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Case Report

Elastophagocytosis and Elastolysis in Leprosy

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Elastophagocytosis is the engulfment of the elastic fibres by the histiocytes, multinucleated giant cells, or both. The cutaneous lesions showing elastophagocytosis are annular elastolytic giant cell granuloma, actinic keratoses, persistent insect-bite reactions, elastosis perforans serpiginosa, foreign body granuloma. Occasionally, it may occur in infectious diseases like leprosy, granulomatous syphilis, North-American blastomycosis, bacterial folliculitis, and cutaneous leishmaniasis. We report a case of lepromatous leprosy with necrotic erythema nodosum leprosum with secondary anetoderma. Histopathology from the atrophic macule of anetoderma revealed periappendageal, perineural infiltration, elastophagocytosis and reduction in elastic fibres.

Key words : Leprosy, anetoderma, elastophagocytosis, elastolysis.

Introduction

The clinical and histological diagnosis of leprosy and its classification may be at times difficult with frequent overlap of findings. Further the co-existence of the findings of leprosy and giant cell granuloma (LEGG), could further pose a diagnostic dilemma for clinicians and pathologists. Annular elastolytic giant-cell granuloma (AEGCG), characterized histologically by loss of elastic fibres and elastophagocytosis may have histological picture overlapping with actinic granuloma, sarcoidosis, leprosy and granuloma annulare and this poses diagnostic challenge at times (Le Corre et al 2010). We present this case report to highlight that lepromatous leprosy may coexist with elastophagocytosis and elastolysis.

Case Report

A 60 year old male presented with multiple dark coloured painful raised lesions over both upper and lower extremities on and off since 7 years. There was history of fever associated with the lesions. The patient had a past history of Hansen's disease 7 years back for which he had taken complete multi drug treatment (MDT-MB) for a total duration of 2 years.

On examination, he had multiple erythematous and hyperpigmented nodules with pus discharge over both the upper and lower extremities.

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There were shiny hyperpigmented plaques with atrophic skin over the waist and hyperpigmented icthyotic plaques over both the lower extremities. In addition, multiple hypopigmented oval macules, patches and slightly bulging plaques of varying sizes with smooth atrophied shiny surface over the trunk and shoulder (Fig 1). Stretching the surrounding skin made the plaques unapparent. Sensory examination revealed absent touch and temperature sensations over bilateral lower extremity. Motor examination was within normal limits. Nerve examination showed bilateral thickening of the ulnar and common peroneal nerves.

Skin biopsy from the nodule over the thigh region showed superficial and deep perivascular, periappendageal and perineural infiltration with foamy macrophages, few neutrophils and plasma cells. A skin biopsy from the oval atrophic macule revealed similar periappandageal and perineural infiltration. Additionally, the papillary dermis showed giant cells with engulfed elastic fibres (elastophagocytosis) (Fig 2). Verhoeff van gieson (VVG) stain showed reduction in the elastic fibers in the dermis which was suggestive of anetoderma.

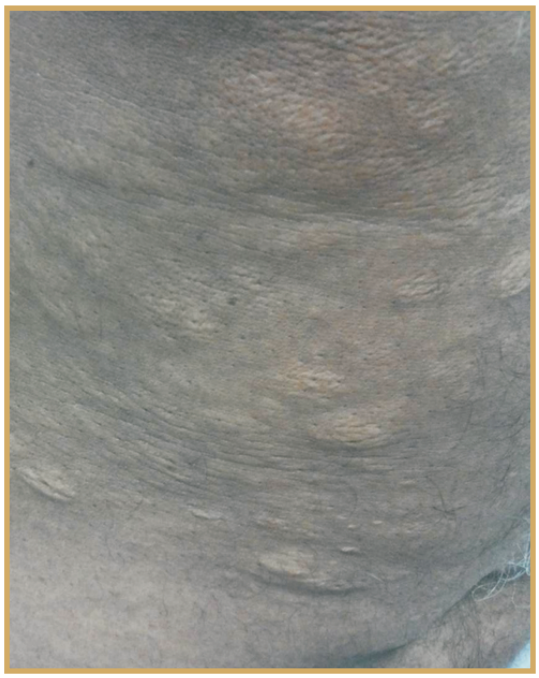

Fig 1 : Oval, hypopigmented macules with smooth atrophied skin

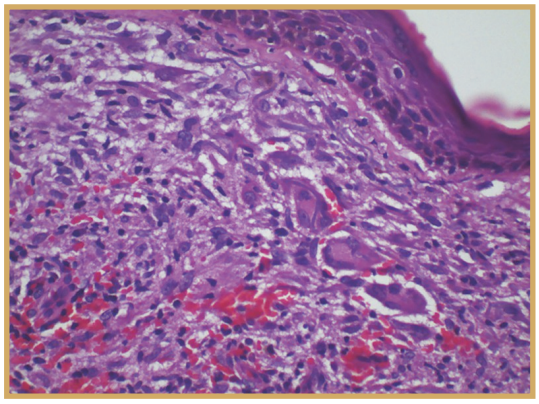

Fig 2 : Elastophagocytosis (Hematoxylin and Eosin x 40)

The patient was diagnosed as lepromatous leprosy with necrotic erythema nodosum leprosum with secondary anetoderma and started on WHO three drug therapy for multi-bacillary leprosy and tablet prednisolone 60 mg daily. The existing lesions subsided and there were no new lesions after initiating treatment.

Discussion

Anetodermas are also known as macular atrophies. They are characterised by oval or circular atrophic lesions and are clinically appreciated as flat or papular areas of thin, hypopigmented and herniated skin which may be protruding. They are located on the trunk, nape of neck or face. They are a result of the destruction of the elastic fibres of the dermis. They may be classified into three groups: hereditary, primary and secondary.

Secondary anetoderma occurs as a result of destruction of elastic fibres secondary to pre-existing skin pathology. It is known to follow resolution of lesions in lupus erythematosus, syphilis, Hansen's disease, tuberculosis, varicella and borreliosis. The lesions may not necessarily appear at the same place as the cutaneous manifestation of the associated disease (Barbosa et al 2002).

Elastic fibre degradation is called as elastolysis. Elastophagocytosis is the engulfment of the elastic fibres by the histiocytes, multinucleated giant cells, or both (El-Khoury et al 2014). The cutaneous lesions showing elastophagocytosis are annular elastolytic giant cell granuloma (Pock et al 2004), actinic keratoses, persistent insect-bite reactions, elastosis perforans serpiginosa, foreign body granuloma, necrobiosis lipoidica, senile purpura, keratoacanthoma, basal cell carcinoma and certain variants of cutaneous T-cell dyscrasia i.e. granulomatous slack skin and mycosis fungoides and adult T cell leukemia. Elastophagocytosis may also be seen in infections including leprosy, granulomatous syphilis, North-American blastomycosis, bacterial folliculitis, and cutaneous leishmaniasis (El-Khoury et al 2014).

Elastolytic giant cell granuloma (EGCG) is a descriptive terminology which includes granulomatous infiltration by many giant cells, histiocytes, lymphocytes, scattered epithelioid cells, and no necrobiosis. Elastic tissue is reduced at the borders of the granuloma and is totally absent in the centre (El-Khoury et al 2014). The co-existence of the findings of leprosy and giant cell granuloma (LEGG), could pose a diagnostic dilemma for clinicians and pathologists. There may be misinterpretation of the pattern of granulomatous elastolysis or granulomatous elastophagocytosis as other granulomatous diseases like annular elastolytic giant cell granuloma or granuloma annulare. (Le Corre et al 2010).

From this case report, it becomes necessary to remember that the findings of elastolysis, elastophagocytosis and elastorrhexis may sometimes be seen in the lepromatous pole of leprosy.

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