Case report

Recurrent angiokeratoma scroti: a case report

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Abstract
Introduction
Angiokeratoma is a broad term that describes various conditions of asymptomatic hyperkeratotic vascular disorders with a histologic combination of hyperkeratosis and superficial dermal vascular ectasia. Here we are describing a case of recurrent angiokeratoma scroti.

Case report
A 34-year-old patient presented to the surgery department with multiple small papules over the scrotum with a previous history of two recurrences. They were slowly progressive, itchy and not associated with any bleeding. Examination revealed multiple soft compressible papules of 3–4 mm diameter with excoriation marks. Systemic examination and routine investigations were normal. Histologically, they were composed of numerous dilated thin walled congested capillaries mainly in the papillary dermis. Epidermis revealed variable degree of acanthosis and elongation of rete ridges and mild hyperkeratosis. Electrocauterisation of lesions was done and the follow-up period of three months was uneventful.

Discussion
Patients of angiokeratoma usually present with small, bright red, vascular papules rarely larger than 3–4 mm. Angiokeratoma of scrotum are true ectasia of blood vessels. Initially, the lesions may be red, soft and compressible but later on may be firm, keratotic and sometimes warty.

These vascular lesions may be single and discrete or arranged in clusters. The most frequent symptom is a sensation of heaviness, tension in the scrotum accompanied by irritation along with bleeding of varying degree and superadded infection. Progressive worsening of symptoms, extension of the involved area, repeated episodes of cellulitis and infection cause psychological and emotional distress in the patient; sometimes requiring hospitalisation.

All authors abide by the Association for Medical Ethics (AME) ethical rules of disclosure.

Patients may even seek medical attention to rule out a sexually transmitted disease or malignancy.

There are four types of localised angiokeratoma which include: (1) Angiokeratoma circumscriptum, (2) Angiokeratoma of Mibelli (in childhood or adolescence), (3) Solitary or multiple angiokeratomas (in young adults, most commonly on the lower extremities) and (4) Angiokeratoma of Fordyce (most common) consists of multiple vascular papules 2–4 mm in diameter which are present on the scrotum. Similar lesions have been described on the vulva.

Angiokeratoma corporis diffusum (Fabry disease) is a widespread form of X-linked genetic disorder associated with deficiency of galactosidase.

The pathophysiology of angiokeratoma remains uncertain. However, some reports and case reviews suggest that increased venous pressure proximal to the site of lesion is responsible. In men, varicoceles have been implicated as a common cause, although the data are variable. In women, increased venous pressure is noted during pregnancy and in vulval varicosity, post-partum and post-hysterectomy. Although it is often a benign condition, it causes considerable worry and distress to the patient. A representative lesion must be biopsied if there is a diagnostic doubt for confirmation.

Histological features are hyperkeratosis of the epidermis, rete ridge elongation and marked ectasia of the upper dermal vessels with dilated post-capillary venules within the dermal papillae connected by very short channels. Gross appearance and clinical presentation of these benign lesions can vary widely among patients, and proper diagnosis may often be difficult. The long list of differentials includes both benign and malignant conditions, thus emphasising the importance of representative excisional biopsy and histological examination.

Treatment can range from conservative management such as reassurance with close monitoring and follow-up to more active approaches, especially for bleeding or discomfort. It also depends on the site and size of the lesion and availability of surgical equipment such as electrocautery, radiofrequency, cryotherapy or ablative lasers. The major reasons for morbidity of this disease are bleeding, anxiety and overtreatment due to misdiagnosis by the treating physicians. Therefore, definitive and accurate diagnosis is imperative for ensuring appropriate management.

The patient must be reassured regarding the benign nature of the disease. All of the treatment modalities are unsatisfactory either because of scarring or recurrence. However, good response has been reported with treatments using the CO2 laser, pulsed-dye laser and Nd:YAG laser resulting in minimal scarring, transient purpura, crust over the lesion and minimal procedural bleeding. Sclerotherapy with repeated local injections of 0.5% ethanolamine oleate or 0.25% sodium tetradecyl sulphate has demonstrated mild pain and epithelial sloughing with minimal scarring.

**Conclusion**

Although angiokeratoma scroti is not an uncommon entity, a better treatment modality with minimal adverse effects in terms of complete resolution of diffuse lesions, recurrence, scarring, crusting, bleeding or cosmetic results is still not available.

![Figure 1](image-url)
Consent
Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References