Read Item - Graham-Little Syndrome

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Abstract: Doctor's resource on Graham-Little Syndrome

Graham-Little Syndrome

Definition
This is a distinctive condition that resembles lichen planopilaris. It has been suggested that Graham-Little syndrome is a variant of lichen planus, however the absence of typical lichen planus elsewhere, the bland histology and the lack of response to therapy suggest a valid distinction between these two conditions.

Epidemiology
The condition is rare. Most patients are women between the age of 30 and 70.

Pathogenesis
Unknown

Clinical features
The essential features of the condition are progressive cicatricial alopecia of the scalp occasionally with prominent follicular plugging, loss of pubic and axillary hair without scarring and the rapid development of keratosis pilaris. The keratosis pilaris is dramatic with horny papules prolonged into conspicuous spines resembling lichen spinulosus. It occurs in plaques, often on the trunk and limbs, occasionally involving the eyebrows and cheeks.

Pathology
Histology shows a dilated follicular orifice filled with a keratin plug. The follicles beneath the plugs are progressively destroyed and eventually an atrophic epidermis covers a sclerotic dermis. The biopsy is remarkable for the lack of inflammation.

Investigation
Scalp Biopsy.

Differential Diagnosis
The condition needs to be distinguished from lichen planopilaris.

Associated Features
Nil.

Prognosis
Poor. The condition tends to be progressive.

Treatment
Many agents have been tried, but none is effective.

Key Points
The triad of cicatricial alopecia with or without follicular plugs, loss of pubic and axillary hair, and widespread plaques of keratosis pilaris produces this characteristic syndrome.