

# Nuchal-type fibroma of the shoulder: A case report and review of the literature

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**Abstract.** Nuchal-type fibroma, initially described in 1988 by Enzinger and Weiss, is a rare clinical entity associated with distinct subcutaneous and dermal fibrous tissue proliferation. The etiology of nuchal-type fibroma largely remains to be elucidated. Typical characteristics of this entity include hypocellular, haphazardly arranged collagen with entrapped adipose tissue, paucity of elastin and entrapped small nerves, on which the pathological diagnosis is based. Magnetic resonance imaging (MRI) is the preferred imaging modality for the detection of nuchal-type fibroma, due to its superior soft tissue resolution and multi-planar capabilities. The present study presents the unique findings of a nuchal-type fibroma arising in the shoulder of a 48-year-old man. Distinct features of the nuchal-type fibroma in the present case included hyperintensity on T1- and T2-weighted MRI. Microscopic examination revealed marked mucoid tissue degeneration. To the best of our knowledge, this is the first case report of nuchal-type fibroma presenting with these distinct features. The present findings may therefore assist with the general and differential diagnosis of nuchal-type fibroma.

## Introduction

Nuchal-type fibroma is a rare, benign fibrous proliferation of unknown pathogenesis involving the dermis and subcutaneous tissues. The mass is characterized by hypocellular bundles of collagen with entrapped adipocytes and increased numbers of small nerves. Nuchal-type fibroma is generally located predominantly in the interscapular and paraspinous regions (1). In the majority of cases, the lesion is solitary; only a small number of cases involving multiple lesions have been reported (2-4). Extranuchal sites include the extremities, lumbosacral area,

buttocks and face (4-11). The tumor typically presents between the third and fifth decades of life, but lesions have additionally been reported in patients aged 3-74 years. Nuchal-type fibroma has been observed to be strongly associated with diabetes mellitus and Gardner's syndrome (12). There is a ratio of 4:1 male to female predominance, although this is not observed in Gardner's syndrome-associated patients (4).

Clinically, nuchal-type fibroma characteristically presents as an asymptomatic, firm, poorly circumscribed, subcutaneous mass, which requires wide surgical excision (1-4). However, the unencapsulated nature of this tumor makes complete resection difficult, which may contribute to its propensity for local recurrence. Alternatively, reoccurrence may be due to the persistent presence of factors that triggered the initial development of the lesion, for example, repetitive trauma (3,7). Nuchal-type fibroma is frequently misdiagnosed and under-reported due to a close histopathological similarity to other benign fibrous tumors, including dermatofibrosarcoma protuberans, giant cell fibroblastoma, mammary and extramammary myofibroblastoma, spindle and pleomorphic cell lipoma, and elastofibroma (13). Due to the rarity of nuchal-type fibroma, at present the mortality rate remains unknown.

To the best of our knowledge, <80 cases of nuchal-type fibroma have been recorded in the literature, including 20 reports of sporadic cases and one clinicopathological study of 52 cases (4). Only 3 studies describing magnetic resonance imaging (MRI) findings of nuchal-type fibroma have been published (6-8). The present study presents unique MRI findings of a nuchal-type fibroma arising in the right shoulder of a 48-year-old man.

## Case report

A 48-year-old man presented at The First Hospital of Jilin University (Changchun, China) with a palpable mass in his right shoulder in February 2014, which had been present for ~30 years. Clinical examination revealed a 20x10x5 cm solid lump in his right shoulder. Overlying skin was observed to possess a normal color and texture (Fig. 1). The patient had no significant medical history and no history of trauma in the affected area. MRI (Ingenia 3.0T; Philips Medical Systems, Inc., Bothell, WA, USA) revealed a mass with mixed signal intensity T1- and T2-weighted images; however, the high signal intensity was observed to be variable and non-unified. The

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Figure 1. Image showing a large subcutaneous mass located over the right shoulder of the patient.

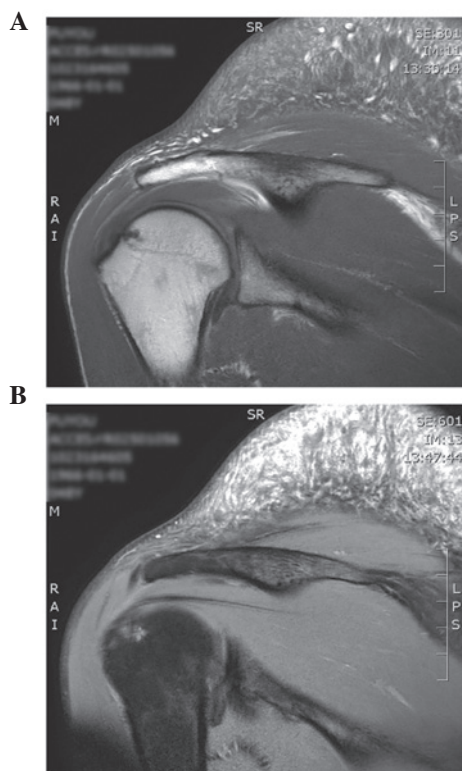


Figure 2. Magnetic resonance imaging scans. (A) Coronal T1-weighted image showing a mass with mixed signal intensity, with a strip-like pattern in the high signal intensity zone. (B) Coronal T2-weighted image with fat suppression showing mixed signal intensity, with a predominant high signal in a strip-like pattern.

high signal demonstrated a strip-like pattern. On T2-weighted images, the area of the high signal was markedly larger than that on T1-weighted images (Fig. 2). Marginal excision of the tumor was performed. A longitudinal incision was made

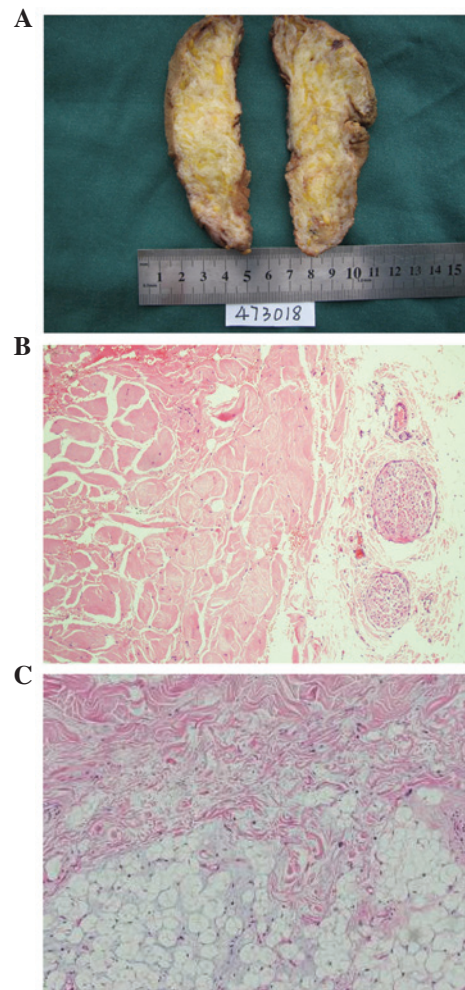


Figure 3. (A) The gross specimen was characterized by a fat-like tissue in the neoplasm. (B) Slide showing thick fibrous tissue and nerve tracks, which are features of nuchal-type fibroma. Hematoxylin and eosin staining; magnification, x100. (C) Slide showing typical collagen fibers, as well as mucoid tissue degeneration, which had not been previously reported in the literature. Hematoxylin and eosin staining; magnification, x200.

to reveal the mass. The lesion was not well demarcated and did not display a definite capsule. The tumor was resected completely with a large surrounding margin of normal tissue. Intraoperative fast frozen pathology resulted in the diagnosis of fibroma, and the incision was sutured without extensive excision. Light microscopic examination (DP20; Olympus Corporation, Tokyo, Japan) of the pathological specimen revealed following staining with hematoxylin and eosin revealed that the mass was composed of typical collagen fibers. Mucoid tissue degeneration was observed. The clinicopathological features of the resected tissue were consistent with a diagnosis of nuchal-type fibroma (Fig. 3). At the time of writing, the present patient remained free of recurrent disease at the 8-month follow-up appointment. Further observation of the patient will be performed in the outpatient department.

## Discussion

Nuchal-type fibroma typically presents as an asymptomatic, relatively slow-growing, superficial mass (1-2,11). The mean tumor diameter is 3.5 cm, but tumors have been observed to

reach 8 cm in diameter (4). To the best of our knowledge, the largest nuchal-type fibroma recorded in the literature was 16.5x15.0x6.5 cm (10). The present study reports a case of a nuchal-type fibroma with a size of ~20x10x5 cm. The mass was large, visible under the skin of the patient's right shoulder and easily palpable.

MRI is the preferred method to visualize nuchal-type fibroma and its components and borders. Previously published MRI of nuchal-type fibromas demonstrated masses with a low or mild signal intensity (6-8). The present MRI revealed a mass that displayed mixed signal intensity on T1- and T2-weighted images, which was a prominent difference compared with previously reported findings. The present MRI findings may assist in the differential diagnosis of nuchal-type fibroma of the shoulder, particularly for those with high signal intensity, which potentially indicates mucoid degeneration of the nuchal-type fibroma (14,15). To the best of our knowledge, this is the first report of mucoid degeneration in nuchal-type fibroma. Since nuchal-type fibromas and Gardner-associated fibromas resemble one another, it has been suggested that a subset of nuchal-type fibromas that occur in multiple sites, unusual locations or in children may be Gardner-associated fibroma, in which the fibromatosis may present a sentinel event for identification, which requires further genetic analysis (16).

The etiology of nuchal-type fibroma remains to be elucidated, but its microscopic features have been described and confirmed by several studies (1-4,11-13,17,18). Microscopically, the majority of nuchal-type fibromas are composed of haphazardly arranged thick collagen fibers. Fibroblasts are sparsely scattered between the collagen fibers (13,18,19). In addition, entrapped islands of adipose tissue and skeletal muscle are typically observed (1,3,13). In certain cases, the lesions may contain enlarged peripheral nerves with perineural fibrosis (1,13). Microscopic examination of the pathological specimen from the present patient revealed typical nuchal-type fibroma collagen fibers and mucoid tissue degeneration. Additional research is required to confirm the factors involved in the pathogenesis of nuchal-type fibroma.

Gross total resection within healthy tissue is the preferred treatment for nuchal-type fibroma (3). Michal *et al* (4) reported that nuchal-type fibroma has a tendency to recur following excision. From 25 reviewed cases of nuchal-type fibroma, 3 patients presented with a single recurrence, 2 patients with lesions that recurred twice and 1 patient with a lesion that recurred 3 times (4). In the present case, complete surgical removal was performed with a margin of surrounding normal tissue. A longitudinal incision was made to reveal the entire mass. As the tumor was not well demarcated, localization and definition of the tumor margin was very important. Intraoperative fast frozen pathology resulted in the diagnosis of fibroma, and the operation was concluded without extensive excision. Retrospectively, the procedure may appear inappropriate for the treatment of nuchal-type fibroma, since incomplete resection may be associated with an increased recurrence rate. The final pathological diagnosis indicated nuchal-type fibroma, emphasizing that accurate intraoperative diagnosis is essential in such cases. At the time of writing, the present patient remained free of recurrent disease at the 8-month follow-up appointment.

In conclusion, the present nuchal-type fibroma demonstrated unique MRI findings and microscopic features

that differed from previously reported cases. Even though nuchal-type fibroma is generally asymptomatic, it should be considered as a potential diagnosis for a subcutaneous mass with mixed signal intensity on T1- and T2-weighted images, particularly when the high signals appear in a strip-like pattern, with additional findings of typical collagen fibers and mucoid tissue degeneration observed by microscopy. The present findings provide useful information that may improve the clinical management of this uncommon lesion.

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