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# ANGIOKERATOMA CIRCUMSCRIPTUM

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Three cases, a 20-year-old male patient with gradually spreading verrucous and crusted nodular lesions on the left lower limb of two years duration, a 60-year-old female with linear verrucous lesions on the left lower extremity for many years and a 36-year-old man with localized, asymptomatic, papulo-nodular, verrucous lesions on the left leg for many years, showed characteristic features of angiokeratoma circumscriptum, on histopathology.

**Key words : Angiokeratoma circumscriptum.**

Angiokeratoma circumscriptum (AC) is a very rare disorder.[1,2] It is also called angiokeratoma circumscriptum neviforme.[1] In its early stage, it consists of one or occasionally several aggregates of purplish papules and blood-filled cystic nodules. Young vascular lesions are non-keratotic and compressible.[3] The lesions gradually become verrucous and coalesce into one or several plaques. Sometimes these show a linear arrangement.[4] The exact aetiology is not known. Imperial and Helwig[5] are of the view that AC represents telangiectasia of papillary vessels acquired as a result of trauma. There are only sporadic reports of AC in the contemporary Indian[2,6-8] and international literature.[9] We are reporting 3 cases of AC.

## Case Reports

### Case 1

A 20-year male patient reported for lesions on the left leg 2 years ago which started as nodules and were removed surgically. Soon after the surgery, the lesions started reappearing and gradually increased in size. A few isolated nodular lesions were present on the same limb, one in the popliteal fossa, another on the thigh and also on an atrophic scar of the past surgery. The lesions were verrucous and crusted papules and nodules (Fig. 1). These were friable and the keratotic material could be removed with little difficulty. Histopathology showed hyperkeratosis, papillomatosis and irregular acanthosis. Greatly dilated capillary spaces filled with RBCs were seen in the papillae. A few dilated spaces showed thrombi.

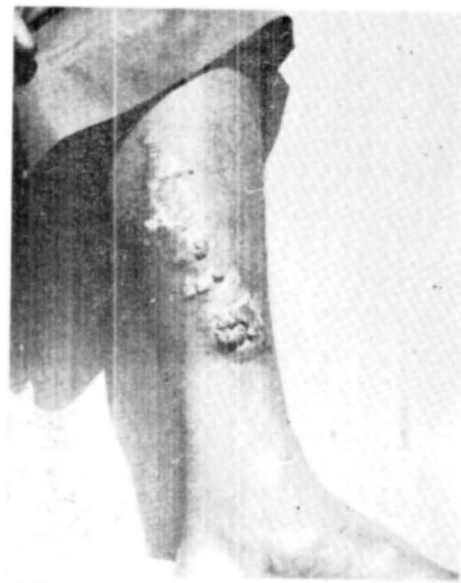

**Fig. 1.** Papules, nodules and plaques with a verrucous surface situated on an atrophic scar on the left lower limb.

### Case 2

A 60-year-old woman came for lesions on the left lower extremity, present for many years, which were spreading gradually. The lesions extended from mid-thigh upto the mid-calf in a linear fashion. A few lesions were discrete while others had coalesced to form plaques.

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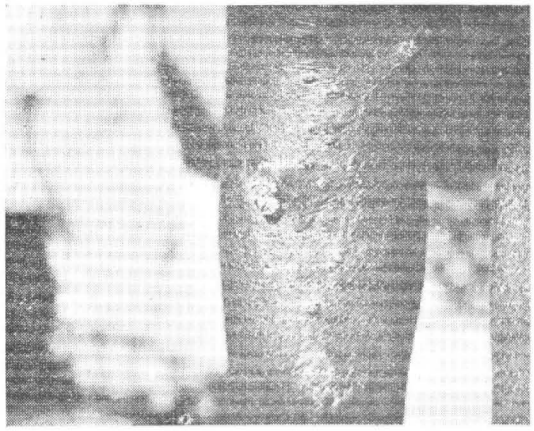

**Fig. 2.** Papules, nodules and plaques on the left leg showing verrucous surface.

These were verrucous, nodular and friable. Histopathology was similar to case 1. The patient was advised surgical treatment.

### Case 3

A 34-year-old man presented with localized, asymptomatic, papulo-nodular lesions on the left leg present for many years. The lesions had coalesced to form a plaque. The surface was dry, rough and verrucous (Fig. 2). Blood oozed whenever the lesion got traumatized. The histopathology was similar to that in case 1.

### Comments

Usually the lesions of AC are present at birth but in some cases, these do not appear until childhood or adulthood.[5] The majority occur on the lower leg and foot. These may be associated with varicosity of the superficial veins and hemangiectatic hypertrophy. All 3 of our cases started in adulthood and none was associated with varicosity or hypertrophy. The histopathology was characteristic in all cases. Patchy parakeratosis may be seen over the crests of the epidermis,[3] and when thrombosis occurs there is parakeratosis rather than hyperkeratosis.[9] Thrombosis and the resultant parakeratosis in angiokeratoma has the optical qualities of colour, intensity and surface reflection which mimic nodular melanoma.[9]

Imperial and Helwig[3] reported presence of keratin cysts in 3 out of 21 of their cases. In a third of their patients, the hemangioma extended laterally under the non-papillomatous epidermis. This may account for a high rate of recurrence when excision is limited to the clinically verrucous area. Just as lymphangiomas have dilated vessels angiokeratomas occasionally have dilated lymphatics.[4]

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