Case Report

Erythema elevatum diutinum with lower limb neuropathy: A rare presentation

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Abstract

Nodular lesions can be seen in a wide range of conditions such as infections, granulomatous conditions, neutrophilic disorders, vasculitis, benign tumours and malignant conditions. These numerous aetiologies behind the occurrence of nodular lesions signify the importance of thorough work-up of the patient to diagnose the underlying reason behind the clinical condition. Erythema elevatum diutinum (EED) is a rare vasculitis with variable clinical presentation, the diagnosis of which can be challenging. Extracutaneous signs such as arthralgias, oral and penile ulcers, involvement of the eye and neuropathy have all been linked to it. Various systemic illnesses, infectious diseases, autoimmune diseases, haematological abnormalities and plasma cell dyscrasias are also associated with EED. Such extracutaneous signs in EED patients indicate that the condition may involve several organ systems. Patients with EED should be assessed for systemic manifestations to ensure focused care, since extracutaneous forms of EED may comprise deposition of circulating immune complexes.

Keywords: Erythema elevatum diutinum, leukocytoclastic vasculitis, neuropathy, painful nodules

Introduction

Erythema elevatum diutinum (EED) is a rare, chronic dermatosis with less than a thousand cases being reported. EED was first described in the 1880s, but Henry Radcliffe-Crocker first used the term "erythema elevatum diutinum" in 1894.[1] The name EED is due to the characteristics of the cutaneous lesions, which are red (erythema), elevated (elevatum) and persistent (diutinum, in Latin). It can mimic many cutaneous disorders and hence becomes a diagnosis of a challenge for physicians and here we report this case of EED with subsequent development of sensory and motor neuropathy of bilateral lower limbs. The authors through this report hope to bring to the knowledge of the physicians about this rare case and the numerous systemic manifestations that are usually associated with it.

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Case Report

A 54-year-old female patient presented with complaints of multiple painful, dark-coloured raised lesions over bilateral feet, arms and dorsum of fingers for the past 18 months which was associated with the throbbing type of pain. She also complained and tingling and numbness over bilateral lower limbs for the past 6 months. No history of itching, discharge, atopy or drug intake prior to the onset of lesions was noted.

There was no significant abnormality detected on systemic examination. On cutaneous examination, multiple red-brown coloured nodules ranging from size of three millimetres to one centimetre were seen over the extensor aspect of the arms, forearms, dorsal aspect of hands, bilateral feet, over the elbows

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and knees [Figure 1]. A few of the lesions showed erosions and crusting. Nodules were soft in consistency. Nail dystrophy was noted in bilateral toenails. Examination of the scalp, oral cavity, palms and soles did not show any abnormality.

Clinically, differential diagnoses of tuberous xanthoma, multicentric reticulohistiocytosis, sweet's syndrome, Hansen's disease and EED were made in the order of preference respectively.

Routine investigations like complete blood count, peripheral smear, ESR (erythrocyte sedimentation rate), CRP (C reactive protein) and Fasting lipid profile (FLP) were normal. Serology for HIV (Human immunodeficiency virus) was non-reactive. No significant abnormality was detected in the X-ray of the hands and feet and ultrasound abdomen. Histopathological examination showed flattening of the epidermis, effacement of rete ridges and a well-circumscribed dermal infiltrate in scanner view. Subcutaneous tissue was normal when visualised under scanner view [Figure 2]. It showed leukocytoclastic vasculitis in high power view which was characterised by fibrosis of blood vessels with abundant neutrophilic infiltrate, plenty of foamy histiocytes and a few eosinophils [Figure 3]. Staining for acid-fast bacilli was negative. A nerve conduction study done in view of the tingling over bilateral lower limbs showed sensory and motor (axonal and demyelinating) neuropathy of both lower limb nerves. Superior fibular nerves were not stimulatable and S1 radiculoneuropathy was noted.

Lesions in our case were painful and FLP was normal, ruling out tuberous xanthoma. In multicentric reticulohistiocytosis, skin lesions are usually associated with arthritis, associated anaemia and raised ESR is present and histopathology shows lymphocytic infiltrate.

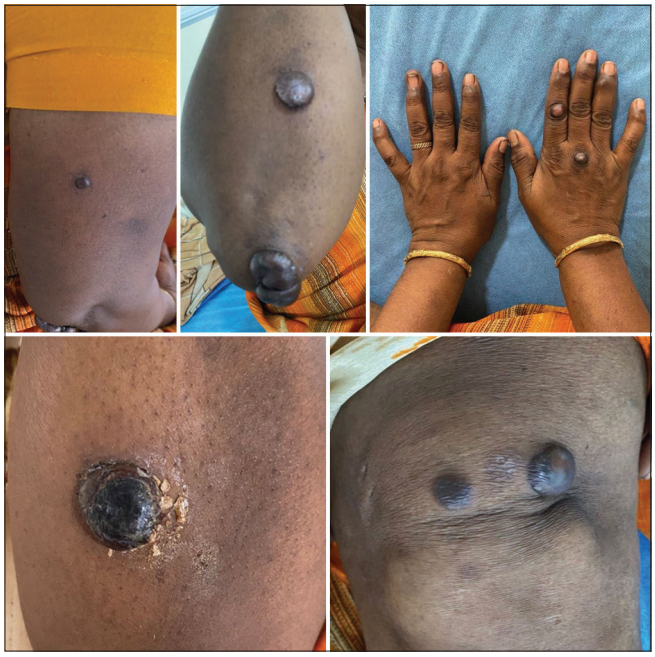

Figure 1: Clinical presentation is characterised by multiple red-brown-coloured nodules seen over the extensor aspect of the arms, forearms, dorsal aspect of hands, bilateral feet, over the elbows and knees

All these features were not seen in our case and hence it was ruled out. The absence of fever, normal levels of total count, ESR, CRP and presence of leukocytoclastic vasculitis which is uncommon in sweet's syndrome helped us to rule it out. Although xanthoma-like presentation is one of the rare manifestations of Hansen's disease, there were no hypopigmented anaesthetic patches over the body, nerve examination was normal and staining for acid-fast bacilli was not seen in histopathological examination and hence it was ruled out.

Finally, with the presentation and histopathological findings diagnosis of EED was made. The patient was started on dapsone 100 mg once a day.

Discussion

EED is an uncommon form of dermal leukocytoclastic vasculitis. It is assumed to be connected to the deposition of a vascular immune complex in small vessels. Adults between the third and sixth decades are typically affected.[2]

Clinically, it is distinguished by symmetrically spaced painful, persistent, red to reddish-brown papules and nodules on the extensor surfaces of the acral regions and joints.[3] The lesions wax and wane over time. It is linked to extracutaneous involvement of the eyes including uveitis and keratitis, as well as the joints, which manifest as arthritis.

EED has been linked to infections such the HIV, hepatitis B or syphilis. Numerous studies associate EED with autoimmune connective tissue illnesses like lupus erythematosus, dermatomyositis, chronic inflammatory bowel disease and recurrent polychondritis as well as haematological disorders like clonal gammopathy.[4] A few reports by *G Fiorillo et al.,* and *Bushra Muna et al.,* have suggested the presentation of EED post-covid vaccination.[5,6] However, further studies have to be done to justify its correlation, especially in this era post the covid pandemic. Despite the fact that EED is linked to a number of disorders, our patient's diffuse neuropathy was unusual. Nerve atrophy can be a result of direct injury due to autoimmune infiltrative neurologic illness involving tiny fibres or indirect damage in relation to chronic or intermittent swelling leading to nerve compression or entrapment.[7]

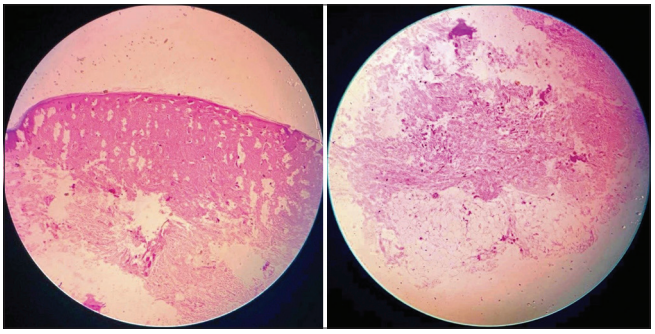

Figure 2: Scanner view of the histological picture demonstrating the epidermis, dermis and subcutaneous tissue. Flattening of the epidermis, effacement of rete ridges, a well-circumscribed dermal infiltrate and normal subcutaneous tissue are seen

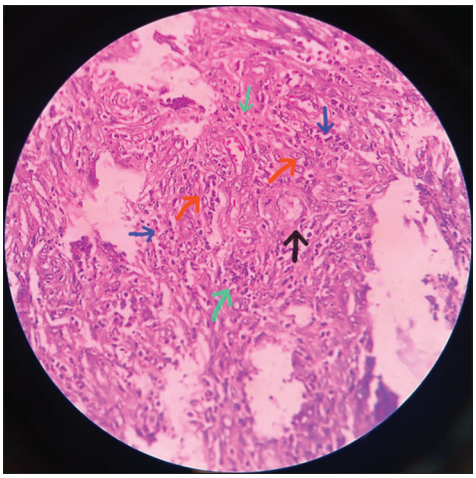

Figure 3: High power of the histological picture showing leukocytoclastic vasculitis characterised by fibrosis of blood vessels (black arrows) with abundant neutrophilic infiltrate (green arrows), plenty of foamy histiocytes (orange arrows) and a few eosinophils (blue arrows)

Early lesions of EED show leukocytoclastic vasculitis on histological analysis, along with neutrophilic infiltration and a few eosinophils. Extracellular cholesterolosis and fibrosis may be present in the late-stage of the disease.

Sweet's syndrome, dermatitis herpetiformis, rheumatoid neutrophilic dermatitis rheumatoid nodules, multicentric reticulohistiocytosis, tuberous xanthoma, and granuloma annulare should all be considered in the differential diagnosis.[8]

Oral dapsone is the most popular form of treatment and work by preventing neutrophil chemotaxis. Systemic corticosteroids, colchicine, methotrexate and chloroquine are second-line treatments. Tetracyclines and nicotinamide have also been used. Newer therapies include topical 5% dapsone preparations and plasmapheresis.[9]

Conclusion

EED represents a rare cause of chronic leukocytoclastic vasculitis. It can present with several extracutaneous findings and systemic diseases like neuropathy making it important to work-up for the underlying disease to optimise management. Finally, the main objective of this report is to alert physicians to consider the possibility of EED in such a clinical presentation because it is frequently diagnosed after ruling out umpteen number of conditions and to the best of our knowledge, there has only been a singular case report of EED associated with neuropathy.

Key points

- EED has an umpteen number of differentials and hence physicians should consider it as a possibility in such clinical presentations
- Due to extensive systemic associations physicians should make sure to do a detailed work-up in EED cases to avoid further complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. Sandhu JK, Albrecht J, Agnihotri G, Tsoukas MM. Erythema elevatum et diutinum as a systemic disease. Clin Dermatol 2019;37:679-83.
2. Galeone M, Arunachalam M, Bassi A, Scarfì F, Difonzo EM. Erythema elevatum diutinum. QJM 2014;107:227-8.
3. Keyal U, Bhatta AK, Liu Y. EED involving palms and soles: A case report and literature review. Am J Transl Res 2017;9:1956-9.
4. Wollina U, Krönert C, Koch A, Schönlebe J, Vojvodic A, Lotti T. Erythema elevatum diutinum-Two case reports, two different clinical presentations, and a short literature review. Open Access Maced J Med Sci 2019;7:3039-42.
5. Fiorillo G, Pancetti S, Cortese A, Toso F, Manara S, Costanzo A, *et al.* Leukocytoclastic vasculitis (cutaneous small-vessel vasculitis) after COVID-19 vaccination. J Autoimmun 2022;127:102783.
6. Muna B. A case of erythema elevatum diutinum after COVID-19 vaccination. J Eur Acad Dermatol Venereol 2023.
7. Nguyen GH, Guo EL, Norris D. A rare case of erythema elevatum diutinum presenting as diffuse neuropathy. JAAD Case Rep 2017;3:1-3.
8. Mançano VS, Dinato SLME, Almeida JRP, Romiti N. Erythema elevatum diutinum. An Bras Dermatol 2018;93:614-5.
9. García-Meléndez ME, Martínez-Cabriales SA, Eichelmann K, Gómez-Flores M, Ocampo-Candiani J. Erythema elevatum diutinum: An atypical presentation. Am J Med Sci 2015;349:374-5.