Multiple Nuchal-Type Fibromas on the Scalp: A Case Report

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Nuchal-type fibroma is a rare, benign, fibrous, tumor-like proliferation characterized by dense hypocellular bundles of collagen, with sparsely scattered fibroblasts, interspersed fat tissue, and entrapped nerve fibers. It usually develops in the posterior neck as a solitary, firm, poorly circumscribed, subcutaneous mass. Herein, we report about a 32-year-old man who presented with a 6-year history of multiple nodules on the scalp. Histopathological features were consistent with those of nuchal-type fibroma. The tumors were surgically excised. (Ann Dermatol 27(2) 194~196, 2015)

Keywords
Nuchal-type fibroma, Scalp

INTRODUCTION

A nuchal-type fibroma (NTF) is a rare, benign, fibrous, tumor-like proliferation of unknown pathogenesis. An NTF is characterized by dense hypocellular bundles of collagen interspersed with fat tissue, entrapped adnexal structures, and peripheral nerve fibers. NTFs usually develop as solitary, hard, poorly circumscribed, subcutaneous masses in the posterior aspect of the neck. However, approximately one-third of all cases occur at other sites. Most extranuchal tumors arise in the back, scapular region, shoulder, and face, but cases have been reported to occur in other parts of the body, including the axilla, forearm, trunk, anterior neck, knee, ankle, and scalp. NTFs present as single lesions in most cases; only a few cases involving multiple lesions have been reported. Herein, we report an unusual case of NTF that arose in the scalp, an uncommon extranuchal site, and presented as 6 distinct lesions.

CASE REPORT

A 32-year-old man presented to our department with a 6-year history of multiple nodules on the scalp. The patient had experienced intermittent pain and an itching sensation on the lesions. On physical examination, 6 protruding, subcutaneous, firm nodules of varying size were observed on the scalp (Fig. 1). The nodules were several centimeters in size. An earlier ultrasonographic examination
at a different hospital had indicated that the lesions could be suspected lipomas. For a further diagnosis, an excisional biopsy was performed for a single lesion. Histopathological examination revealed a poorly circumscribed, paucicellular lesion composed of thick, haphazardly arranged collagen fibers with sparsely scattered fibroblast-like cells in the subcutis (Fig. 2A, B). Adipose tissue islands and peripheral nerve fibers were entrapped by the collagen fibers (Fig. 2C, D). These findings were consistent with those of NTF. The other lesions were also excised, and they displayed the same histopathological features.

**DISCUSSION**

Nuchal fibromas are rare, benign, fibrous tumors, first described in 1988 by Enzinger and Weiss. Nuchal fibromas characteristically present as asymptomatic, firm, poorly circumscribed, subcutaneous masses that generally develop in the posterior neck. Histopathologically, nuchal fibromas are composed of thick, haphazardly arranged collagen fibers, with sparsely scattered fibroblasts. The entrapment of adnexal structures, adipose tissue, and peripheral nerve fibers by the collagen bundles is a notable histologic feature. Nuchal fibromas typically develop in the nuchal region, but in approximately one-third of the cases, it occurs at extranuchal sites. The extranuchal lesions are morphologically and histologically indistinguishable from those of the nuchal region; therefore, in 1999, Michal et al. proposed the term ‘nuchal-type fibroma’ to encompass all lesions histologically similar to nuchal fibroma, irrespective of their site of origin. The most common extranuchal sites of NTF are the back (particularly the interscapular region), shoulder, and face. In addition, cases have been reported to occur in other parts of the body including the axilla, forearm, trunk, anterior neck, knee, ankle, and scalp.
In most cases, NTF presents as a single lesion. However, 3 cases with 2 distinct lesions have been described. Recently, a case of NTF presenting as multiple lesions on the posterior neck and upper back was also reported. The pathogenesis of NTF remains unknown, but some cases of NTF have been associated with trauma, diabetes mellitus, and Gardner syndrome. When NTF occurs at multiple sites or unusual locations, the possibility of a Gardner-associated fibroma prior to NTF should be considered. However, our patient had neither diabetes mellitus nor Gardner syndrome.

Wide surgical excision is the most acceptable treatment for NTF. However, the unencapsulated nature of NTF lesions makes complete excision difficult, potentially contributing to their propensity for local recurrence. Alternatively, recurrence could be owing to the persistent presence of factors that triggered the initial development of the NTF lesion, including repetitive trauma.

Many cases of NTF have been misdiagnosed because of its indolent clinical course and histopathological similarity to other benign fibrous tumors. NTF needs to be distinguished from other fibrous tumors and tumor-like conditions, including desmoid-type fibromatosis; circumscribed storiform collagenoma; connective tissue nevus; collagenous fibroma; fibroma of the tendon sheath; scleroderma; and scar tissue. NTF differs fromtheses tumors by its marked hypocellularity, poor circumscription, and presence of entrapped adipose tissue.

The case described here was a rare NTF that presented as multiple lesions on the scalp, an uncommon extranuchal site.

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