Zosteriform Collagen Nevus in an Infant

Connective tissue nevi (CTN) are dermal hamartomas characterized by an imbalance in the amount and distribution of the normal components of the extracellular dermal matrix, specifically collagen, elastin, and/or proteoglicans. The term "CTN" was first mentioned by Lewandowsky in 1921 (1), although it was not accepted until the review by Gutmann in 1926 (2). Classification of CTN was established by Uitto *et al*. (3) in 1980 according to clinical, genetic, and histopathological features. But this classification did not include zosteriform nevi. The more recent Pierard and Lapiere (4) classification seems to be a more suitable method of classification for zosteriform nevi. They classified CTN into two groups: (1) reticular and (2) adventitial. Zosteriform nevus is a rare form of reticular CTN that is diagnosed according to its clinical distribution. Here we report a collagen nevus in an infant that followed a zosteriform pattern.

An 8-month-old girl presented with flesh-colored plaques on the right buttock in a zosteriform distribution, which had been present since birth. The plaques appeared to be well-defined cobblestone-like nodules on palpation (Figure 1). Systemic examination, laboratory tests and radiologic examinations did not reveal any abnormalities. The patient had no associated disease and no history of similar skin findings among family members.

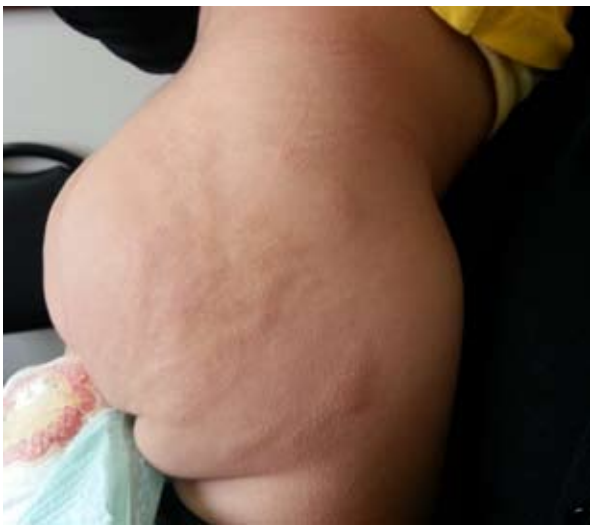

Figure 1. Zosteriform, flesh-coloured plaques on the right buttock.

A skin punch biopsy was performed from one of the nodules. The histopathologic examination showed significantly increased density of thickened collagen fibers in the lower dermis and subcutaneous tissue. Verhoeff-van Gieson and orcein stains demonstrated the presence of dense collagen fibers with diminished elastic fibers (Figure 2).

Four subtypes of collagen tissue nevus have been described: (I) familial cutaneous collagenoma, (II) shagreen patches in tuberous sclerosis, (III) eruptive collagenoma, (IV) and isolated collagenoma (5). Isolated collagenoma with lack of family history is fairly rare. It is sporadic, localized to only one body region, and not associated with any disease. In confluent plaques it has the appearance of "peau de chagrin" or a cobblestone-like pattern. The reported presentations include paving stone nevi, plantar fibromatosis, papulolinear lesions, and zosteriform lesions (5). Zosteriform distribution is an extremely rare variety of connective tissue nevus. Steiner (6) was the first to describe the condition in 1944, in a 5-year-old girl who presented with nevi in a zosteriform distribution. The histopathology of the lesion revealed an abnormality in both collagen and elastin fibers. Only 11

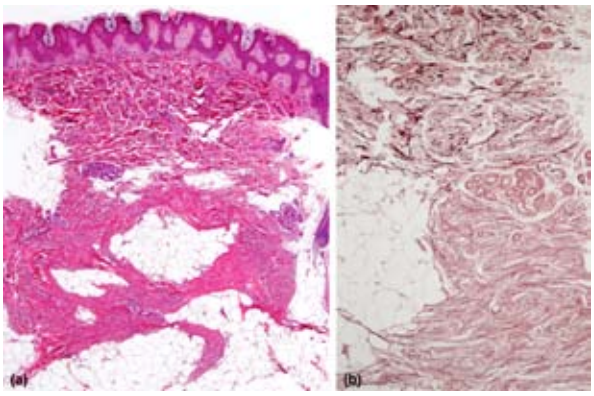

Figure 2. (a) Dense, coarse, thick collagen fibers in the lower dermis and subcutaneous tissue (HE; x 40). (b) Diminished elastic tissue fibers with dense collagen fibers (Orcein; x 100)

other cases have been reported as zosteriform CTN in dermatological literature. To the best of our knowledge, only 3 collagen nevi in a zosteriform distribution have been previously described in the literature. De *et al.* (7) described the first case in a 25-year-old man presenting a collagen tissue nevus with zosteriform distribution located over the lower back. Subsequently, Kumari *et al.* (8) described the second case in a 20-year-old man presenting a large, flesh-colored, well-defined plaque in a zosteriform distribution on his right buttock since birth. Topal *et al.* (9) reported another case of a 10-year-old boy with a zosteriform collagen tissue nevus on his right arm as sclerotic papules and plaques. Clinically, zosteriform CTN has similar morphology and distribution to nevus lipomatosus superficialis (NLS) or segmental neurofibromatosis. The latter differential possibility needs to be excluded due to its association with gliomas. The histopathologic findings of NLS make it easy to differentiate from zosteriform CTN. The peculiar finding of ectopic fat in the dermis is considered to be almost pathognomonic of NLS (10).

In conclusion, the zosteriform distribution of CTN is very rare, especially in the variety with collagen predominance. As the lesion remains asymptomatic, with only cosmetic effects, the condition needs no specific treatment. The present case is a rare type of isolated collagenoma with zosteriform distribution presenting over the right buttock with no associated abnormalities and family history.

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