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Authors
López, Diego
Beltramo, Pilar
Acosta, Agustina

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Papulolinear collagenoma: a rare entity in children

Diego López[1] MD, Pilar Beltramo[2] MD, Agustina Acosta[3] MD

Affiliations: [1]Pediatric Dermatology Unit, Pereira Rossell Hospital, Montevideo, Uruguay, [2]Pereira Rossell Hospital, Montevideo, Uruguay, [3]Pediatric Dermatology Unit, Pereira Rossell Hospital, Montevideo, Uruguay

Corresponding Author: Diego López MD, Pediatric Dermatology Unit, Clinic Hospital Dr Manuel Quintela, Avenida Italia 11600, Montevideo, Uruguay, Tel: 598-99330186, Email: diegolodamar@gmail.com

Abstract

Papulolinear collagenoma is a rare kind of connective tissue nevus. It is a dermal hamartoma characterized by an increase in collagen. We report a young girl's collagen nevus with a papulolinear distribution.

Keywords: collagenoma, connective tissue nevus, dermatopathology, hamartoma

Introduction

Connective tissue nevi (CTN) are dermal hamartomas characterized by an alteration in the components of the extracellular dermal matrix, including collagen, elastin, and/or glycosaminoglycans. When collagen is elevated, they are called collagenomas and although no comprehensive classification is yet available they may be divided into two groups: inherited and acquired. There are two subtypes of inherited collagenomas: familial cutaneous collagenoma (FCC) and shagreen patches of the tuberous sclerosis complex (TSC). The acquired forms include eruptive collagenomas (EC) and isolated collagenoma. In the latter, the reported presentations include paving stone nevus, zosteriform collagenoma, and papulolinear collagenoma [1]. There are a few reports of isolated collagenoma but papulolinear distribution is extremely rare with only a few reports in the literature [1-4].

Case Synopsis

An 8-year-old girl presented with asymptomatic sclerotic flesh-yellowish colored papules and nodules in a linear distribution over her right popliteal space, not following a specific dermatome. The cutaneous changes were present since birth and been slowly increasing in size (**Figure 1**). The rest of the physical examination was normal. She had no systemic symptoms or associated disease, and no family history of similar dermatoses.

A punch biopsy was performed. Hematoxylin and eosin stain showed a normal epidermis, increased thick collagen bundles in the dermis, and dermal thickening (**Figure 2**). Trichrome stain demonstrated haphazardly arranged thick collagen bundles in the dermis (**Figure 3**) and orcein stain showed the presence of dense collagen fibers with diminished elastic fibers (**Figure 4**).

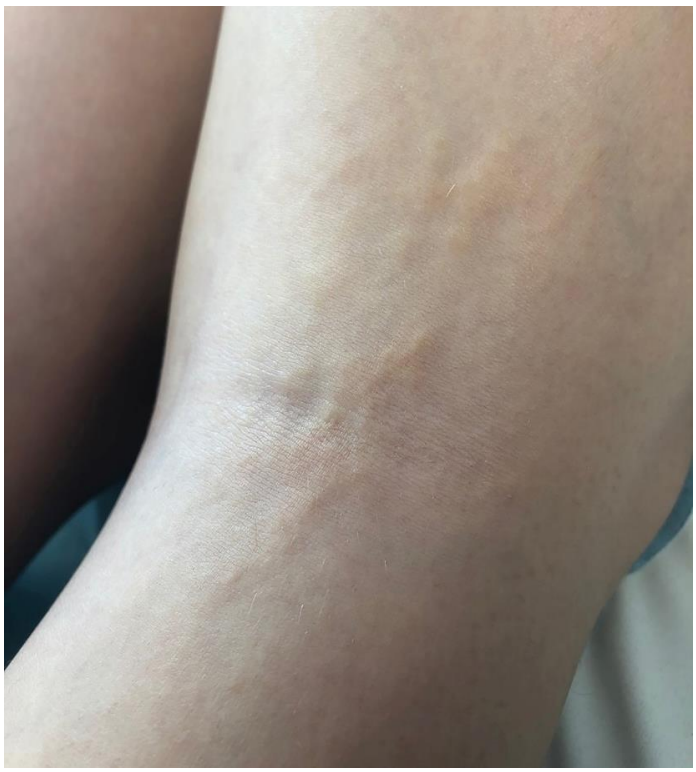

Figure 1. *Flesh-color-yellowish papules and nodules in lineal distribution over right popliteal space.*

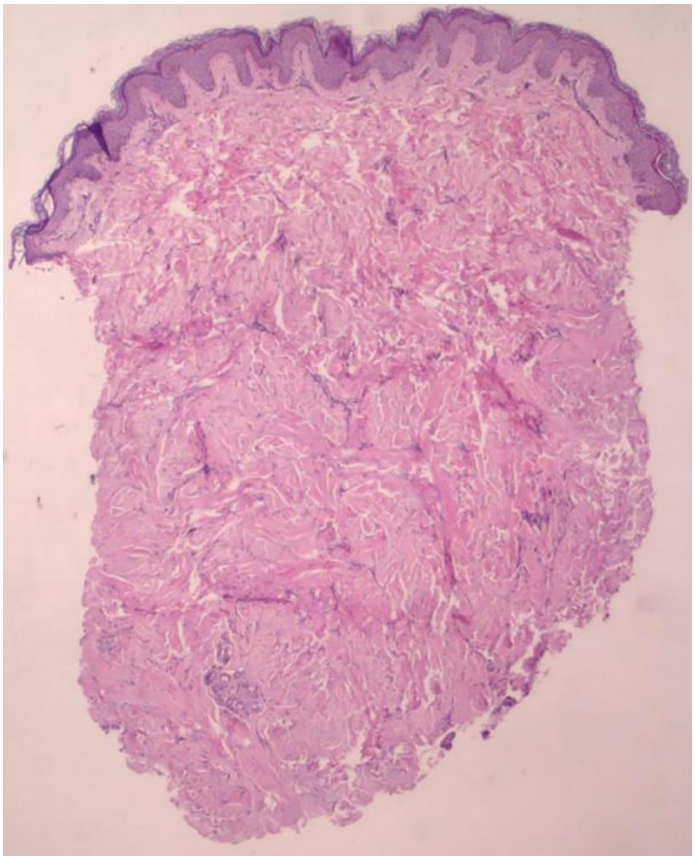

Figure 2. *A normal epidermis, increased thick collagen bundles in the dermis, and dermal thickening. H&E, 20×.*

The lesion was diagnosed as papulolinear collagenoma according to its clinic-pathological findings. Conservative management with observation was the treatment plan because of the benignity of the entity and its asymptomatic nature.

Case Discussion

Papulolinear collagenoma is a type of CTN. Connective tissue nevi are dermal hamartomas characterized by an alteration in the components of the extracellular dermal matrix, including collagen, elastin, and/or glycosaminoglycans [5]. When collagen is the one increased, they are called collagenomas as in our patient. Its pathogenesis is not well known but it is believed to relate to an imbalance between collagenase activity and collagen production by fibroblasts [1].

Within the different collagenoma types FCC is an autosomal dominant disease characterized by symmetrically distributed multiple flesh-colored nodules in the skin, positive family history of the same lesions, and associations with extracutaneous abnormalities, especially cardiac. Shagreen patches appeared in the context of the TSC and they consist of flesh-colored plaques located specially in the lumbosacral area of young children. Eruptive collagenoma is an acquired entity similar to FCC but without family history or extracutaneous involvement [1,6].

Our case can be distinguished from the above mentioned since there was no family history of the same lesions, an absence of other TSC characteristics, and a lack of systemic involvement. The characteristic linear distribution differs from the above mentioned but the clinical differential

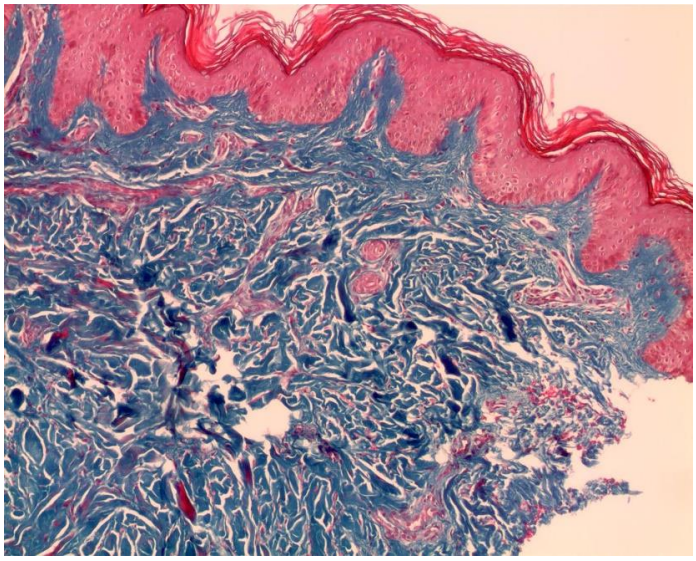

Figure 3. *Haphazardly arranged thick collagen bundles in the dermis. Trichrome, 100×.*

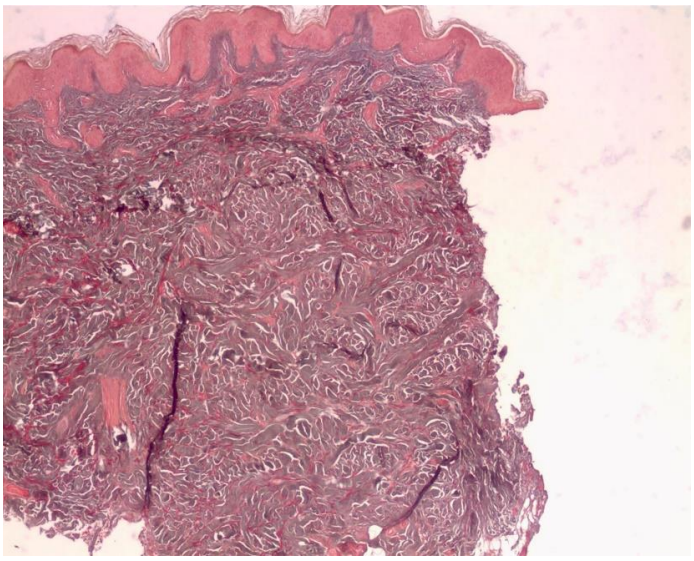

Figure 4. *Presence of dense collagen fibers with diminished elastic fibers. Orcein, 100×.*

diagnosis may include nevus lipomatosus superficialis or segmental neurofibromatosis which must be ruled out with histopathology [1,7].

Within the isolated collagenomas the reported presentations include paving stone nevus, papulolinear collagenoma, zosteriform collagenoma, and other nonspecific varieties. There are a few reports of isolated collagenomas but papulolinear distribution is extremely rare with only a few reports in the literature. Within the isolated collagenomas it does not have a dermatomal distribution; accordingly, this case cannot be classified as zosteriform collagenoma. It is not a paving stone nevus which is most commonly characterized by flesh-colored cerebriform tumors especially localized on the soles and almost always associated with Proteus syndrome [1,7-10]. The diagnosis of papulolinear collagenoma is most frequently made in children as it was in our patient. Lesions are similar as the ones described in literature but topography varies among different case reports [2-3].

Diagnosis is clinicopathologic, as in most cases is difficult to diagnose it by biopsy alone. As it is a benign entity, treatment should be conservative unless it is cosmetically unacceptable for the patient, in which case surgery is the best option. In our case no specific treatment was given [5].

Conclusion

Isolated papulolinear collagenoma is very rare. Diagnosis is made by clinicopathological correlation and as it is a benign and asymptomatic condition, and does not require specific treatment. We report this case because of its rarity.

Potential conflicts of interest

The authors declare no conflicts of interest.

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