ANGIOKERATOMA OF THE SCROTUM (FORDYCE)
A Case Report on Response to Surgical Treatment of Varicocele

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Abstract. In a patient with multiple angiokeratomas of the scrotum and double varicocele, radical surgical treatment of the varicocele was followed by regression of the angiokeratomas. A direct communication between the angiokeratomas and the underlying veins was demonstrated by phlebography. These facts indicate a probable causal relation between increased pressure in the veins and angiokeratoma of the scrotum.

The term angiokeratoma has been applied to several conditions, all of them characterized by some combination of angiomatosis and hyperkeratosis. Apart from angiokeratoma corporis diffusum (Fabry), which is a disorder of phospholipid metabolism, at least four types of purely cutaneous angiokeratoma are recognized: The Mibelli type, the Fordyce type, angiokeratoma circumscriptum and “solitary and multiple angiokeratoma” (Imperial–Helwig). The types differ from each other by location and clinical manifestation, but are histologically similar. Recently a survey has been published by Imperial & Helwig (5).

In angiokeratoma of the scrotum (Fordyce) (=a.k.f.) solitary or multiple nodular growths, rarely larger than 3–4 mm in diameter, are found on the scrotum, located along the superficial veins. The colour ranges from bright red to blue or blackish. The young lesions are soft and compressible while the older lesions may be firm and non-compressible, often keratotic and scaly, sometimes warty. Rarely, similar lesions may be found on the penis, the upper part of the thighs and the buttocks. Profuse bleeding from the lesions may occur.

Fordyce’s own patient (4) had a bilateral varicocele. Several authors (6, 9, 10) believe that vascular dilatation and angiomatous growths are induced by increased pressure in the veins, the epidermal changes being a secondary reaction. This theory is supported by the regression of the angiokeratomas following radical surgical treatment of varicocele in the patient reported here.

CASE REPORT
A 35-year-old man in good health was referred to the skin clinic in June 1968 because of several episodes of profuse bleeding from small blue-redish growths on the scrotum during the previous 2–3 months. Varicocele had been present on both sides for several years but caused no complaints except periodic pains in the groins. The skin lesions had developed during the previous two years.

Examination revealed a large varicocele on both sides, most pronounced on the left side, and multiple angiokeratomas equally distributed on both sides (Fig. 1).

In October 1968 division and ligation of the internal spermatic vein was performed on both sides just above the internal inguinal annulus as described by Ivanisevich (8). During the operation a retrograde phlebography was performed on the right side. This showed communications between the angiokeratomas and the underlying veins (Fig. 2).

Biopsy from a skin lesion showed some parakeratosis, but no acanthosis and no hyperkeratosis. In the papillary layer of the dermis a markedly increased number of dilated capillaries with normal endothelium was found.

The patient was reexamined in October 1969. The varicocele had disappeared, the angiokeratomas had regressed (Fig. 5), and the patient was without any complaints; especially, there had been no bleeding.

DISCUSSION
Some authors suggest that the primary cause of a.k.f. might be a pathological alteration either in
the walls of the vessels or in the supporting tissue (4, 10). Wile & Belote (11) stress the similarity of the histological picture of senile angiomas and a.k.f.

In a statistical study of a.k.f., Isaki (7) found a close relation to senile angioma. Bean (3) observed scrotal angioma in 70 out of 500 men. All but one were more than 50 years old. Both authors regard a.k.f. as nothing more than senile changes.

By comparison only 2 of 35 patients studied by Imperial & Helwig (6) were more than 40 years old. In 24 of the patients the angiomas were associated with conditions alleged to increase the pressure in the scrotal veins (varicocele, hernia, prostatitis, lymphogranuloma venereum, carcinoma of the bladder, thrombophlebitis and beri-beri). Twenty-three of the patients had multiple angiokeratomas and 12 of these had episodes of intermittent bleeding. The authors suggest that increased venous pressure can induce a.k.f.

In varicocele there is an increased pressure in the scrotal veins. This can be caused by tumors in the kidney or the colon, lymphnode metastases, hydronephrosis, etc. (secondary varicocele). This, however, is extremely rare. Varicocele is nearly always primary, i.e. venous dilatation without any obstruction, caused by valvular insufficiency in the internal spermatic vein. The scrotal veins empty when the patient is lying and retrograde flow will be noted if the patient stands. Furthermore, a pressure wave can be felt when the patient coughs (8).

The valves in the internal spermatic veins have been studied radiographically by Ahlberg et al. (1). These authors, too, have studied the competence of the valves in the internal spermatic vein in an autopsy-material comprising 30 men (2). They found lacking or incompetent valves in 50% on the left side and in 40% on the right side, which means that such a condition can exist without varicocele. Probably the venous pressure will also be increased in such cases. Furthermore, the anatomical difference on the two sides will cause a higher pressure on the left side. The left internal spermatic vein empty into the renal vein,
which is slightly compressed between the aorta and the superior mesenteric artery, while the right internal spermatic vein empty directly into the inferior caval vein. The above-mentioned might explain that in a.k.f. the left side is most often involved, and that varicocele almost invariably occurs on the left side.

In the material presented by Imperial & Helwig (6) two patients with varicocele and a.k.f. were operated upon. “Excision and ligation of the involved veins resulted in temporary regression of the angiookeratomas. However, in one of the patients the angiookeratomas not only reappeared, but developed on the skin of the adjacent upper thigh.” Unfortunately nothing is noted about the method of operation, nor the effect on the varicocele.

High ligation of the internal spermatic veins is the rational treatment of varicocele (8). In the patient reported here this operation resulted in disappearance of the varicocele and regression of the angiookeratomas. A direct communication between the angiookeratomas and the underlying veins could be demonstrated by phlebography. These facts render a causal relation between increased pressure in the veins and a.k.f. probable.

Possibly, high ligation of the internal spermatic vein should be carried out in the treatment of multiple a.k.f., especially if bleeding occurs, even if varicocele is not present.

REFERENCES


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