

## ANGIOKERATOMA CIRCUMSCRIPTUM (A case report)

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### Summary

A case of angiokeratoma which is a rare disease has been reported.

### Review of Literature

Angiokeratoma is a vascular malformation of the vessels of papillary layer of dermis. It represents telangiectasia of papillary vessels with hyperkeratosis of overlying epidermis. The term angiokeratoma is derived from three Greek words meaning vessels, horn and tumour respectively. Mibelli<sup>1</sup> reported the first case of angiokeratoma on the basis of histological appearances of marked hyperkeratosis and vascular dilatation of papillary capillaries. Though angiokeratoma clinically resembles wart or keratoma, Mibelli established it as a definite dermatological entity on the basis of precise clinical and histological findings. Five distinct clinical varieties of angiokeratoma are now recognised.

1. Angiokeratoma of Mibelli
2. Angiokeratoma of Fordyce.
3. Angiokeratoma of Fabry.
4. Angiokeratoma circumscriptum of Fabry.
5. Solitary and multiple angiokeratoma.

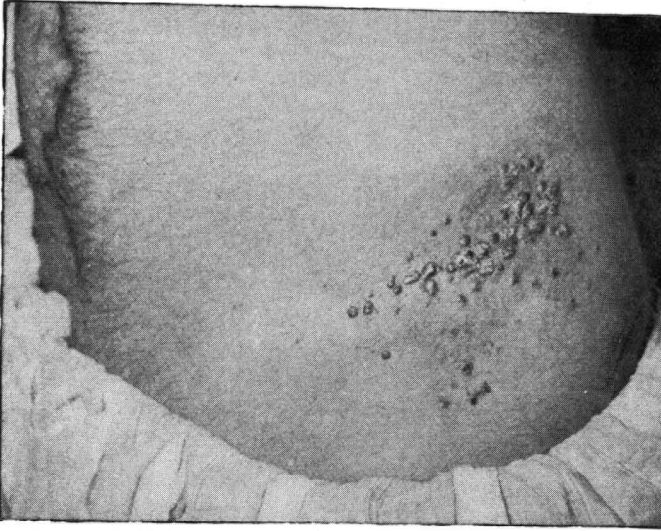
Angiokeratoma circumscriptum is a very rare disease and about 50 cases were reported in world literature till 1967. The term angiokeratoma circumscriptum is reserved for lesions corresponding clinically and histologically to Fabry's description<sup>2</sup>. Lesions appear in early infancy and are unilateral with a predominance in females (M:F::1:3). Sites of predilection are lower limbs, trunk, and upper limbs in that descending order. Clinically the condition presents as multiple papules, grouped often in irregular linear or zosteriform distribution. Some papules enlarge to become nodules with surfaces uneven and warty. There is no tendency for spontaneous involution. Following factors are important in the histological diagnosis:

1. Marked dilatation of dermal papillary vessels forming large lacunae.
2. Acanthosis and elongation of rete-ridges, partly or completely encasing vascular lacunae forming blood cysts.
3. Intimate vascular and epidermal relationship.
4. Absence of many dilated vessels or capillaries in the deeper dermis unlike in verrucous haemangioma.

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**Fig. 1** Multiple papules are distributed on left side of abdomen in Zosteriform pattern.

5. Varying degree of hyperkeratosis, acanthosis not being a prominent feature of lesions on trunk, arm and thigh.
6. Collarette formation which is seen in 25% of cases.
7. Organised and organising thrombi which are present in 30% of cases.

Lynch et al<sup>3</sup> and Cavanagh<sup>4</sup> have described this condition under the name of congenital angiokeratoma. Radia-

tion angiokeratoma of scrotum following grenz radiation for lichen simplex chronicus has been described by Abe et al<sup>5</sup>. Exact aetiology of angiokeratoma circumscriptum is not known. Imperial and Helwig<sup>6</sup> are of the view that angiokeratoma represents telangiectasia of papillary vessels acquired as a result of trauma. They presume that injury to papillary capillaries causes impairment in contractility and this leads to their dilatation. Since the papillary capillary is composed of



**Fig. 2** Shows hyperkeratosis, acanthosis, dilatation of papillary vessels and their intimate relation with epidermis.

only endothelial cells, a thin membranous sheath and a layer of pericytes, stasis will induce back pressure and dilatation of vessels. Later secondary fragmentation of the elastic tissue due to over distension leads to permanent damage and dilatation. The role of trauma is supported by the presence of lesions on lower limbs and on trunk mainly at the site of binder and their short duration. Elastic tissue support of vascular lacunae and venules in upper dermis is deficient or degenerated at the affected site but is normal in adjacent deep corium. They further explain that increase in number of dilated vessels is due to dilatation and not due to formation of new vessels. This view is also supported by Loria<sup>7</sup> who opined that angiokeratomas arise from preformed capillaries and are acquired with a congenital predisposition. Wertheim's view<sup>8</sup> also favours this and he has reported that there is actual increase in vascular elements in verrucous haemangioma in contrast to angiokeratoma of Mibelli where dilatation of capillaries takes place.

However, other authors do not agree with the above view. Thus, Lynch et al<sup>3</sup> think that congenital factors play a part. Their view is supported by the facts that lesions are distributed roughly along the course of a nerve or vessel and they appear at an early age. Occurrence of angiokeratoma in association with limb enlargement is cited as yet another supportive evidence<sup>9</sup>. Bruce<sup>10</sup> reported association of angiokeratoma with systemic hemangioma and points out that angiokeratoma presents a definite picture of angioma. Lever<sup>11</sup> has included verrucous hemangioma under the heading of angiokeratoma *circumscriptum*.

### Case History

A 15 years old boy presented with localised, mildly itchy papular eruptions. His parents noticed redness at the left iliac crest region in his early infancy.

Two months later purplish papules appeared at this site and gradually increased in size. Still later, surface of the papules turned dry, rough and scaly. Blood oozed whenever the lesion got traumatised. Intense itching resulted subsequent to topical indigenous medication and this brought the patient to the department of Dermatology of this Medical College Hospital. On interrogation, personal, family and past history were non-contributory. General physical examination and systemic examination revealed nothing abnormal.

### Local Examination

Local examination revealed purplish papules of variable size ranging from 1 to 5 mm on the left lateral aspect of abdomen in zosteriform pattern. Most of the papules were covered with scales and a few had blood crusts on their top.

### Histopathology

There was hyperkeratosis, acanthosis and elongation of rete ridges. Rete ridges appeared to enclose a vascular space. An intimate vascular and epidermal relationship was seen. Superficial dermis showed endothelial lined vascular spaces which were absent in the deeper region.

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