Journal of Pediatric Sciences

Case Report:
A Child with Acquired Reactive Perforating Collagenosis

Farah Sameem, Sheikh Javeed Sultan, Ummer Yaseen

Journal of Pediatric Sciences; 2010; 2(2); e36

How to cite this article:

Sameem F., Sultan S.J., Yaseen U. Case Report: A Child with Acquired Reactive Perforating Collagenosis. Journal of Pediatric Sciences 2010;2(2):e36

CASE REPORT

Case Report:
A Child with Acquired Reactive Perforating Collagenosis

Farah Sameem[1], Sheikh Javeed Sultan[2], Ummer Yaseen[1]

Abstract:
A nine year old boy presented with a four year history of a pruritic papular eruption predominantly involving the face. Histopathological examination revealed extrusion of collagen through the epidermis with no inflammation of the dermis favouring a diagnosis of reactive perforating collagenosis.

Key words: Perforating dermatosis, Koebners phenomenon, Reactive perforating collagenosis
Received: *13/06/2010; Accepted: 07/07/2010*

Introduction

Perforating dermatosis are a heterogeneous group of disorders characterized clinically by papulonodular lesions with keratotic plugs or crusts and by elimination of some dermal connective tissue component through a normal looking epidermis [1]. First described by Mehregan *et al*. reactive perforating collagenosis (RPC) is a type of perforating disorders characterized by elimination of altered collagen through the epidermis [2]. It occurs in both inherited and acquired forms [3,4]. We report a nine year old male child with RPC with predominant facial involvement.

Case Report

A nine year old boy product of a non-consanguineous marriage, with one normal sibling presented with multiple painless, skin coloured and reddish lesions on the face and limbs since the last four years (Figure 1). There was history of pruritus, but no history of photosensitivity or summer exacerbation. The child was born of a fullterm, normal delivery and had normal developmental milestones. There was no other significant family or medical history. Height and weight were normal for age. General physical and systemic examination was non contributory. On cutaneous examination multiple, bilaterally symmetrical, erythematous to skin colored papules, in different stages of development, with central umbilication in few and keratotic plugging in others were seen on extensor aspect of knees, elbows and trunk but predominantly on cheeks. A few residual scars were also visible.

Farah Sameem[1], Sheikh Javeed Sultan[2], Ummer Yaseen[1]

[1]Government Medical College, Srinagar
[2]SKIMS Medical College, Srinagar

Corresponding Author:
Sheikh Javeed Sultan, MD

Shalimar – Srinagar Kashmir (J & K) India. 191121
e-mail: sjsultan@gmail.com

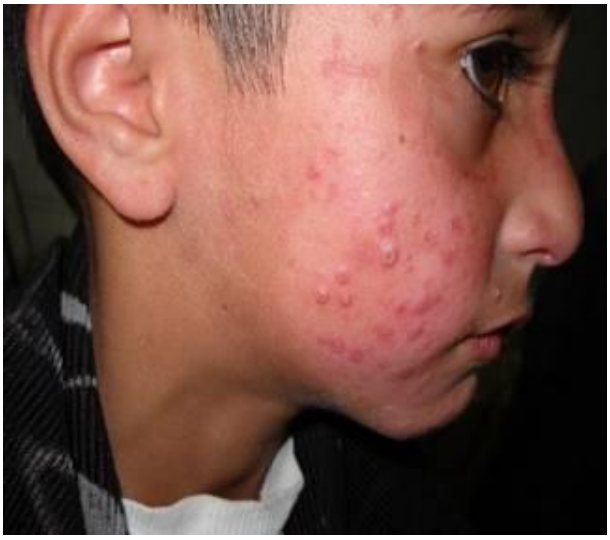

Fig 1. Erythematous papules with umblication and keratinous plug.

No exudation, vesiculation or pustules were seen. Marked Koebnerisation was evident. Nails, hair and teeth were normal. Hemogram, liver function tests, kidney function tests, thyroid function tests, blood sugar, chest X-ray and abdominal ultrasonography were normal. Histopathological examination (Hematoxylin and eosin staining) of a skin biopsy specimen showed a cup-shaped area of depression of epidermis filled with plug of parakeratotic keratinocytes, few inflammatory cells, necrotic material, and fine fibers (Figure 2).

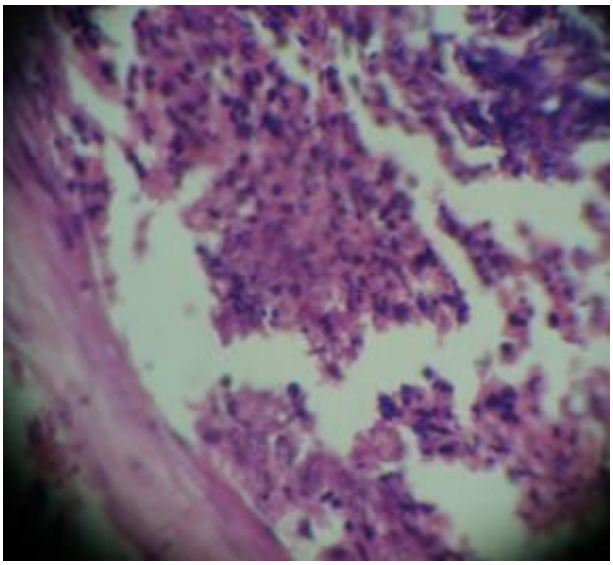

Fig 2: Showing collagen band engulfed and surrounded by focal epidermal proliferation.

The fibers stained positive for collagen. In view of these clinical and histopathological findings, a diagnosis of reactive perforating collagenosis was made with marked facial involvement and koebnerisation. The patient was advised to avoid trauma and was given topical retinoids (tretinoin 0.025%) for three weeks with a good response.

Discussion

Reactive perforating collagenosis is a rare skin disorder characterized by elimination of altered collagen through the epidermis [1,2]. It occurs in two distinct forms- an inherited form manifesting in childhood and an acquired sporadic form occuring in adulthood [1,3,4]. Both autosomal dominant and autosomal recessive modes of inheritance have been reported for the inherited form [3,4]. The acquired form usually occurs in patients with diabetes or chronic renal failure; other systemic associations such as hypothyroidism, hyperparathyroidism and liver dysfunction; malignancies (including lymphoma, hepato-cellular carcinoma, peri-ampullary carcinoma, and thyroid cancer); and drugs (Indinavir) have also been reported [4-9]. There are no racial variations in the incidence of this disorder and it occurs equally in both sexes.

Typical lesions of reactive perforating collagenosis are flesh-colored, umbilicated, dome-shaped papules or nodules with an adherent, keratinous plug. These may occur in a linear configuration, exhibiting the Koebner phenomenon. The lesions are intensely itchy, sometimes leaving residual scarring after resolution. They occur most commonly on the extensor surfaces of the limbs and the dorsa of the hands but may also occur on the trunk and the face. The course of the disease is variable. Lesions may regress spontaneously in a few weeks or persist chronically for years [1-4].

The major abnormality in reactive perforating collagenosis is focal damage to collagen and the elimination of the disrupted collagen through the epidermis [10]. The underlying cause is unknown, but an abnormal response to superficial trauma has been suggested. Lesions have been reported following scratches, insect bites and scabies [11-13].

The histological findings in reactive perforating collagenosis are variable depending upon the stage of the lesion. In early lesions epidermal hyperplasia is seen associated with underlying degenerate basophilic collagen fibers. Well established lesions reveal a cup-shaped depression of the epidermis associated with a keratin plug containing parakeratosis, inflammatory debris and collagen fibers. [1-3].

The unique features in our case were the young age, marked koebnerisation, predominant facial involvement, absence of positive family history and absence of any other systemic association. Hydroa vacciniforme and lichen planus were considered in the differential diagnosis. Hydroa vacciniforme was excluded by the absense of vesiculation, extension of lesions beyond the face and absence of photoaggravation. Lichen planus was excluded by the characteristic histopathology. Hence a diagnosis of reactive perforating collagenosis was made.

References

1. Rapini RP. Perforating diseases In: Bolognia JL, Jorizzo JL, Rapini RP (eds). Dermatology 2nd ed. Elsevier 2008:1461-67.

2. Mehregan AH, Schwartz OD, Livingood CS. Reactive perforating collagenosis. Arch Dermatol 1967;96:277-82.

3. Ramesh V, Sood N, Kubba A, Singh B, Makkar R. Familial reactive perforating collagenosis: a clinical, histopathological study of 10 cases. J Eur Acad Dermatol Venereol. 2007;21:766-70.

4. Faver IR, Daoud MS, Su WP. Acquired reactive perforating collagenosis. Report of six cases and review of the literature. J Am Acad Dermatol. 1994;30:575-80.

5. Pedragosa R, Knobel HJ, Huguet P, Oristrell J, Valdıs M, Bosch JA. Reactive perforating collagenosis in hodgkin's disease. Am J Dermatopathol 1987;9:41-4.

6. Kiliç A, Gönül M, Cakmak SK, Gül U, Demiriz M. Acquired reactive perforating collagenosis as a presenting sign of hepatocellular carcinoma. Eur J Dermatol. 2006;16:447.

7. Chae KS, Park YM, Cho SH, Cho BK. Reactive perforating collagenosis associated with periampullary carcinoma. Br J Dermatol. 1998;139:548-50.

8. Yazdi S, Saadat P, Young S, Hamidi R, Vadmal MS. Acquired reactive perforating collagenosis associated with papillary thyroid carcinoma: a paraneoplastic phenomenon? Clin Exp Dermatol. 2009;35:152-55

9. Calista D, Morri M. Acquired reactive perforating collagenosis induced by indinavir in 2 patients with HIV disease. Eur J Dermatol. 2008;18:84-5.

10. Millard PR, Young E, Harrison DE, Wojnarowska F. Reactive perforating collagenosis: light, ultrastructural and immunohistological studies. Histopathology. 1986;10: 1047-56.

11. Bovenmyer DA. Reactive perforating collagenosis. Experimental production of the lesion. Arch Dermatol. 1970;102:313-7.

12. Kim EJ, Kim MY, Kim HO, Park YM. Acquired reactive perforating collagenosis triggered by insect bite. J Dermatol. 2007;34:677-9.

13. Ikezaki E, Sugita K, Kabashima K, Tokura Y. Scabies-induced acquired reactive perforating collagenosis. J Eur Acad Dermatol Venereol. 2008;22:120-1.