinger and Weiss first characterized it in 1988, highlighting its distinct histopathological features. ***Description of the Case Report*:** We present a case of 71-year-old South Asian male patient with painless swelling over the nape of neck, highlighting the challenges in diagnosis of NTF. Clinical examination, imaging, and cytological findings initially suggested a lipomatous lesion. Histopathological evaluation revealed characteristic features consistent with NTF, emphasizing the importance of histopathological assessment for accurate diagnosis. Despite its benign nature, vigilance is essential due to the potential for recurrence. ***Conclusion*:** This case underscores the need for a comprehensive understanding of NTF's clinical and histopathological features to facilitate optimal management and improve patient outcomes.

Keywords: Nuchal-type fibroma, posterior neck, benign fibrous proliferation, collagen bundles.

INTRODUCTION

Nuchal-type fibroma (NTF) was initially described as originating from the dorsocervical neck region [1, 2]. Enzinger and Weiss introduced it as a distinct entity in 1988, describing it as a rare benign fibrous proliferation typified by hypocellular bundles of collagen interspersed with fatty tissue.3 Association with conditions such as diabetes mellitus, scleroderma, and Gardner's syndrome has been seen, suggesting potential underlying pathophysiological mechanisms [2, 4]. Additionally, trauma has been proposed as another possible cause [4, 5]. While the majority of NTF cases are found in the posterior neck region, reports of occurrences in extra-nuchal sites have been rarely reported [6]. This diversity in presentation underscores the need for a comprehensive understanding of NTF's clinical features, histopathological characteristics, and associated conditions to facilitate accurate diagnosis and optimal management.

CASE REPORT

Here, we present a case of 71-year-old South Asian male patient who presented to the General Surgery outpatient department with painless swelling over the nape of neck for the past two years which gradually increased in size without any associated neurological deficits. There was no significant past or present history and routine investigations were within normal limits. Clinical examination revealed a firm, non-tender swelling measuring approximately 1.7x1.0 cm on the left side of the posterior neck. Importantly, there was no fixation to the underlying structures or the overlying skin, suggesting a benign etiology. Ultrasonography of the neck confirmed the presence of a well-defined, ovoid hypoechoic lesion measuring approximately 1.7x1.0 cm in the subcutaneous plane of the left neck region. The patient was then sent for fine needle aspiration. The aspiration was difficult and scant aspirate was obtained. Cytological findings included few fat fragments with interlacing capillaries, so a diagnosis of lipomatous lesion was suggested and an excisional biopsy was requested for the confirmation of the diagnosis. Surgical excision of the mass was performed via a transverse neck

incision, and the specimen was subjected to histopathological evaluation. Gross examination revealed a well-defined, but non-encapsulated swelling measuring approximately 1.7x1.0 cm with a grey-white, firm to hard cut surface (Figure 1). Microscopic examination demonstrated hypocellular, thick bundles of collagen with interspersed blood vessels and scattered lymphocytes (Figures 2A & B). Notably, attached adipose tissue appeared unremarkable. The absence of diabetes, polyposis of the colon, or family history of Gardner's syndrome further supported the diagnosis of isolated NTF. The histopathological features observed in our case align with the characteristic findings of NTFs, consisting predominantly of collagenous stroma with varying degrees of vascularity and inflammatory infiltrate. The post-operative course was uneventful without any complications and a follow-up examination at one month revealed satisfactory healing progress. The patient is currently under follow-up to monitor for any signs of recurrence or other related complications.

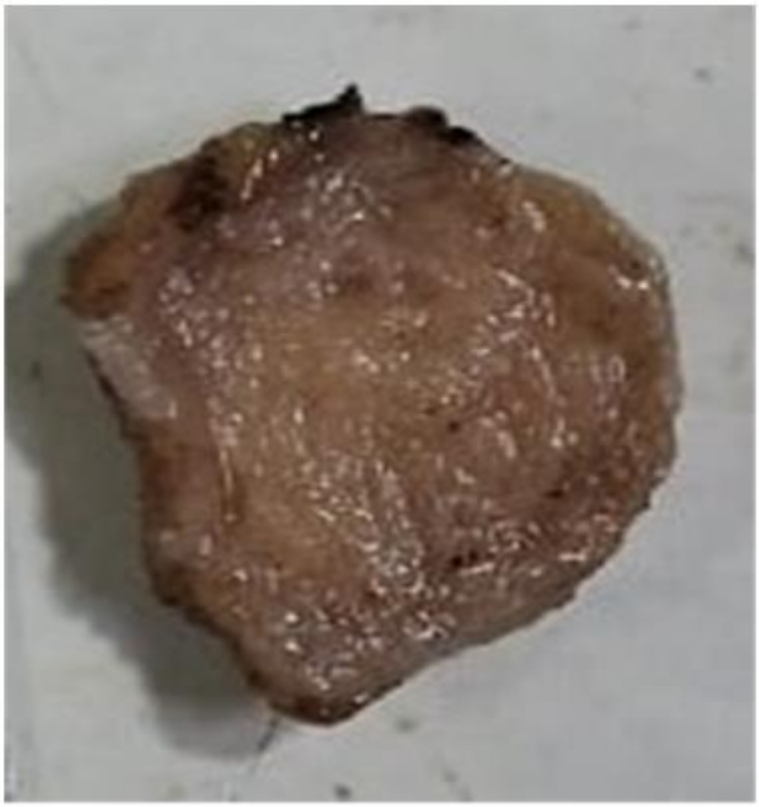
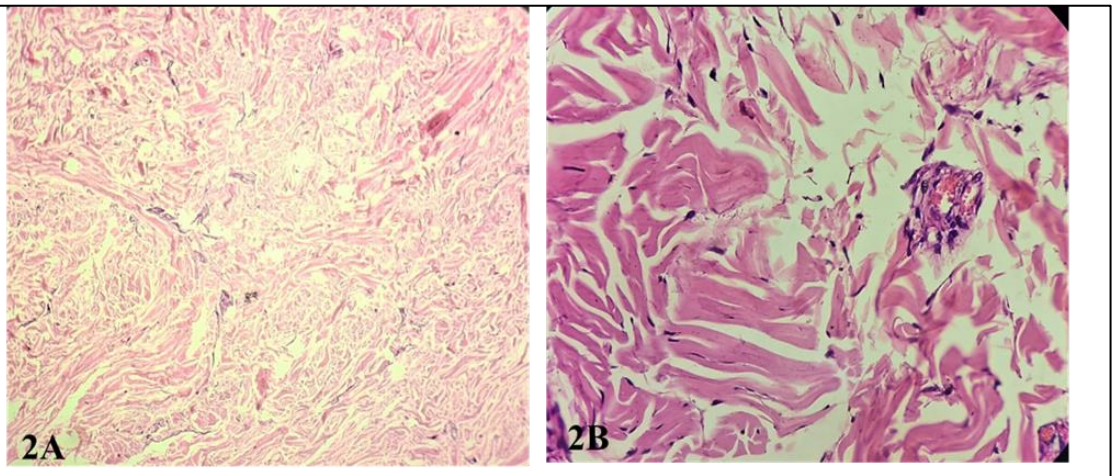


Figure 1: A circumscribed, but non-encapsulated swelling measuring 1.7x1.0 cm with a grey-white, firm to hard cut surface



Figures 2 A&B: Sections show a hypocellular lesion composed of thick bundles of collagen with interspersed blood vessels and scattered lymphocytes. A: (H&E, 4x10 X), B: (H&E, 40x10)

DISCUSSION

NTF was first described by Enzinger and Weiss in 1988 [3]. The rare benign neoplasm arises usually within the dermis and subcutaneous tissue of the posterior neck or upper back. Approximately, one-third of the nodules may appear within extra-nuchal sites such as the upper and lower back, buttocks, extremities, lumbosacral, scapular, or facial region, shoulder, axilla, forearm, torso, anterior neck, knee, ankle, or scalp [2, 6, 7]. The majority of neoplasms are solitary, although multiple lesions on the upper back or posterior neck have been observed.6 In our case, the lesion was solitary and had a typical nuchal location.

It is predominantly seen in males (male-to-female ratio of 4:1) as was seen in our case.2,8 It typically arises between the third to fifth decades with a mean age of 40 years, although tumefaction can appear within 3 years to 74 years [2, 8]. In the present study, the patient was a 71-year-old male. The size of the tumor ranges between 0.7 cm to 20 cm, and in our case, the tumor measured approximately 1.7 cm in maximum diameter [8].

The pathogenesis of NTF has not been established till date but comorbidities like diabetes mellitus and Gardner's syndrome are documented in literature [1, 3, 9]. There may be a genetic predisposition, especially in patients with Gardner's syndrome.9 Repeated trauma has also been suggested as a possible etiopathogenetic factor [4, 5]. No association with the aforementioned etiologic factors was observed in the present case.

Clinically, the lesion is usually asymptomatic, slow-growing and presents as a non-tender, mobile, poorly circumscribed mass [2, 6]. When symptomatic, pain is the most common complaint with a restricted range of movements of the cervical spine and upper limb 10]. In our case, the patient presented with a slowly progressive non-tender swelling.

The existence of the lesion cannot always be precisely ascertained with imaging techniques [11]. On ultrasonography, the lesion can be misinterpreted as a lipoma. Magnetic Resonance Imaging (MRI) is a preferred imaging modality for discerning nuchal-type fibroma which appears as ill-defined low signal intensity lesions in T1 and T2 weighted images [4, 11]. In the present study, USG was done which showed a hypoechoic lesion suggesting a possibility of lipoma [12].

Fine needle aspiration cytology may not always provide a satisfactory diagnosis as the hypocellular nature of this lesion will lead to a lack of diagnostic material [10]. In the present study, aspiration was difficult, and scant aspirate was obtained. Cytological findings included few fat fragments with interlacing capillaries, so a diagnosis of lipomatous lesion was suggested.

An accurate diagnosis using imaging studies and aspiration cytology is usually not possible. Histopathological evaluation is therefore imperative for the differentiation of nuchal fibroma from other soft tissue tumors arising in the posterior neck [11].

Grossly, nuchal-type fibromas are circumscribed masses with a firm to hard consistency and are grey-white in color.1 Extra-nuchal lesions appearing at diverse sites are morphologically similar to nuchal-type fibromas.6 Similar gross morphology was noted in the present study. Microscopically, NTF consists of haphazardly arranged collagen fibers with scattered fibroblasts and entrapped adipose tissue, nerve fibers, and adnexal structures [6]. Similar microscopic findings were observed in our case. However, no entrapment of adnexal structures, nerve fibers, or adipose tissue was observed.

Possible histopathological differentials include fibrolipoma, extra-abdominal fibromatosis, elastofibroma, keloid, collagenous and desmoid fibromas, and dermatofibroma [4, 11]. Fibrolipoma, the most common differential diagnosis can be distinguished by the presence of features like encapsulation, greater proportion of mature adipose tissue, and lack of entrapped nerves. Subcutaneous location, paucity of cells, and lack of cartilaginous metaplasia can differentiate NTF from extra-abdominal fibromatosis and nuchal fibrocartilaginous pseudotumor [3, 13]. Elastofibroma arises in the deep soft tissue of the scapula in the inferomedial portion and contains characteristic fragmented elastic fibers. Keloid scar develops as a reaction to injury and has denser, thicker collagen than NTF [14]. Collagenous fibromas are typically negative for CD34, and desmoid fibromas often show a positive reaction to beta-catenin. These immunohistochemical characteristics help differentiate them from NTF. Dermatofibroma is distinguished by the presence of pseudoepitheliomatous hyperplasia and hypertrophied basal cells in the epidermis [4].

A significant limitation of the study is the absence of immunohistochemistry, a valuable tool for differentiating NTF from other similar soft tissue tumors. Immunohistochemical staining for markers like CD34, beta-catenin, and others can provide critical diagnostic information and help confirm the diagnosis more robustly.

Although the tumor is benign, it tends to recur, therefore complete surgical excision remains the treatment of choice [9].

CONCLUSION

To conclude, while NTF is mostly benign, it presents challenges in both diagnosis and management.

Characterized by slow growth and typically asymptomatic presentation, accurate diagnosis often requires histopathological evaluation due to inconclusive imaging and cytological findings. While treatment primarily involves surgical excision, vigilance is necessary due to its propensity for recurrence. Further research into its etiology and improved diagnostic modalities could enhance patient care and outcomes.

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Data Availability: The data has not been made public, and are open from the corresponding author on reasonable request.

Ethics Approval: The subjects gave their informed consent for inclusion before they participated in the study.

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