Connective Tissue Nevus: Case Report

Konnektif Doku Nevüsü

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ABSTRACT Connective tissue nevus is defined as hamarthomatous malformations of dermal connective tissue components including collagen, elastic fibers or glycosaminoglycans. The mechanism of the increased connective tissue components remains uncertain. Collagen type of connective tissue nevus is the most common type. We herein report a collagen type of connective tissue nevus with clinical and pathological features in a 20-year-old man with a 7 year history of lesions on parasacral region. This case has been presented since it is a rare and benign lesion, and may be important for the differential diagnosis.

Key Words: Nevus, connective tissue, skin and connective tissue diseases

ÖZET Konnektif doku nevüsü kollajen, elastik lifler ve glikozaminoglikanlar gibi dermal konnektif doku komponentlerini içeren hamartomatöz bir lezyon alarak tanımlanır. Konnektif doku komponentlerinin artmasının mekanizması kesin bilinmemektedir. Konnektif doku nevüsünün en sık görüleni kollajen tipidir. Biz parasakral bölgede 7 yıldır mevcut lezyonu bulunan, klinik ve patolojik olarak konnektif doku nevüsünün kollajen tipi tanısı alan 20 yaşında erkek hastayı, nadir ve benign lezyonlarda ayırıcı tanıda önemli olduğu için sunmaktayız.

Anahtar Kelimeler: Nevüs, konnektif doku, deri ve konnektif doku hastalıkları

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Connective tissue nevus (CTN) is defined as hamartomatous malformations of dermal connective tissue components including collagen, elastic fibers or glycosaminoglycans. CTN is a rare disease and may be familial or acquired.[1] The predominant element of the extracellular connective tissue within an individual nevus can be collagen, elastic fibers, or proteoglycan (mucin).[2,3] The mechanism of the increased connective tissue components remains uncertain.[4] A unifying theory regarding connective tissue was proposed, in which abnormalities of collagen, elastic fibers and mucopolysaccharide synthesis were considered, but it could not explain in all cases.[5] We presented a case report with CTN that we considered as clinical and histopathological.

CASE REPORT

A 20-year-old man had a 7 year history of lesions on parasacral region. The lesions showed gradual enlargement after its initial occurrence in addi-

tion to slight itching. Physical examination showed multiple oval cutaneous plaques on the parasacral area (Figure 1). Individual lesions were soft, well demarcated, and ranged in size from 0.5 to 6 cm. Plaques have always been flesh-colored with a yellowish tinge. There were no hypopigmentes macules, café au lait spots and adenoma sebaceum. The remaining physical examinations including neurological evaluation, the results of routine laboratory studies, and radiological examinations were normal. There was no family history of similar lesions. A punch biopsy was taken from the lesion. The specimen showed minimal epidermal changes and thickening of the dermis due to increased collagen deposition (Figure 2). In the upper and mid-dermis, an accumulation of dense, coarse, thick collagen fibers was present (Figure 3). Masson's trichrome stain showed a significant increase of thick collagen bundles (Figure 4). Verhoeff's elastic tissue stain showed normal elastic fibers (Figure 5). The patient was defined as collagen type of CTN with clinical and pathological features of lesions.

DISCUSSION

In 1921, Lewandowsky first reported four cases of CTN as "*neavus elasticus regionis mammariae*".[5]

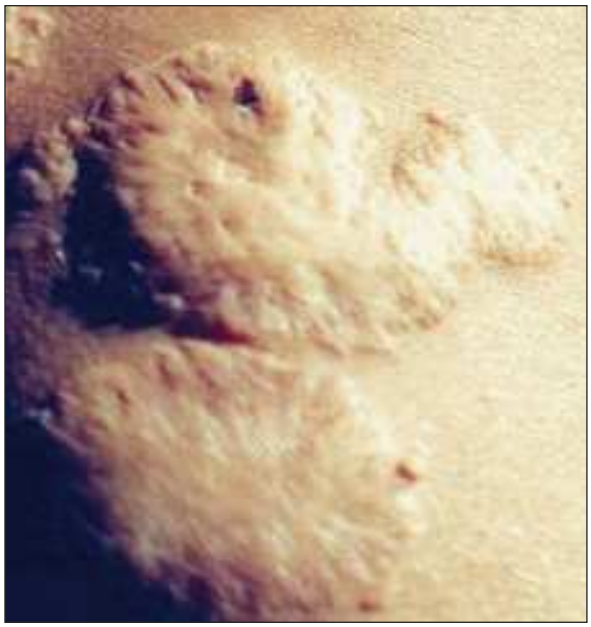

FIGURE 1: Multiple flesh-colored with yellowish tinge oval cutaneous plaques on the parasacral area of the patient.

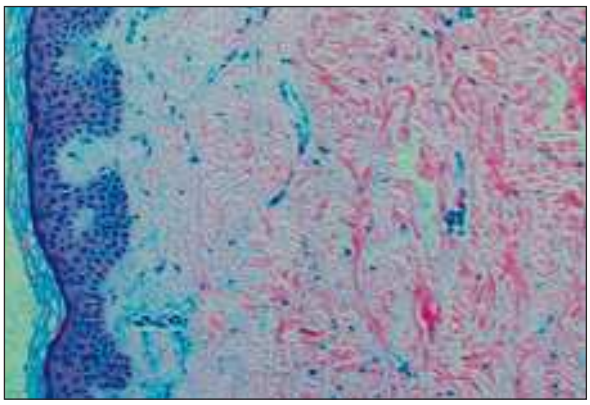

FIGURE 2: An increased amount of collagen in the dermis. (Hematoxylin-eosin, X10)

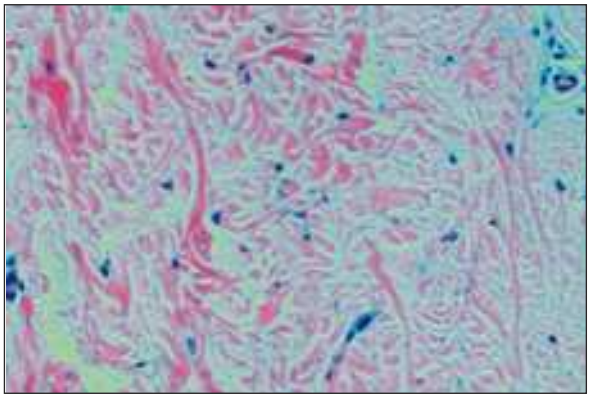

FIGURE 3: Biopsy specimen showing many dense, coarse collagen fibers. (Hematoxylin-eosin, X20).

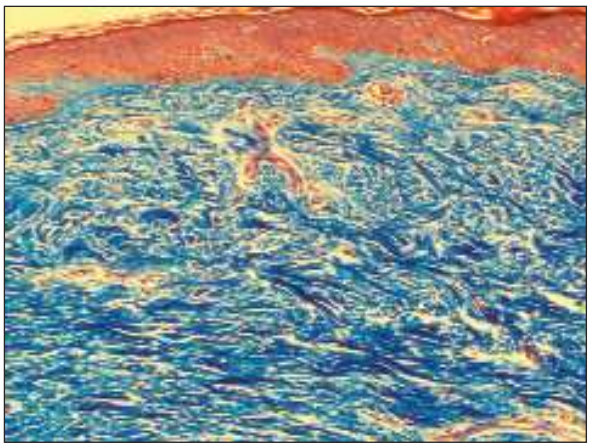

FIGURE 4: An increased amount of collagen in dermis (Masson's trichrome, x20).

The following year, Lipschütz reported four cases as pavingstone connective tissue naevus. The term "*connective tissue nevus*" became more commonly accepted when Gutmann's review appeared

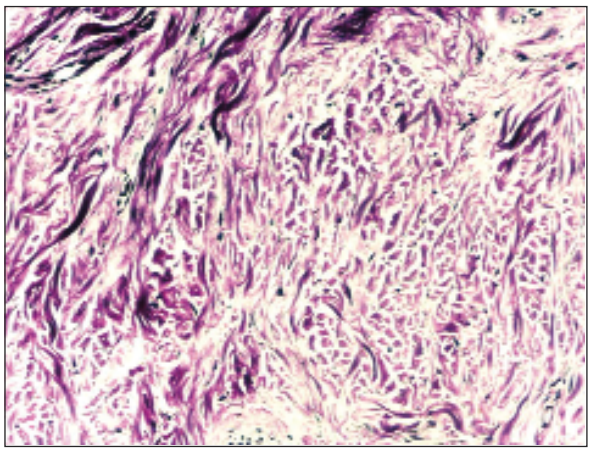

FIGURE 5: Elastic tissue stain demonstrates normal amounts of elastic tissue (Verhoeff's elastic, x 20).

in 1926. In this review, CTN in which elastic tissue are predominant were referred to as the Lewandowsky type, while those in which collagen changes are predominant were referred as the Lipschütz type.[5,6] On the basis of clinical, histological, and genetic consideration, the CTN can be classified into defined categories. Pierard and Lapiere classified nevi of connective tissue into two main basic subgroups according to the portion of the dermis principally involved; nevi of reticular connective tissue and nevi of the adventitial connective tissue.[2] Uitto et al. proposed another classification based on clinical, genetic, and histopathologic considerations and accordingly they can be either inherited (familial cutaneous collagenoma, shagreen patches in tuberous sclerosis) or acquired (eruptive collagenoma, isolated collagenoma).[1,7]

CTN is also subdivided into collagen, elastin, proteoglycans and mixed type, based on their particular histopathologic features.[8] Collagen type of CTN that is most common is characterized by an excessive accumulation of dense, coarse collagen fibers in the dermis. Elastic fibers may appear normal or diminished.[9-11] Elastic type CTN is characterized by a marked increase in elastine. In the proteoglycan (mucin) type, mucin deposition is the main histological feature.[3]

Clinically CTN is elevated, soft to firm, varying from 0.5 to several centimeters in diameter papules, nodules or plaques. It may be solitary or multiple.[11] CTN can be defined as eruptive, papular, papulolinear, zosteriform according to appearance and location of the lesion. CTN may be skin-colored, yellowish, brownish or hypopigmented. The back and thighs are usually involved, less frequently the upper extremities, flexures and face.[8] Familial cutaneous collagenoma is an autosomal dominant inherited condition characterized by the presence of numerous, asymptomatic, symmetrical cutaneous nodules or papules on the trunk, upper arms; most are located on the upper two thirds of the back in distribution.[2,7,12] The hereditary types of connective tissue nevus include dermatofibrosis ventriculares disseminata in the Buschke Ollendorf syndrome, familial cutaneous collagenoma and shagreen patches in tuberous sclerosis.[6]

The differential diagnosis of CTN might include Buschke-Ollendorf syndrome, dermatofibroma, lipoma, nevus lipomatodes superficialis (Hoffmann-Zurhelle), Pseudoxanthoma elasticum and tuberous sclerosis.[7] In our case, classic skin findings of Shagreen patches in tuberous sclerosis (adenoma sebaceum, periungal fibromas, and ash-leaf macules) were not found. Buschke-Ollendorf syndrome is characterized by multiple CTN and osteopoikilosis, a benign disease of the bone characterized by circumscribed areas of increased bone density.[10-12] The other diagnoses may be easily reached histopathologically.

CTN is benign lesions and at present, there is no recommended effective therapy for those cutaneous lesions except the surgical removal of those apparent papules and nodules.[1] Therefore, surgical excision may be performed when the patient would like the lesion removed for cosmetic reasons; however, surgery may not be advised when multiple or large lesions are present.

In our case, the lesion was first seen when the patient was 20 years old and he had no significant medical or family history, there was no other lesion except parasacral region and surgical removal was not applied since the patient did not give consent. This case has been presented since it is a rare and benign lesion, and may be important for the differential diagnosis.

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