INDIAN J DERMATOL VENEREOL LEPR VOL. 46, NO. 5, 1980

ANGIOKERATOMA OF IMPERIAL AND HELWIG
(A case report)

K. K. RAJA BABU, * P. HARINARAYANA, † B. VIJAYAKUMAR, ‡
T. SURYAPRAKASH RAO, § B. SEKHAR REDDY, || P. SYAMASUNDARA RAO ¶

Summary

An unusual case of angiokeratoma of Imperial and Helwig type with detailed clinical and histological features is presented. The difficulty in the clinical recognition of this type of vascular malformation is emphasised.

Angiokeratomas are not tumors in the real sense of the term, but are discrete or clustered telangiectasis with a tendency for secondary epithelial changes like acanthosis and hyperkeratosis. While some occur without any apparent cause, others may follow trauma or chronic irritation, and in quite a few there is some underlying vascular abnormality.

Although histologically all angiokeratomas show almost the same features, clinically five distinct types are recognised today[1].

1. Angiokeratoma of Mibelli
2. Angiokeratoma of Fordyce.
3. Angiokeratoma corporis diffusum (Fabry's disease)
4. Angiokeratoma corporis circumscriptum of Fabry
5. Solitary and multiple angiokeratomas of Imperial and Helwig.

The histological features of angiokeratomas are

1. Marked vascular dilatation of papillary vessels forming large lacunae in the papillary area of the dermis.
2. Altered appearance of the epidermis with acanthosis and elongation of rete ridges, partially or completely encasing the vascular lacunae forming the so-called blood cyst.
3. Intimate vascular and epidermal relationship.
4. The absence of many dilated vessels in the underlying dermis or of lobules of capillaries suggestive of haemangiomas.

Though histologically angiokeratomas are essentially similar, they differ clinically from each other by the sites of involvement, age of onset, sex predelection, presence or absence of systemic involvement, inheritance and cause.

The first four varieties of angiokeratomas mentioned above have been known of for a long time and their clinical

* Professor of Dermatology
† Additional Professor of Surgery
‡ Asst Professor of Dermatology
§ Tutor in Dermatology
|| Additional Professor of Pathology
¶ Professor of Pathology
Kakatiya Medical College and MGM Hospital, Warangal (AP)
Received for publication on 12—10—1979

and histological features were fairly well delineated. In 1967 Imperial and Helwig in an extensive clinical and histological study of 116 cases[2], recognised a fifth variety of angiokeratoma. They claimed this variety differed clinically in several ways from the known types of angiokeratoma. While these lesions could occur on any part of the body, more than half were located on the lower extremities. The age of onset was the 2nd to the 4th decades of life. Both sexes were affected. In 83% of the patients the lesions were solitary and in 17% they were multiple. There was no inherited predisposition for its occurrence. The lesions were comparatively smaller than those of angiokeratoma circumscriptum and on the average measured about 3mm. The colour of the lesions was variable. Most lesions were asymptomatic although in a few there was mild to moderate pain and in some intermittent bleeding occurred after mild irritation. In many patients the lesions were of less than 6 months' duration. Most of the patients were treated by local excision, and the rate of recurrence was 4.3%.

Imperial and Helwig believed that this type of angiokeratoma was basically a response to injury to the wall of papillary vessels by various factors such as trauma and chronic irritation. They considered that the pathological changes were the result of telangiectasia rather than the result of true angioma or naevus.

To our knowledge there have been no reports of angiokeratoma of Imperial and Helwig type in the Indian literature. This is understandable because angiokeratomas are not common and a clinical and histopathological diagnosis can not always be made with much certainity. We here under report a case of angiokeratoma which we believe is of the type described by Imperial and Helwig.

Case Report

A 50 year old farmer presented in early November of 1978 with a warty, hyperkeratotic greyish black nodule, 5 cm. in diameter and covered with hemorrhagic crusts, on the front of the right fore-arm and a similar nodule on the left lateral chest wall (Fig. 1). Both nodules were freely mobile not being fixed to underlying structures. Both the lesions had been present for less than one month. The lesions were not tender but bled easily on touch. The regional lymph nodes were enlarged and firm but not tender. The patient did not manifest any systemic abnormality. There was no history of trauma preceding the appearance of the lesions, and there was no history of cold sensitivity.

Fig. 1 Angiokeratomatous lesions of the trunk and Right fore-arm. Note the bleeding from the lesion on the trunk.

Examination of urine, stool and blood did not reveal any abnormality. Skiagrams of the chest were normal.

Under local anaesthesia total excision of both the lesions were done and skin from the front of the right thigh was grafted on the excised areas.

Histopathological examination of the excised tissue revealed the following features. The epidermis showed hyperkeratosis, acanthosis and marked elongation of retepegs. Within the epidermis there were large spaces filled with blood (Fig. 2). There was marked dilatation of dermal capillaries, essentially confined to the papillary dermis (Fig. 3.) There was no proliferation of the endothelial cells of capillaries, and there was no cellular atypia. There was scattered inflammatory cellular infiltrate.

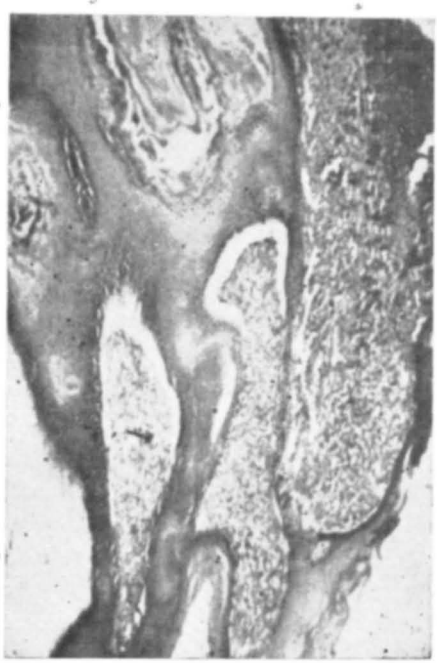

Fig. 2 Note the Hyperkeratosis, marked acanthosis and epithelial "blood cysts". (H & E × 100)

Lymphnodal biopsy from the left axilla showed only reactive hyperplasia.

A month after the excision of the lesions, small discrete nodular lesions appeared on the periphery of the excised area on the chest wall.

Comment

Although clinically a diagnosis of angiokeratoma was not suspected in this case the diagnosis was evident on histology. The histological features did not suggest either a granuloma pyogenicum or a verrucous hemangioma[3]. But the difficulty had been to fit it into one of the five types of angiokeratomas. The age of onset, the location of the lesions, and the absence of cold sensitivity, helped to exclude angiokeratoma of Mibelli. Because of the absence of the lesions on the scrotum, angiokeratoma of Fordyce was readily excluded. Absence of a familial history of the disease and absence of systemic involvement ruled out angiokeratoma corporis diffusum. Because most cases of angiokeratoma corporis circumscriptum appear at or immediately after birth[4] and because the lesions are almost always unilateral affecting mostly the

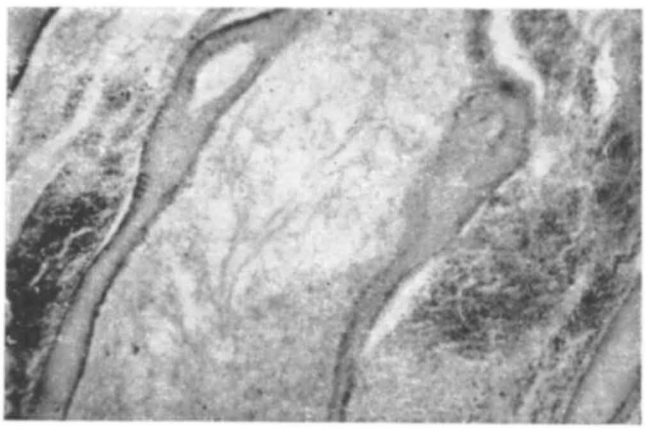

Fig. 3 Note the dilated capillaries in the papillary dermis. (H & E × 100)

lower extremities, this diagnosis was not considered in the present case. The present case thus seems to be one of angiokeratoma described by Imperial and Helwig. This case differs from the Imperial Helwig type in the size of the lesions. While most lesions studied by Imperial and Helwig were small ranging in size between 2 and 10 mm. in diameter, the sizes of the lesions in the present case was about 5 cm. each. As solitary and multiple angiokeratomas are only reactive telangiectasia due to trauma, the rapidity of growth and the consequent large size of the lesion is explainable.

But what we are not able to explain fully, is the prompt recurrence of one of the lesions. There were no features suggestive of a malignant angio-endothelioma[5]. Recurrences in angiokeratoma are by no means rare, and the recurrence in the present instance is probably due to incomplete excision of the reactive tissue.

References

1. Agarwal RR and Gupta S: Angiokeratoma circumscriptum, Indian J Dermatol Venereal Lepr, 1979, 45 : 122.
2. Imperial R and Helwig EB : Angiokeratoma - A histopathological study, Arch Derm 1967, 95 : 165.
3. Imperial R and Helwig EB : Verrucous hemangioma- a Clinicopathological study of 21 cases, Arch Derm 1967, 96 : 247.
4. Lynch CPJ, Kosanovich CM and Ga FB : Angiokeratoma circumscriptum, Arch Derm 1967, 96 : 665.
5. Griest MC and Callaway JL: Angio-endothelioma, Arch Derm 1978, 114:1690.

IX Annual Conference of
Indian Association of Dermatologists, Venereologists and Leprologists
at Indore
on January 8 - 10, 1981