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Case Report

Erythema elevatum diutinum - Case report of a rare entity

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ABSTRACT

Erythema elevatum diutinum (EED) is a rare entity of chronic cutaneous vasculitis occurring in association with a variety of conditions including autoimmunity, infectious disease, and hematological abnormalities. The exact pathogenesis of EED is unknown.

A 59-year old female presented with multiple, itchy, eczematized erythematous papules and plaques over both upper and lower limbs present since 5 months. Histopathological changes were consistent with EED. Patient was started on oral Dapsone 100mg bid along with topical betamethasone diproprionate cream. Within 6 weeks lesions regressed completely leaving post-inflammatory hyperpigmentation. Dapsone was stopped at the end of 3 months. Minor recurrences are managed by topical steroids.

Key Messages: EED is a rare form of vasculitis which is difficult to diagnose clinically, histopathology can be diagnostic. Dapsone is the treatment of choice.

1. Introduction

Erythema elevatum diutinum (EED) is a rare, chronic, cutaneous leukocytoclastic vasculitis characterized by violaceous to red -brown papules and nodules on extensors of extremities.[1] The first description of the disease was by Hutchinson and Bury in the 1880s and the condition was later named in 1894 by Radcliffe-Crocker and Williams.[2] EED is most commonly seen in adults although occasional childhood cases are reported with no gender and ethnic groups predilection.[2] It has been associated with many infections, hematological disorders, connective tissue disorders, and inflammatory disorders. IgA monoclonal gammopathy is the commonest association and myeloma has the strongest association. EED may precede years before hematological abnormality.[2]

2. Case Report

A 59- year old female patient presented with itchy reddish skin lesions over arms, legs, buttocks and lower trunk present since 5 months. Lesions started on forearms bilaterally and gradually involved other areas.

Patient was diabetic and hypertensive and was controlled on metformin and amlodipine since 5 years. She had no history of similar lesions in the past and had no significant past history of any cutaneous or systemic disease other than diabetes and hypertension. No complain of joint pains or fever was present. Patient had consulted dermatologist and was prescribed topical steroids without any significant improvement in lesions.

On clinical examination, multiple, erythematous, papulo-nodular lesions coalescing into plaques at places were present over extensor aspects of bilateral lower and upper limbs, lower abdomen and gluteal region.(Figure 1 a,b). No oral or genital mucous membrane lesions were seen. No hair or nail changes were evident. No hepato-splenomegaly or

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lymphadenopathy were evident.

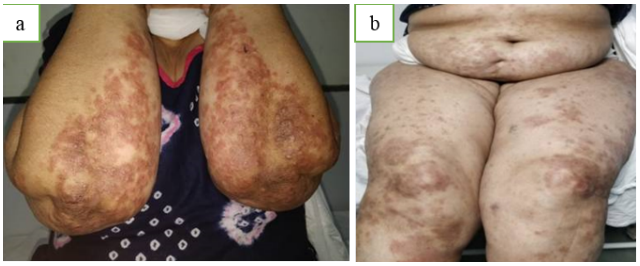

Fig. 1: a,b:

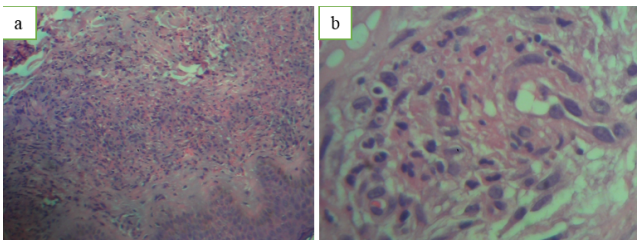

Fig. 2: a,b

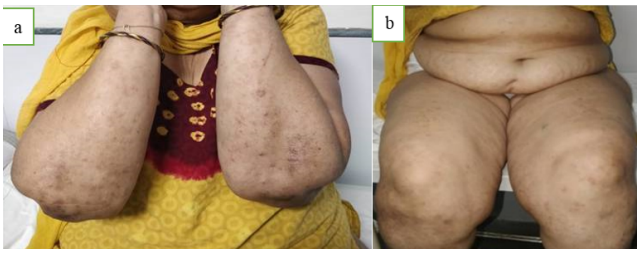

Fig. 3: a,b

All the routine haematological investigations including lipid profile were within reference range. Antinuclear antibody titre, HIV, hepatitis B & c serologies were negative. Rapid plasma reagin test was nonreactive. Antistreptolysin O titers was <200 mIU/mL. Chest radiograph, ultrasonography of abdomen, electrocardiogram were normal. Mantoux test was negative. S. electrophoresis did not show any abnormality.

Keeping in mind differential diagnosis of keratosis lichenoides chronica, lichen planus, unsual form of bullous pemphigoid and erythema elevatum diutinum biopsy for routine histopathology and direct immunofluorescence were performed from erythematous forearm lesions and perilesional skin respectively. Histopathology of erythematous plaque revealed leukocytoclastic vasculitis; dilated, thickened dermal blood vessels with neutrophilic infiltration within and around the dermal blood vessels, occasional eosinophils, nuclear dust, extravasation of red cells and fibrin deposition around blood vessels (Figure 2 a,b). Direct immunofluoroscence was negative.

Considering the clinico-pathological correlation diagnosis of EED was considered. After normal reports of G-6PD level; oral Dapsone 100mg twice daily and topical betamethasone diproprionate cream were presecribed. Patient started improving symptomatically within one week and complete resolutions of lesions were seen after 6 weeks with post-inflammatory hyperpigmentation. Dapsone was tapered to 100mg daily and continued for 3 months by which time PIH also near completely resolved. (Figure 3a,b). Patient is followed up regularly and had no recurrence of lesions for 2 months. Minor recurrences after two months are managed by topical steroids only.

3. Discussion

EED is a rare, chronic dermatosis that can occur at any age with no gender and ethnic groups predilection though it is commonly seen in adults in the fourth to seventh decade.[2] Clinically, lesions present as firm, tender, brownish-red to purple, papules, plaques, or nodules. Extensor aspects of the extremities, usually near joints such as the fingers, hands, elbows, ankles, and knees are the preferred locations. However, occurrences at atypical sites have been reported, including truncal, retroauricular, palmar, and plantar areas.[1] The Lesions of EED are usually asymptomatic, but pruritus, pain and arthralgia may be present.[2]

Early lesions are characterized histologically by leukocytoclastic vasculitis, with neutrophilic perivascular infiltration in the mid-dermis admixed with eosinophils, lymphocytes, plasma cells, and nuclear dust. Late lesions are characterized by mixed inflammatory cell infiltrate, fibrin deposition, cholesterol deposits in histiocytes, and extracellular tissue (extracellular cholesterolosis).[2]

Many reported cases of EED have been described to be associated with a number of systemic diseases including Infections like streptococcal infection, HIV, hepatitis B and syphilis,[3,4] autoimmune diseases such as inflammatory bowel disease, Wegener's granulomatosis, relapsing polychondritis, lupus erythematosus and rheumatoid arthritis,[5–7] hematological disorders such as plasma cell dyscrasias (multiple myeloma), IgA monoclonal gammopathy, lymphomas, and leukemias.[7–10]

Basic workup and serum electrophoresis in our patient did not reveal any systemic illness. Patient is counseled and is followed up regularly for later development of any systemic disease.

Dapsone is first line management option for EED. Dapsone acts by inhibiting neutrophilic chemotaxis and complement deposition in the vessel wall and suppresses the dysregulation of processes involved in immune complex-induced EED. However, 20% patients do not respond to dapsone or do not tolerate the drug and recurrence after stopping the drug is seen in around 32% cases.[11] The present case responded to dapsone with complete resolution and only minor recurrences after stopping dapsone till date in

follow up over 2 years.

4. Conflict of interest

The authors declare they have no conflict of interest.

5. Source of f unding

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