

Dermato-venereological Quiz

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A 28-year-old gentleman presented with multiple itchy, mildly tender papules and annular plaques on the posterolateral aspect of his neck for 3-4 years. The lesions started as small erythematous papules that gradually increased in size and coalesced into irregular serpiginous or annular plaques (Figure 1). Some of them were excoriated. He was treated with

topical steroid in various clinics of the public sector without any effect. Concerning his past health, he had Wilson's disease and was on regular treatment with penicillamine for a few years. Investigations including skin scraping for fungal culture, ANF and complete blood picture showed no abnormalities. A skin biopsy was also performed (Figures 2 & 3).



Figure 1. Itchy lesions over the neck for 4 years.

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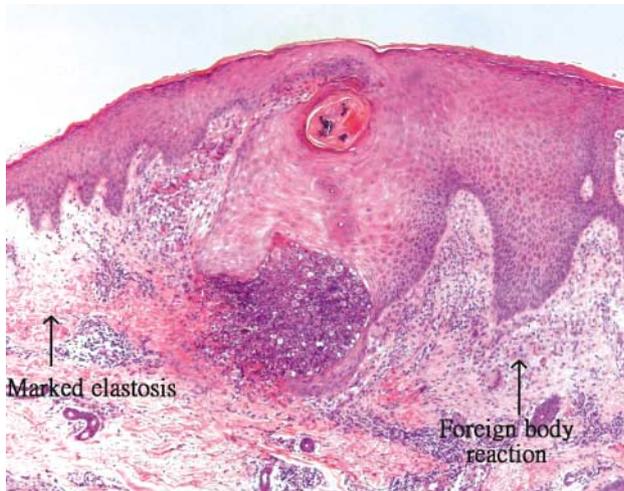


Figure 2. A collection of basophilic debris is protruding into the hair follicle. Marked elastosis with foreign body reaction is present in the upper dermis. (H&E, original magnification x 5)

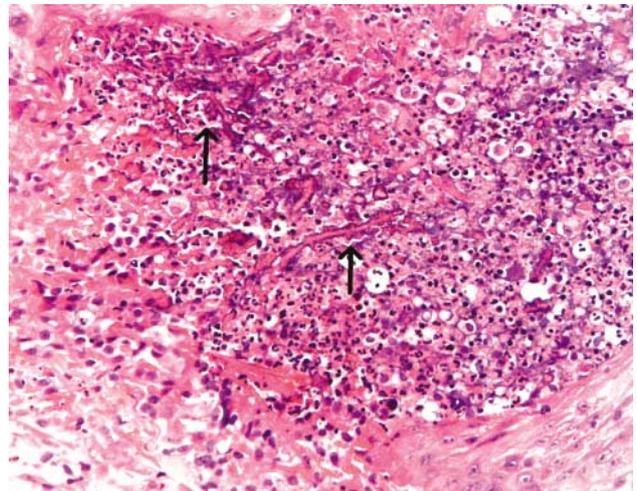


Figure 3. Elastic fibers (arrows) are found in the debris. (H&E, original magnification x 10)

Questions

1. What are the diagnosis and differential diagnoses for this patient?
2. What are the histopathological findings of this lesion?
3. What is the commonest site of involvement of this disease?
4. What are the treatments of choice?
5. What is the prognosis of this disease?

Answers to Dermato-venereological Quiz on pages 206-207

1. The diagnosis is elastosis perforans serpiginosa (EPS). Forty percent of cases occur in association with Down's syndrome, Ehlers-Danlos syndrome, Marfan's syndrome, osteogenesis imperfecta, and acrogeria. It can be a complication of penicillamine treatment. The differential diagnoses include perforating granuloma annulare, discoid lupus erythematosus, tinea corporis, porokeratosis, prurigo nodularis and actinic granuloma.
2. The skin biopsy in this case revealed a hyperplastic epidermis and an obliquely cut hair follicle featuring keratin plugging. In the upper dermis, there was markedly increased outline irregularity, fragmented and brightly eosinophilic elastic fibres. Focal collection of basophilic debris, polymorphs and elastic fibres were found protruding into the hair follicular epithelium. In the papillary dermis, there were aggregates of histiocytes engulfing the elastic fibres. PASD and Grocott's stains showed no fungus. The immunofluorescence study was negative.
3. The commonest site of involvement of EPS is the posterolateral aspect of the neck as seen in this patient. Other sites include the chin, cheeks, mandibular areas of the face, antecubital fossae, elbows, and knees.
4. The treatments of choice are cryotherapy, curettage, laser, keratolytic agents, and as in our case, stopping penicillamine.
5. The lesions may spontaneously resolve, but they tend to persist for up to several years.