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Asymptomatic Papulonodules on Elbows and Hands: A Rare Chronic Fibrosing Leukocytoclastic Vasculitis

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Abstract: ***One of the pathogenic causes of cutaneous inflammatory pseudotumors is chronic localized fibrosingleukocytoclastic vasculitis (CLFLCV), seen in Granuloma Faciale (GF) and Erythema ElevatumDiutinum (EED). EED is a rare type of leukocytoclastic vasculitis, most commonly effecting young and middle age groups. EED is thought to be an Arthus type reaction with immune complex deposition secondary to a number of infections, autoimmune diseases, both benign and malignant haematological disorders. This is a rare, chronic, cutaneous eruption characterized by red, purple or yellowish papules, plaques and nodules distributed acrally and symmetrically over extensor surfaces. Here, we report a case of thirty-five year old male with EED.***

Keywords: Erythema elevatumdiutinum, Leukocytoclasticvasculitis

1. Introduction

One of the pathogenic causes of cutaneous inflammatory pseudotumours is chronic localized fibrosingleukocytoclastic vasculitis (CLFLCV), a vasculitic reaction pattern seen in Granuloma Faciale (GF), a localised vasculitis and Erythema Elevatum Diutinum (EED), a generalised vasculitis.[1]Erythema elevatumdiutinum is a rare, chronic, skin eruption characterized by red, purple or yellowish papules, plaques and nodules distributed acrally and symmetrically over extensor surfaces.[2]Here, we report a case of thirty five year old male with EED.

2. Case Report

A 35year old male presented with multiple, hyperpigmented, papulonodules and plaques over extensor surface of both elbows and hands since one and half years. The lesions first started as hyperpigmented papules initially over the right elbow which gradually increased in size, number and also spread to involve other sites in a span of 6 months. Lesions were initially soft in consistency, but later progressed to become hard. The lesions were asymptomatic but there was occasional sensation of numbness and pain over fingers in winter season and on exposure to cold.

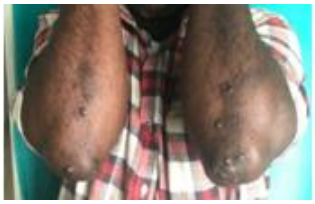

Figure 1

Figure 2

Figure 3

Figure 4

On cutaneous examination, multiple, firm, non tender, hyperpigmentedpapulonodules and plaques, of varying sizes, smallest being 1x1cms and largest being 2X3cms noted over the extensor aspects of both elbows (Figure: 1), dorsum of hands and fingers (Figure:2 and Figure:3) (MCP and IP) and palms (Figure: 4) in a symmetrical fashion. The lesions were immobile and there is no change in size or shape of the lesion on application of pressure. Few lesions were smooth and few lesions were hyperkeratotic. A differential diagnosis of Multicentric Reticulate Histiocytosis, Erythema ElevatumDiutinum and Rheumatoid Nodules were considered.

Routine investigations, HIV, C-Reactive protein, Antinuclear antibodies, RA factor, Prostate Specific Antigen, Mantoux test, Slit Skin Smear, ultrasound abdomen and x-ray chest, both elbows, hands and knees revealed no significant abnormality except for positive ASO titres and reactivity for HbsAg. Histopathology of hyperpigmented papule showed features of fibrosing neutrophilic leukocytoclastic vasculitis (Figures: 5, 6, 7).

Figure 5

Figure 6

Figure 7

A final diagnosis of EED was made and patient was put on dapsone 100mg/day with complete resolution of the lesions in 3 months (Figures: 8, 9, 10, and 11)

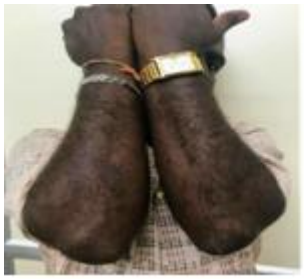

Figure 8

Figure 9

Figure 10

Figure 11

3. Discussion

EED is a rare type of cutaneous vasculitis, most commonly effecting young and middle age groups.EED was first described in 1888 by Hutchinson and 1889 by Bury. This was later named in 1894 by Radcliffe Croker and Williams.[3]The latin name describes the characteristic lesions seen in EED; red (erythema), elevated (elevatum), persistent (diutinum). It is mediated by immune complexes (Arthus type reaction) and has been associated with auto immune, neoplastic and various infections like Streptococcus, hepatitis, syphilis, tuberculosis, etc.[3][4]In addition, EED has been associated with hypergammaglobulinemia, IgA monoclonal gammopathies, as well as with myelodysplasia, pyoderma gangrenosum, and relapsing polychondritis.[4**]** Erythema elevatumdiutinum is very rare with only a few hundred cases described.[3]

Characteristic skin lesions are asymptomptomatic plum coloured erythematous nodules and plaques of variable sizes distributed symmetrically over extensors of extremities particularly over the joints, dorsal hands and feet, knees and elbows, buttocks and achilles tendon and occasionally face and ear pinnae. The initial soft lesions eventuate to become fibrotic and develop atrophic scarring.[5]Characteristic histologic features of leukocytoclastic vasculitis of the mid- and upper-dermal vessels with fibrinoid necrosis, a dense dermal neutrophilic infiltrate, and, in older lesions, fibrosis.[6]

Dapsone has remarkable therauptic effect and remains the mainstay of the treatment. However, resurgence is common on its discontinuation. Sulfpyridine, colchicine or cyclophosphamide are other alternative treatments. [5]

4. Conclusion

This case is reported for its rarity and to underscore its importance in the differential diagnosis of asymptomatic papulonodules on elbows, knees and extremities and its possible underlying systemic associations.

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