Original Research Paper

Volume - 14 | Issue - 04 | April - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Internal Medicine

REACTIVE PERFORATING COLLAGENOSIS IN A YOUNG FEMALE – A CASE REPORT

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ABSTRACT This is a case report about 28-year-old female with multiple ulcero-nodular lesions with relapsing and remitting course diagnosed with reactive perforating collagenosis. These ulcerative lesions occurring at sites of minor/ trivial trauma. The histopathological diagnosis of these ulcerative lesions – Reactive perforating collagenosis has a characteristic transepidermal elimination of altered collagen through the epidermis. This very rare dermatological condition has been discussed in the case report.

KEYWORDS : Abnormal collagen; Elimination; Collagenosis; Central keratotic plug

INTRODUCTION:

Reactive perforating collagenosis is a very rare inherited disease. Fewer than 50 cases have been reported in the literature since it was first described in 1967. Most patients will have a relapsing and remitting course of lesions throughout their life. The lesions will most commonly resolve spontaneously in six to eight weeks, leaving behind a hypopigmented area or scar. The patient will most likely complain of pruritus of the lesions.

CASE REPORT:

A 28-year-old female with no comorbidities had ulcerative skin lesions all over the body for 7 months. She is a non-pregnant married woman with no children or abortion. Her menstrual cycles were in normal cycle and flow. She does not have any history of fever, rash, loss of weight or appetite, joint tenderness, or bite marks.

She had multiple discrete ulcerating papulonodular lesions; which were increasing in size over the time distributed in extremities as well as trunk. These lesions evolve form sites of minor trauma/ abrasions. A large dark keratotic plug was found in the central portion of the ulcer. There was no accompanied induration around the ulcer. No signs of inflammation such as redness or tenderness. Sensation was intact. Ulcers were associated with Intense pruritis.

Few weeks back she was treated with dapsone in view of high clinical suspicion for Hansen's disease. Methylprednisolone has also been given. However, there was no clinical resolution on ulcers with medications.

Her nutritional status was fairly good. After examining the patient; basic investigations have been carried out. There was mild normocytic normochromic anemia due to variant hemoglobin. Creatinine – 0.6; AST/ALT in normal limits with maintained Albumin / Globulin ration (4.3 / 3.3); Mild elevation of GGTP - 82; No proteinuria or microhematuria. X ray chest and USG of abdomen were normal. She was clinically euthyroid but mild elevation of laboratory TSH.

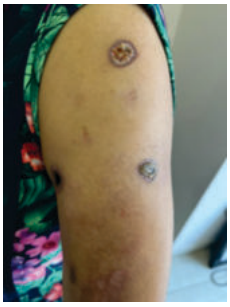

Figure 1

Her ulcer has been biopsied and stained which showed subacute inflammatory exudate with debris and trans epidermal elimination of collagen; Perivascular inflammatory infiltrate and fibrosis. BY Histopathological examination, diagnosis of Reactive perforating collagenosis has been made.

She has been managed with colchicine; antipruritic agent and antibiotics. Retinoids has not been tried as she is a newly married young lady.

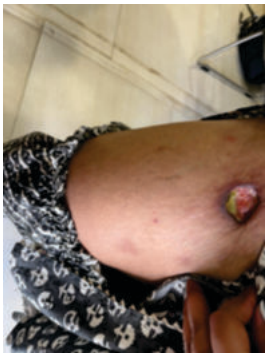

Figure 2

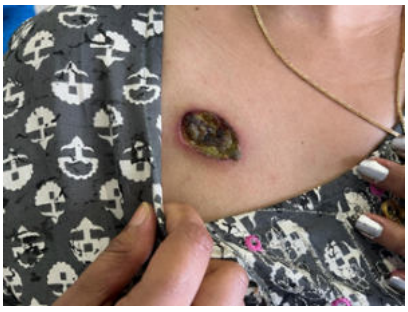

Figure 3

All the above figures 1,2 and 3 representing evolving ulcerative lesions with large central keratotic plugging.

DISCUSSION:

Reactive perforating collagenosis has a characteristic trans epidermal elimination of altered collagen through the epidermis. Two forms has been identified; an inherited form in childhood (very rare) and an acquired form (adulthood). Adult form more commonly found in patients with diabetes mellitus and end-stage renal disease.

Perforating dermatosis is a skin disease that is characterized by the

trans epidermal elimination of material in the dermis that can be divided into two groups: primary and secondary. Primary perforating dermatoses include reactive perforating collagenosis, Kyrle disease, elastosis perforans serpiginous, and perforating folliculitis. Secondary perforating dermatoses was coined by Rapini in 1989 and is called acquired reactive perforating collagenosis. Commonly, patients will have hyperkeratotic papules on the extensor areas of extremities and hands, most likely over areas of superficial trauma. The lesions are very pruritic and can grow into large, umbilicated papulonodules with a central keratotic plug that relapse and remit over the span of the patient's lifetime. Most cases are self-limited and do not require treatment. Patients can be treated with avoidance of superficial trauma and control of pruritus in most cases.

Pathophysiology:
No specific gene defect has been found. Overexpression of TGF-Beta has been hypothesized. Serum and tissue concentrations of fibronectin have been reported to be elevated, which may contribute to epithelial migration. Superficial trauma, as well as a cold environment, induces degeneration of collagen. The histology of reactive perforating collagenosis classically is trans epidermal. Vertically oriented perforating bundles of basophilic collagen are present at the bases of the invagination of the epidermis with focal extrusions. A keratin plug most likely is found in the cupping of the epidermis. Histology is variable depending on the stage of the lesion. New lesions will show epidermal hyperplasia, degenerated basophilic collagen fibers with later lesions showing invagination of the epidermis with a keratin plug. The vertically oriented basophilic collagen fibers are highlighted with Verhoeff-Van Gieson staining, which stains the collagen fibers red. This differentiates reactive perforating collagenosis to elastosis perforans serpiginosa, which will not show any staining of elastic fibers.

The small, hyperkeratotic papules of reactive perforating collagenosis can be present in any area of the body. The inherited form begins in infancy as papules located on the extensor surfaces of the hands, elbows, and knees, most likely after superficial trauma to the area. Reactive perforating collagenosis lesions then grow into larger, umbilicated papulonodules with central adherent keratin plugging. The lesions will most commonly resolve spontaneously in six to eight weeks, leaving behind a hypopigmented area or scar. The patient will most likely complain of pruritus of the lesions. Koebner phenomenon of the lesions can occur.

Management:
Reactive perforating collagenosis lesions are commonly self-limited and rarely problematic. The mainstay of treatment is avoiding trauma to decrease risk factors for acquiring reactive perforating collagenosis lesions. Treatment is also aimed at controlling pruritus with topical corticosteroids, emollients, and systemic antihistamines. The use of keratolytics, topical and oral retinoids, phototherapy, liquid nitrogen, methotrexate, and allopurinol have been reported without much improvement of the lesions. No randomized controlled trials have evaluated the treatments for reactive perforating collagenosis, and all treatment suggestions are the result of case reports. It appears that treatment is patient-specific and can vary with the degree of severity of the lesions. Most lesions will regress spontaneously in six to eight weeks with hypopigmentation, scarring, or post-inflammatory hyperpigmentation. Unfortunately, most patients will have a relapsing and remitting course of lesions throughout their life.

Declaration:
Funding- None

Conflict Of Interest- None

Ethical Approval- Not required

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