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Case Report

Elastosis Perforans Serpiginosa - A Rare Side Effect of D-Penicillamine Therapy

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Abstract

Penicillamine can cause various cutaneous adverse effects. Elastosis Perforans Serpiginosa (EPS) is a rare complication of long-term penicillamine therapy. We report a case of 21 year old male patient, diagnosed as a case of Wilson's disease 10 years back, and taking D-penicillamine for last 10 years. Now, presented with non-itchy skin lesions over the neck region and bilateral axillae. Skin biopsy showed the presence of abnormal elastic fibers surrounded by acanthotic epidermis compatible with the diagnosis of elastosis perforans serpiginosa (EPS). EPS is a rare complication of long-term penicillamine therapy causing transepidermal elimination of abnormal elastic fibers.

Keywords: Wilson's Disease; Elastosis Perforans Serpiginosa; D-Penicillamine

Introduction

Penicillamine can cause various cutaneous adverse effects like acute hypersensitivity reactions, elastosis perforans serpiginosa, pseudo-pseudoxanthoma elasticum, drug induced lupus erythematosus [1,2]. EPS is a rare complication of long-term penicillamine therapy causing transepidermal elimination of abnormal elastic fibers [3].

Case Report

We report a case of 21 year old male patient, diagnosed as a case of Wilson's disease 10 years back, and taking D-penicillamine (250 mg thrice daily) for last 10 years. Patient was diagnosed as a case of Wilson's disease following clinical symptoms in the form of tremors in bilateral hands, and slowness of activities. Patient had rest tremors, bradykinesia and rigidity on examination. Following investigations including elevated 24 hour urinary copper and reduced serum ceruloplasmin levels, the diagnosis of Wilson's disease was confirmed. He was put on D-penicillamine as a chelation therapy 10 years ago. Now, presented with non-itchy skin lesions over the neck region and bilateral axillae. The lesions had been gradually increasing in size with central clearing. There was no history of any other systemic illness or medication changes. On cutaneous examination, there were multiple reddish brown papules coalescing to form serpiginous plaques (Figure 1-3). They were surrounded by a hyperkeratotic rim with central clearing. They varied in size from a few millimetres to a centimetre. Neurologically, there was no sensorimotor deficit or extrapyramidal sign. Kayser-Fleischer rings were absent on slit lamp examination. Skin biopsy showed the presence of abnormal elastic fibers surrounded by acanthotic epidermis compatible with the diagnosis of elastosis perforans serpiginosa (EPS). The diagnosis of penicillamine induced EPS was made and the drug was withdrawn and replaced by zinc acetate. The skin changes did not progress over next one year of follow up.

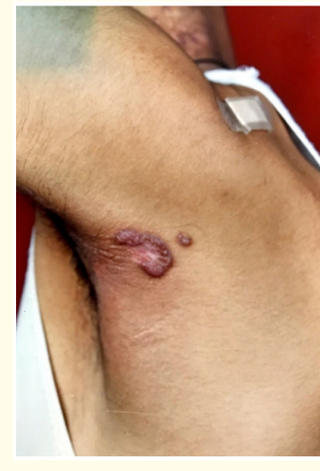

Figure 1: Serpiginous plaque showing central clearing and hyperkeratotic rim in left axilla.

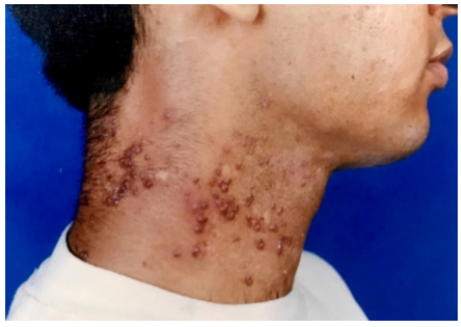

Figure 2: Multiple reddish brown papules over cervical region.

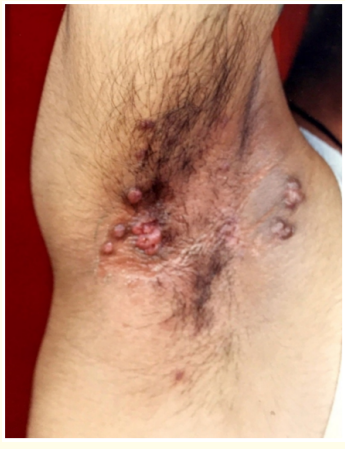

Figure 3: Multiple reddish brown papules with few serpiginous plaques showing central clearing and hyperkeratotic rim in right axilla.

Discussion

EPS is seen more commonly between 6 and 20 years of age. The common sites affected are face, neck and upper limbs [3]. The underlying pathogenesis is not yet clear. Various hypothesis have been proposed including a local copper deficiency in the skin causing impaired cross-linking of collagen fibers, causing abnormal elastic fiber deposition [4]. D-penicillamine does not affect the mature collagen, thus explaining the long period before dermopathy sets in. It has been estimated that ingestion of minimum of 1 g per day of D-penicillamine for more than 5 years is required to induce these changes [5]. The presence of underlying connective tissue disorder such as Ehlers-Danlos syndrome and osteogenesis imperfecta has also been seen in some patients with EPS [3].

Therapeutic management of EPS mainly involves removing the offending agent besides other measures like oral isotretinoin and intra-lesional corticosteroids. Spontaneous resolution of the lesions has been reported within three years of stopping penicillamine therapy [4]. Management of WD warrants replacing penicillamine with other effective and less toxic drugs like trientine and zinc acetate. The cost of therapy is an important limiting factor with trientine in low and middle income countries.

Conclusion

In view of many cutaneous and non cutaneous major adverse effects of penicillamine, other safer drugs should be considered for WD treatment as per clinical and socio-demographic setting.

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