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ERUPTIVE COLLAGENOMA, ORANGE, RUBBERY AND COLOURFUL: A CASE REPORT

Dermatology

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ABSTRACT

Connective tissue nevi of the skin are hamartomas consisting of components of the extracellular matrix, namely collagen, elastic fibres or proteoglycans. Collagenomas, are a type of connective tissue nevi characterized by hamartomatous proliferation of collagen fibres in the dermis, with normal, decreased or increased elastic fibres. Collagenomas, can be familial type or non-familial types which includes eruptive collagenoma and isolated collagenoma. We report a case of eruptive collagenoma, that presented to us with asymptomatic, multiple, firm, rubbery, papules without a positive family history or contributing systemic findings, where histopathologic examination using routine and special stains highlighted proliferation of collagen tissue with decrease elastic fibres.

KEYWORDS

collagen, collagenoma, hamartomas, connective tissue nevi.

INTRODUCTION:

Connective tissue nevi of the skin are hamartomas consisting of components of the extracellular matrix, namely collagen, elastic fibres or proteoglycans [1]. Collagenomas, also known as collagen nevi, are a type of connective tissue nevi characterized by hamartomatous proliferation of collagen fibres in the dermis, with normal, decreased or increased elastic fibres [2]. They typically present as asymptomatic skin coloured papules, plaques, nodules of varying sizes which can be grouped or solitary and may be distributed in a solitary, linear or segmental fashion. We report a case that presented to us with asymptomatic, multiple, firm, rubbery, papules where histopathologic examination using routine and special stains highlighted proliferation of collagen tissue with decrease elastic fibres.

CASE REPORT:

A 25-year-old male was referred for complains of multiple, white, asymptomatic lesions on trunk and both upper limbs. The lesions had an insidious onset and began to appear around 8 years ago but patient chose not to pursue any treatment because of the inconspicuous nature of the lesions. There was no history of trauma or preceding inflammation. There was no history of developmental delay, epilepsy or any other systemic complains. Family history was not contributory.
Clinical examination revealed multiple well define, discrete, firm to soft, white, non-tender papules and plaques, with an uneven surface, akin to skin of an orange, measuring around 2-3 millimetre (mm) in size, situated over the bilateral upper limbs and trunk (fig.1). Examination of hair, nails and mucosa revealed no abnormalities. Systemic examination was uneventful. Differential diagnosis that struck us were, Histoid Hansen's, smooth muscle hamartoma, neurofibroma, elastofibroma and collagenoma. To be sure of the quandary, we proceeded to go for a 4mm punch biopsy after taking an informed consent of the patient.

Histopathologic examination using haematoxylin and eosin stain (fig.2), revealed normal epidermis with a thickened reticular dermis showing haphazardly arranged thickened collagen bundles indicative of collagen nevus. To further highlight the abundance of collagen, special stains like Masson's trichrome and Verhoeff-Van Gieson were used (fig.3, fig.4), which highlighted the paucity of elastic fibres. Thus, we reached a final diagnosis of eruptive collagenoma.

We explained the benign nature of the disease along with available treatment options to the patient, who later expressed, not to pursue any treatment due to asymptomatic nature of his disease.

DISCUSSION:

Uitto *et al*. classified collagenoma based on the pattern of distribution (localised or generalized) and mode of inheritance (familial and non-familial types.) [3]. Familial type includes cutaneous collagenomas associated with cardiological abnormalities, which is inherited in an autosomal dominant pattern and shagreen patches associated with tuberous sclerosis. Non-familial types include eruptive collagenoma and isolated collagenomas.

Eruptive collagenoma, was first described by Cramer in 1966 [4], it generally presents as single or multiple, firm to soft, well defined skin coloured papules and nodules of varying sizes, but generally is less than 1 cm in diameter. Onset is mostly around the first or the second decade of life without any contributing family history or systemic findings.

Pathogenesis of eruptive collagenoma remains elusive. A study done by Uitto et a [5], demonstrated that collagenoma consist almost exclusively of type I collagen and the underlying pathogenic mechanism maybe decreased production of collagenase, leading to decrease degradation of collagen. Some studies have also implicated hormones to have a possible etiologic role in pathogenesis as collagenomas may grow in size during pregnancy or during puberty [6] [1].

On histopathologic examination, lesions are characterized by an excessive accumulation of dense collagen bundles arranged randomly in the dermis. It is postulated that elastic fibres appear reduced or absent due to a dilution phenomenon caused by excessive collagen accumulation [7] [1].

Eruptive collagenoma should be differentiated from other collagenomas like familial cutaneous collagenoma, Isolated collagenomas, nevus anelasticus and papular elastorrhexis.

Familial cutaneous collagenoma, was first described by Henderson *et al*. in 1968. It is characterized by appearance of lesions similar to eruptive collagenoma, that generally begin to appear around the third decade of life, however unlike eruptive collagenoma, it is associated with a positive family history and an associated systemic involvement [8].

Isolated collagenomas are generally acquired sporadically or associated with other diseases like Proteus syndrome or Turner syndrome and are mostly confined to a single body region. It can present as planter cerebriform, linear or zosteriform, knuckle pads and papulo-linear lesions [2].

Nevus anelasticus, presents as multiple, flat firm, perifollicular papules over the trunk. Papular elastorrhexis is considered to be a variant of nevus anelasticus, it presents as multiple asymptomatic, small, white

papules, distributed over the trunk and extremities but without a predilection for the perifollicular areas [9]. On histopathologic examination, both these entities show focal area of decreased and fragmented elastic fibres. Some authors consider eruptive collagenoma, nevus anelasticus and papular elastorrhexis to be a single disease or part of a disease spectrum owing to similar clinical and histopathologic features [10]. Opinions however, differ among experts.

There is no specific treatment, however reports suggest successful use of intralesional corticosteroids to help flatten the lesion. Al be it, it seems wise to be cautious about use of intralesional steroids as there is an associated risk of exaggerated dermal atrophy due to absent or decreased elastic fibres in this disorder.

Our patient was diagnosed as a case of eruptive collagenoma since the lesions presented around the second decade of life, without a positive family history or contributing systemic findings. Histopathology using routine as well as special stains helped in identifying randomly distributed thickened collagen bundles with paucity of elastic fibres.

Eruptive collagenoma is a relatively uncommon disorder coupled with the predicament of fewer case reports in literature. Therefore, there is a need to bring cases like these in spotlight among practicing luminaries along with a more comprehensive research to better understand the pathogenesis of this elusive condition.

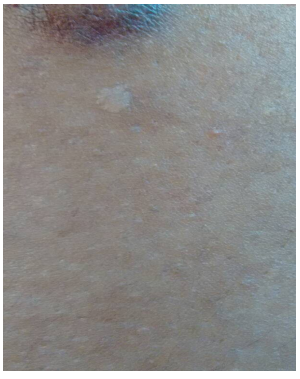

Figure 1: Well defined, discrete, white, plaque, with surface similar to skin of an orange, situated over the trunk.

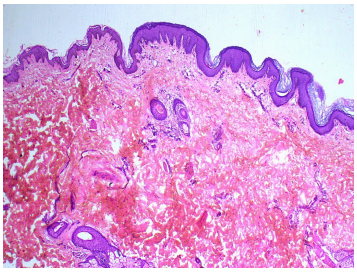

Figure 2: 10x view using haematoxylin and eosin stain demonstrates normal epidermis with a thickened reticular dermis along with haphazardly arranged thickened collagen bundles.

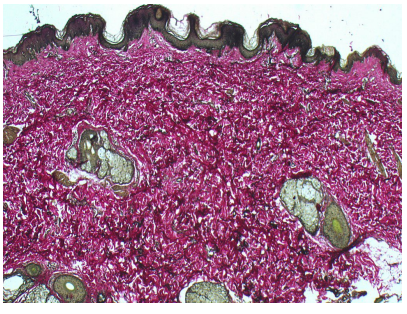

Figure 3: 10x view using Verhoeff-Van Gieson staining shows a normal epidermis along with thickened collagen bundles and scarcity of elastic fibres.

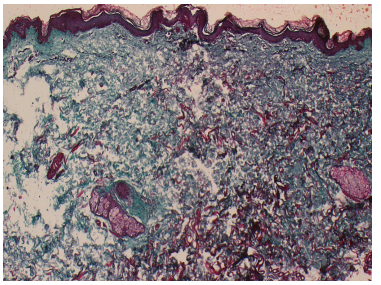

Figure 4: 10x view using Masson's trichrome stain reveals, normal epidermis along with paucity of elastic fibres.

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