

Reactive Perforating Collagenosis in Two Siblings – A Rare Case Presentation

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Abstract: Reactive perforating collagenosis (RPC) is characterized by transepidermal elimination of altered collagen through the epidermis. Two distinct forms of RPC are known ; rare inherited form that starts in early childhood and a common acquired form that occurs in adulthood. We report a rare case of inherited form of RPC in a female child with presence of similar lesions in her elder brother .A 9 year old female child presented with extensive dark colored skin lesions over face and extremities since 6 months. On dermatological examination , multiple closely studded skin colored and hyperpigmented crateriform lesions with central crusting were present on dorsum of hand. Discretely arranged papules with central adherent keratotic plugging were seen over trunk and extremities. Histopathological examination revealed invagination of the epidermis with necrotic inflammatory material and collagen fibres which are seen at the base of the lesion with transepidermal elimination . Masson's Trichrome positive collagen fibres were seen within the perforation. All routine laboratory parameters were within normal limits. The patient was treated with emollients ,tretinoin cream along with oral isotretinoin which showed excellent results on follow up. Patients' 11 year old brother showed similar clinical features and good response was seen with treatment. Familial RPC should be considered in all cases where children present with umblicated papules and keratotic plugs. To confirm the diagnosis histopathologically, special stains for collagen fibres(masson's trichrome and verhoeff-van gieson) should be employed along with H&E staining.

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I. Introduction:

- Reactive perforating collagenosis (RPC) is characterized by transepidermal elimination of altered collagen through the epidermis.
- Two distinct forms of RPC are known ; rare inherited form that starts in early childhood and a common acquired form that occurs in adulthood.
- We report a rare case of inherited form of RPC in a female child with presence of similar lesions in her elder brother.



CASE REPORT:

- A 9 year old female child presented with extensive dark colored skin lesions over face and extremities since 6 months.



- On dermatological examination , multiple closely studded skin colored and hyperpigmented crateriform papules with central crusting were present on dorsum of hand.
- Discretely arranged papules with central adherent keratotic plugging were seen over trunk and extremities.
- Koebners phenomenon was seen .
- Papules with central umbilication was noticed.



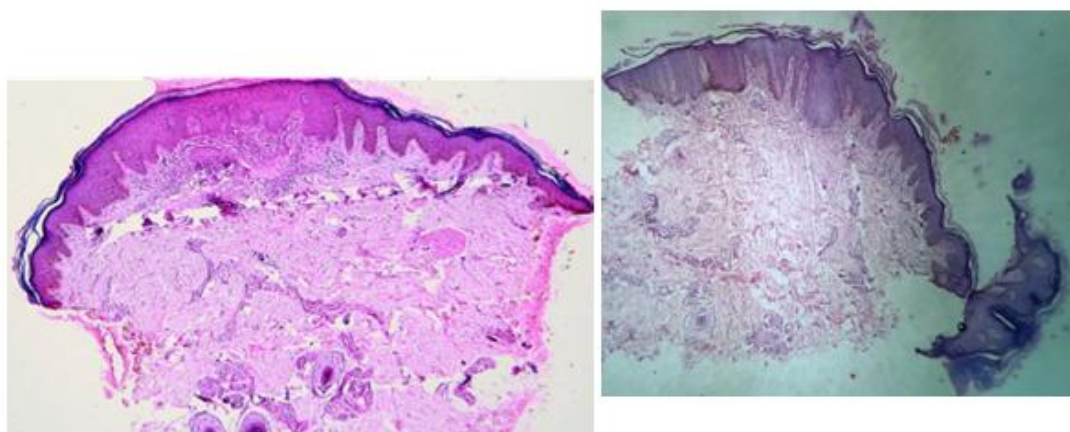
- Patients 11 year old brother also presented with similar keratotic papules over the face trunk and extremities.



- Based on the clinical features , a differential diagnosis Perforating folliculitis , Follicular lichen planus and Reactive perforating collagenosis was considered.

INVESTIGATIONS:

- Laboratory workup : Complete blood count , serum biochemistry profile was within normal limits
- Mantoux , chest x-ray and usg-abdomen did not reveal any abnormality.
- SKIN BIOPSY:
- Histopathological examination revealed invagination of the epidermis with necrotic inflammatory material and collagen fibres which are seen at the base of the lesion with transepidermal elimination
- Masson's Trichrome positive collagen fibres were seen within the perforation.



- These features suggestive of Reactive Perforating Collagenosis (RPC).
- Histopathological features in sibling was also in favour of RPC.

TREATMENT

- Cap.Isotretinoin -20mg daily for 1 month followed by another 6 weeks at night.
- Topical emollient containing urea , lactic acid was given twice daily.
- Multivitamin supplementation.
- Excellent improvement with resolution of all keratotic papules was observed after 8 weeks of treatment in both patient and her sibling.

II. Discussion:

- RPC presents in 2 forms - Rare inherited-familial form and acquired form .
- Acquired form may be associated with diabetes and hemodialysis

- It is usually precipitated by environmental factors such as cold or trauma : Abnormal response to trauma results in focal damage of collagen which is extruded.
- Usually umblicated papules with keratinous plug are seen over extensor surfaces of the extremities which regress spontaneously over 8 to 10 weeks scar formation.
- Histopathology shows Vertically oriented perforating bundles of collagen are present at the base of the invagination.
- Topical Retinoic acid , topical steroids under occlusion , oral isotretinoin and phototherapy was used with satisfactory results.

III. Conclusion :

- Familial RPC should be considered in all cases where children present with umblicated papules and keratotic plugs.
- To confirm the diagnosis histopathologically, special stains for collagen fibres(masson's trichrome and verhoeff-van gieson) should be employed along with H&E staining.

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