

A large asymptomatic lesion on buttock since birth

A 20-year-old Indian male had a large asymptomatic lesion on his right buttock since birth. The lesion started growing in size at puberty and eventually attained the current size. There was no history of a similar lesion in his family.

On physical examination, there was a large 15 x 10 cm, firm, solitary, flesh colored, well - defined plaque with mammillated surface in the S3 dermatome over the left buttock extending onto the inner side of the thigh in a zosteriform distribution

[Figure 1]. The rest of the physical and systemic examination ruled out any other phakomatoses. Skin biopsy was done from the edge of the plaque. On hematoxylin and eosin staining, no apparent histopathological abnormality was seen. Findings of special stain are given in [Figure 2]. Elastic fibers appeared widely spaced and there was no increase in mucopolysaccharides. His skeletal survey, however, was unremarkable.

WHAT IS YOUR DIAGNOSIS?



Figure 1: Zosteriform, flesh coloured, well- defined plaque with mammillated surface in the S3 dermatome over the left buttock

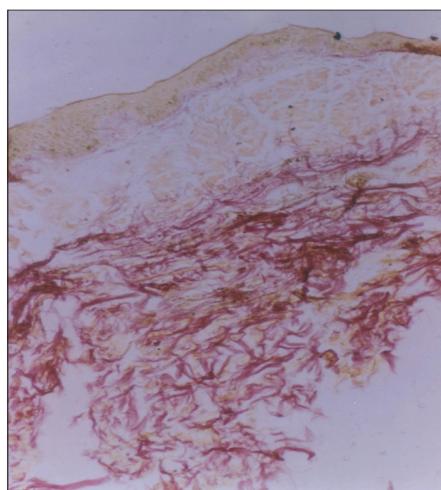


Figure 2: Verhoeff-van Gieson stain (X 1000)

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Diagnosis: Zosteriform collagen nevus

DISCUSSION

Special staining with Verhoeff-van Gieson showed red, thick and homogenized collagen bundles in a haphazard arrangement in the dermis suggestive of collagenoma [Figure 2]. Connective tissue nevi are circumscribed hamartomatous malformations of the dermal extracellular matrix, i.e., of collagen, elastic fibers or glycosaminoglycans. These lesions occur characteristically on the trunk, most often in the lumbosacral area. They may be solitary but often are multiple and may show a zosteriform arrangement.^[1]

In 1921, Lewandowsky first reported four cases of connective tissue nevi as “nevus elasticus regionis mammariae”, in which the collagen tissue was apparently unaltered.^[2] In this series of patients, elastic tissue was absent in the papillary dermis and presented as fragmented or split fibers. In the deeper portion of the dermis, this elastic tissue was aggregated in homogeneous masses. The following year, Lipschütz reported four cases described as “Pflastersteinförmiger bindegewebsnaevus” (pavingstone connective tissue nevus) in which the elastic tissue was essentially normal, but the collagen bundles were hypertrophic, swollen and homogeneous. The term “connective tissue nevus” became more commonly accepted when Gutmann’s review appeared in 1926.^[3] In this review, connective tissue nevi in which elastic tissue changes predominated were referred to as the Lewandowsky type and those in which collagen changes were conspicuous were termed the Lipschütz type (as seen in our case).

Presently the connective tissue nevi classification is based on that proposed by Uitto *et al.*^[4] and Pierard *et al.*^[5] as collagen nevi, elastic nevi, proteoglycan nevi and nevi of adventitial connective tissue. Some cases may have an increased amount of both collagen and elastic fibers when they are called mixed type. Collagen nevi may be acquired or hereditary. Acquired type includes eruptive collagenoma, plantar cerebriform collagenoma, isolated collagenoma or elastoma. The hereditary types of collagen nevi include dermatofibrosis lenticularis disseminata in the Buschke-Ollendorff syndrome, familial cutaneous collagenoma, shagreen patch etc.

In 1944, Steiner^[6] described a 5-year-old white girl presenting a nevus with zosteriform distribution located on the right

side of the lower chest and back with increase in both collagen and elastic fibers on histology. Subsequently, in 1985, Kozminsky^[7] described a second case in a 23-year old Nigerian woman presenting flesh-coloured plaques with cobblestone appearance in a zosteriform distribution on her left upper back and the dorsal aspect of the left arm. In 2003, Yeh *et al.*^[8] reported another case of a zosteriform connective tissue nevus in a 3-year-old boy on the right side of abdomen and flank involving T7-T12 dermatome as a single 9x7 cm cobblestone-like plaque. The present case is an example of this rare non familial collagen nevus in zosteriform distribution that involved the S3 dermatome. This case further supports the view that zosteriform connective tissue nevi should be considered as a separate entity because of their unusual clinical presentation, the lack of a genetic inheritance and the absence of associated abnormalities in other organs.

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