Childhood solitary collagenoma

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ABSTRACT

Familial cutaneous collagenoma is an inherited connective tissue nevus, which presents with asymptomatic symmetrically distributed skin nodules on the trunk or upper limbs. Here, we describe a case of a 12-year-old girl with collagenoma affecting the lower back.

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Collagenomas are connective tissue nevi composed of excessive collagen deposition in the dermis, they may be familial or acquired. The familial type was first described in 1968 by Henderson et al,[1] with a an autosomal dominant mode of inheritance.[2] Patients usually present with multiple asymptomatic dermal nodules distributed symmetrically over the trunk or upper limbs, usually appearing during adolescence.[3,4] The acquired type of collagenomas may be eruptive or isolated. Here, we describe a case of collagenoma with review of literature.

Case Report. A 12-year-old Saudi girl presented to our dermatology clinic with asymptomatic skin colored grouped papular and nodular lesions affecting the lower aspect of her back. According to the mother, the lesions started to appear at the age of 7 years, and continued to increase in number till it reached to its present size. She had no history of previous skin trauma, inflammation or family history of similar condition. Her general health was good. Examination of the affected area revealed multiple cutaneous, rubbery, flesh-colored papules and nodules varying in size from 0.5-1 cm in diameter and coalescing to form oval shaped plaque ranging from 4-12 cm in size running horizontally across the lower back (**Figure 1**). There were no pigmentary changes and the rest of the skin was free of any lesion. Systemic examination was normal. So a punch biopsy of 6 mm was taken and it showed normal epidermis and adnexal structures and there were no signs of inflammation. The dermis was markedly thickened due to increased collagen deposition. In the upper and mid-dermis, an accumulation of dense, coarse, thick collagen fibers was present (**Figure 2**). Collagen bundles were variably oriented with many of them running perpendicularly to the skin surface. The collagen was strongly stained with Masson trichrome stain, showing an intense blue green color. The orcein stain for elastic tissue showed sparse, fragmented, elastic fibers that seemed to be diminished in number in the upper and mid-dermis. The Alcian-blue stain at pH 2.5 and the periodic acid Schiff stain showed no accumulation of mucopolysaccharides. Our most likely diagnosis was connective tissue nevus and specifically collagenoma.

Discussion. Connective tissue nevi are hamartomas characterized by an excess or deficit in the number of the cells or their biosynthetic products which include collagen, elastic fibers, and glycosaminoglycans.[5] Rocha and Winkelsmann[6] suggested that connective tissue nevi are hamartomas of collagen resulting in an alteration in the smooth

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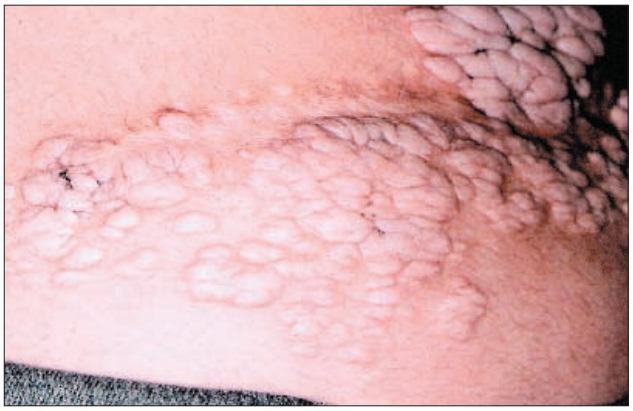

Figure 1 - Grouped papules and nodules running horizontally across the lower back.

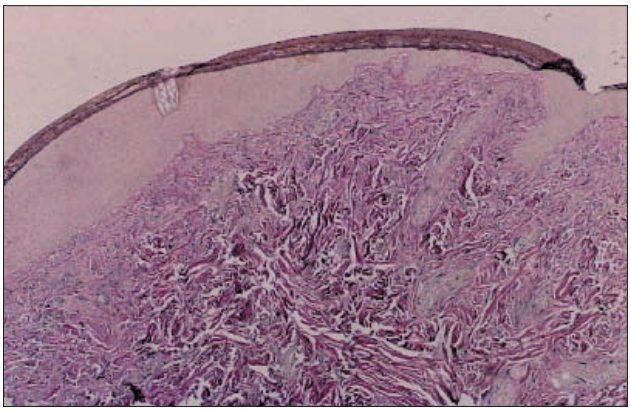

Figure 2 - Biopsy specimen from the lesion showing normal epidermis and accumulation of dense, coarse, thick collagen fibers in the upper and mid-dermis. (Hematoxylin and eosin x 100)

muscle, fat and elastic tissue balance while Pierard and Lapiere[7] classified nevi of connective tissue into 2 main basic subgroups according to the portion of the dermis principally involving: nevi of reticular connective tissue and nevi of the adventitial connective tissue. Uitto et al[8] 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 collagenomas, isolated collagenoma). Familial cutaneous collagenoma (FCC) is an inherited disorder first described by Henderson et al[1] in 1968. Later, an autosomal dominant inheritance was established by Uitto et al[2] contrasting with Hegedus and Schorr[9] who reported a female patient with similar nodules but without documented familial involvement. Clinically, FCC lesions consist of multiple symmetrical, asymptomatic papules, or nodules on the trunk and arms.[3,4] While individual lesions vary in diameter from few millimeters to few centimeters forming discrete, firm, indurated, flesh-colored, round or oval, and slightly elevated lesions. They usually appear during adolescence.[5] Cardiac abnormalities, in particular idiopathic progressive myocardiopathy with congestive heart failure and early R-wave transition can occur in some patients.[3,4] In fact, our case mostly fits into this category with absence of positive family history similar to the case reported by Hegedus and Schorr.[9] Acquired collagenomas are non-familial connective tissue nevi and include eruptive and isolated forms. Eruptive collagenomas have an abrupt onset and present as asymptomatic papulonodules on the trunk and arms while the isolated collagenoma presents as a single plaque on the same areas of the body.[5] Histologically, all collagenomas show an accumulation of dense, coarse collagen fibers in the dermis with an apparent reduction in the number of elastic fibers.[5,10] At present, there is no recommended effective therapy for those cutaneous lesions except the surgical removal of those apparent papules and nodules. In summary, this case could be a familial cutaneous collagenoma starting at an early age with absence of positive family history, which can be explained by the possible occurrence of new mutation in the affected person making it the second reported case in the literature or it can be re-designated into a newly reported solitary collagenoma in a child, which could be supported by further case reports.

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