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Dermatology

A RARE CASE REPORT OF ELASTOSIS PERFORANS SERPIGINOSA WITH HYPOTHYROIDISM

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ABSTRACT Elastosis perforans serpiginosa is a rare perforating dermatosis characterized by transepidermal elimination of abnormal elastic fibers that classically present as small keratotic papules arranged in serpiginous or annular pattern over face, neck, arms or other flexural areas. It is also known as Perforating elastoma and Elastoma intrapapillare perforans. Hereby we report a rare case of elastosis perforans serpiginosa with hypothyroidism.

KEYWORDS : Elastosis Perforans Serpiginosa, Elastic Fibers, Perforating Dermatosis, Keratotic Papules.

INTRODUCTION:

Elastosis perforans serpiginosa is a rare perforating dermatosis characterized by transepidermal elimination of abnormal elastic fibers that classically present as small keratotic papules arranged in serpiginous or annular pattern over face, neck, arms or other flexural areas and are sometimes bilaterally symmetrical [1]. It is also known as Perforating elastoma and Elastoma intrapapillare perforans. The papules are asymptomatic skin-colored to slightly erythematous and may have central scaling, atrophy or hypopigmentation. They can be arranged symmetrically with satellite lesions. The disease occurs predominantly in males and usually in ages between 5 and 20 years.

Here we present a case of elastosis perforans serpiginosa with hypothyroidism in a 72-year-old male.

Case Report:

A 72 year old male presented with complaints of severe itching all over the body for 2 months. On examination multiple discrete keratotic papules of 2-5mm (about 0.2 in) in size arranged in annular pattern were present over face, neck, trunk, upper and lower limbs. There was no family history of similar lesions and no history of drug intake. General and systemic examination are normal; mucus membranes, nails, scalp, palms and soles are normal.

Routine investigations show normal hemoglobin levels of 15g/dl, differential leucocyte count- increased neutrophils, decreased lymphocytes, increased eosinophils, PCV- increased, MCH and MCHC-decreased, ESR-increased, Renal function test-increased uric acid levels, Liver function tests- normal.

Thyroid profile: Suggestive of hypothyroidism: T3: 52.94ng/dl T4: 4.52microg/dl TSH: >100UIU/ml 5mm punch biopsy was taken from the lateral abdominal wall for histopathological examination. Histopathology shows hyperplastic epidermis with transepidermal elimination of dermal elastic fibers. There is a mixture of degenerated, eosinophilic elastic fibers, basophilic debris and mixed inflammatory cells with lymphocytes and macrophages in the dermis. The epidermis surrounding the lesion is acanthotic and hyperkeratotic.

Figures:

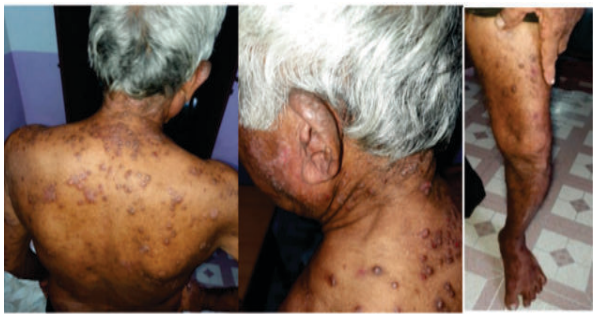

Figure-1 (a,b&c): Multiple hyperkeratotic papules over back, neck and leg.

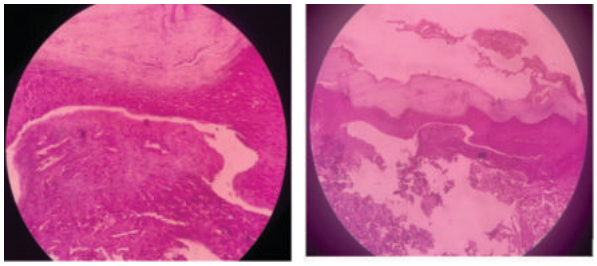

Figure-2 (a & b): Histopathology showing hyperplastic epidermis with transepidermal elimination of dermal elastic fibers.

DISCUSSION:

Elastosis perforans serpiginosa is a rare skin disease in which there is trans epidermal elimination of abnormal elastic fibres. It is more commonly reported in men and usually occurs in childhood and early adulthood. There are three subtypes of EPS.

1. Idiopathic EPS: about 60-70% of cases fall in this category where the exact cause is unknown and is more likely due to autosomal dominant type of inheritance.
2. Reactive EPS: this type is associated with connective tissue diseases, such as Ehlers-Danlos syndrome (type IV), osteogenesis imperfecta[2], Marfan syndrome, Rothmund-Thomson syndrome, acrogeria, cutis laxa and pseudoxanthoma elasticum. It has also been reported in association with Down's syndrome.
3. Drug-induced EPS: this variant is caused by the drug D-penicillamine [3]. It occurs in nearly 1% of patients on long term therapy for Wilson's disease.

Tends to be self limiting and no treatment is of proven benfit. Careful removal of the nodules with a curette under local anesthesia may give cosmetic result.Various modalities have been successfully used such as topical corticisteroids, topical and oral retinoids, imiquimod[4], calcipotriol, electrocautery and cryotherapy with liquid nitrogen. Treatment with pulsed dye, ultrapulsed carbondioxide and Er:YAG lasers [5] have also been advocated.

Prognosis: EPS can persist for long periods but may involute spontaneously to leave reticulate atrophic scars.

Differential Diagnosis:
conditions with annular patterns should be considered such as tinea corporis, granuloma annulare, porokeratosis, sarcoidosis and discoid lupus erythematosus. Other conditions include familial reactive perforating collagenosis, skin calcinosis, cutaneous larva migrans etc.,

CONCLUSION:

We report this case due to its rare occurrence, rare onset in elderly , classical presentation and association with hypothyroidism.

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