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Case Report

Atrophic wrinkled plaque on face: a diagnostic dilemma

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ABSTRACT

We report an interesting case of an eight year old boy presenting with a depressed atrophic wrinkled plaque on cheek for last three years. The clinical finding of wrinkling and soft and pulpy consistency of lesion raised the possibility of elastolytic disorder but was ruled on the basis of normal elastic tissue on Verhoff stain. The histopathological findings were consistent with atrophoderma, but the lesion lacked the classical clinical features, thereby posing a diagnostic dilemma.

Keywords: Atrophoderma, Variant, Face, Wrinkle, Elastolysis

INTRODUCTION

Idiopathic atrophoderma of Pasini and Pierini is a form of dermal atrophy that presents as one or several sharply demarcated brown or violaceous depressed patches, usually on the back of adolescents or young adults. The surface and consistency of the lesion is normal. Herein we describe an unusual case of an eight year old boy presenting with depressed atrophic wrinkled plaque on face which has histological features of atrophoderma but clinically resembling elastolytic disorder, thus posing a diagnostic dilemma.

CASE REPORT

An eight year boy, first full term uncomplicated vaginal delivery child born of non-consanguineous marriage, presented to us with an asymptomatic round depressed lesion on left cheek for last 3 years. There was no history of trauma, infection, ulceration, urticaria, topical or intralesional drug administration, drug allergy. According to the parents, the lesion progressed very slowly and there was no history of ivory white indurated lesion with lilac border. The child was otherwise healthy and family history was not contributory.

On cutaneous examination, there was a single, ill defined, round depressed plaque of size approximately 3×3 cm with fine wrinkles on the lower part present on central region of left cheek as given in Figure 1.

Figure 1: (a) a round depressed plaque on left cheek, (b) slight textural changes on right cheek.

On close careful examination, telangiectasia was seen on the lesion and the skin surrounding the lateral and lower part of the lesion showed mild reticular erythema as shown in Figure 2. On palpation, the lesion was non

tender, non-indurated and felt soft and pulpy. Examination of the right cheek revealed a similar sized ill-defined plaque with irregular surface resembling an orange peel and surrounded by reticular erythema. Nail, hair and mucosa were normal. As per the pediatric referral, systemic examination was normal and child's physical and mental development was appropriate for age. Differential diagnosis of acquired elastolytic disorders (such as cutis laxa acquisita, anetoderma, mid dermal elastolysis) and atrophoderma were kept.

Routine laboratory investigations including complete blood count, ESR, liver and kidney function tests, chest X-ray, ECG were normal. ANA was negative. Skin biopsy showed slightly atrophic epidermis, hyalinization of collagen in deep dermis and mild perivascular mononuclear infiltrate. Appendages and subcutaneous layer were normal as in Figure 3. Normal unfragmented elastic tissue was seen in the entire dermis on Verhoff – Van Gieson stain as shown in Figure 4. The histopathology rules out elastolytic disorder.

Diagnosis of atrophoderma presenting as a variant was made on clinico-histopathological correlation. Patient was prescribed mild topical steroid.

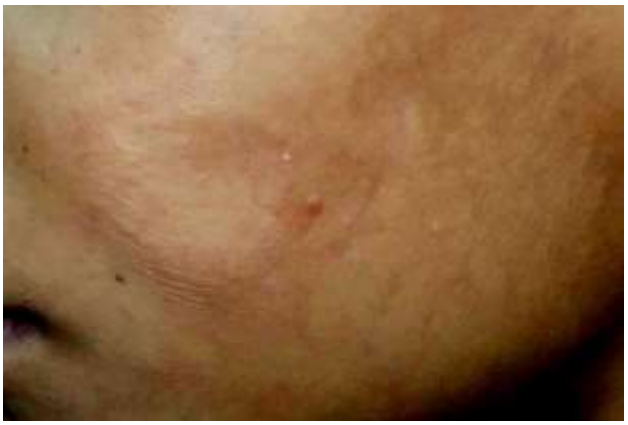

Figure 2: Close up of lesion showing fine wrinkles, telangiectasia and mild reticular erythema.

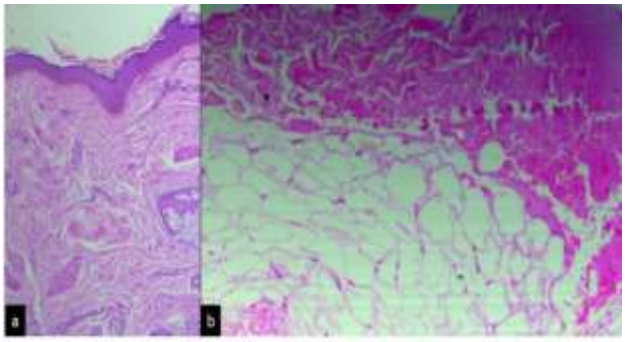

Figure 3: (a) Slight epidermal atrophy with loss of rete ridges (H and E stain, 100 X), (b) homogenization of collagen in deep dermis and normal subcutaneous tissue (H and E stain, 200 X).

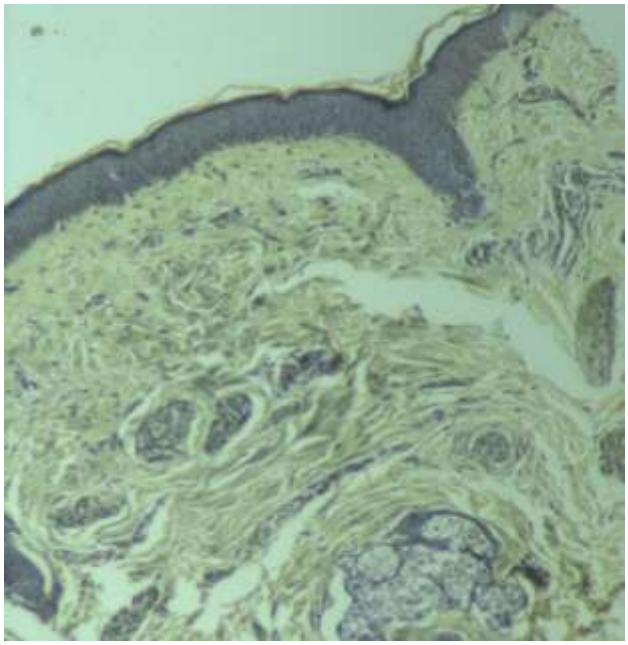

Figure 4: Normal elastin fibers in the dermis (Verhoff Van Gieson stain, 200 X).

DISCUSSION

First described by Pasini in 1923, atrophoderma of Pasini-Pierini is a rare disorder of connective tissue of unclear etiology.[1] It is clinically characterized by single or multiple asymptomatic, depressed, sharply defined round to oval plaques which may be hyper pigmented.[2] There is no associated induration or secondary changes and surface and consistency of the lesion is normal. The surrounding skin is entirely normal and the abrupt edge of atrophoderma is often described as cliff-drop border or footprints in the snow. The back is almost always involved, the chest and abdomen frequently, and the proximal parts of the limbs occasionally. Usual presentation is in the second or third decade and the lesions may gradually increase in number, coalesce to form large plaques and eventually remain unchanged. Female preponderance is seen with ratio 6:1.[3] The classification of atrophoderma is an unsettled issue with some considering it as a separate entity and some believing it to be an abortive variant of morphea as it resembles the late stage of morphea without undergoing the initial stages of induration and inflammation. The histopathology shows subtle non-diagnostic changes with normal or slightly atrophic epidermis, increased pigmentation of basal layer, varying degree of homogenization of collagen bundles and mild perivascular lymphocytic and histiocytic infiltrate.

Acquired elastolyic disorders, also rare disorders of connective tissue, are characterized by decrease or absence of elastin in the dermis and normal collagen. They include cutis laxa acquisita, anetoderma and mid dermal elastolysis. In anetoderma, there is a wrinkled atrophic plaque which yields on pressure. Localized cutis

laxa is clinically indistinguishable from anetoderma. Mid dermal elastolysis (MDE) is an acquired condition seen mostly in middle aged females and presents in three clinical patterns.[4] Type 1 MDE has patches of fine wrinkles and in type 2 patterns there are perifollicular popular protrusions resembling peau-d-orange appearance. A rare variant referred to as type 3 MDE presents as reticular erythema on upper trunk.[5] Trunk and proximal extremities are the most commonly involved sites. Although etiology is not fully known, it is associated with excessive sun exposure, multiple autoimmune disorders and inflammatory conditions.

The case described here has a round depressed non indurated plaque which suggests atrophoderma. However, presence of fine wrinkles and reticular erythema in the peri-lesional skin and soft and pulpy consistency of the lesion partially favor the diagnosis of elastolytic disorders. Moreover, the classical cliff drop border and hyperpigmentation of atrophoderma is absent. The lesion is rather ill defined and of skin color. Also, the site of the lesion (face) is perplexing as face, hand and feet are invariably spared in atrophoderma and rarely involved in elastolytic disorders.[6] The lesion on the contralateral cheek, unnoticed by the parents, probably represents the early stage of the disease pathogenesis.

To summarize, the finding of normal elastic fibers on special stain rules out elastolytic disorders and the clinical findings do not entirely correspond with classic atrophoderma. As the diagnosis of atrophoderma is primarily a clinical one and the histopathological features are consistent, the case described here could be considered as a novel variant of atrophoderma presenting at an unusual site in a male patient at an unusual age.

Previous reports have shown uncommon patterns of atrophoderma such as atrophoderma elastolytica discreta, unilateral and zosteriform patterns.[7-9] Also, hypo-pigmentation instead of hyperpigmentation and lesions over the extremities have been described.[10] However, we could not find literature on involvement of face in atrophoderma.

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