

CASE AND RESEARCH LETTERS

Unilateral Angiokeratoma of Fordyce[☆]

Angioqueratoma de Fordyce unilateral

To the Editor:

We present the case of a 66-year-old man with a past history of ischemic heart disease, systemic hypertension, and type 2 diabetes mellitus, who consulted for the appearance 8 months earlier of multiple lesions on the left side of the scrotum. The lesions had progressively increased in number and size.

On physical examination there were numerous, sessile, erythematous-purpuric unilocular papules of vascular appearance that only affected the left side of the scrotum and did not cross the midline. In some areas the lesions grouped together to form clusters. In addition, bluish cords were observed, which indicated dilatation of superficial blood vessels (Fig. 1).

Several lesions were biopsied. All revealed the presence of dilated vascular lumens in the papillary dermis in intimate contact with the epidermis, which was hyperplastic and hyperkeratotic. There was evidence of thrombosis and recanalization of some of the blood vessels (Fig. 2).

We were able to obtain a small blood sample from the dilated scrotal vessels, which allowed us to compare the levels of cortisol in the scrotal blood with those of the general circulation. The cortisol concentration in the scrotal blood was 20.6 µg/dL, considerably higher than that of the general circulation (15.3 µg/dL; normal range, 6.2–19.4 µg/dL).

The urology department confirmed the presence of a grade II varicocele on the left side of the scrotum.

Fordyce first described this type of angiokeratoma in 1896 in a 60-year-old man with bilateral varicocele. They are the most common form of angiokeratoma. They typically arise on the scrotum bilaterally, although they have been reported at other sites, such as the vulva¹ and on the body of the penis.²

The etiology and pathogenesis of scrotal angiokeratomas have not been clarified. It has been suggested that increased venous pressure could favor their appearance, and this would explain the association with varicocele.^{1,3} However, a study by Orvieto et al.⁴ found no association between varicocele and angiokeratomas. A limitation of that study was



Figure 1 Clinical appearance of the lesions, which affect only the left half of the scrotum. The right side is unaffected.

that the maximum age of inclusion was 55 years and the data were obtained by means of a questionnaire. Some authors consider that alterations of the blood vessel wall must be present for angiokeratomas to develop, though this may not be the only factor.¹

Varicocele more commonly affects the left testicle. This is attributed to anatomical differences in the venous drainage of the testicles. While the right testicular vein (also

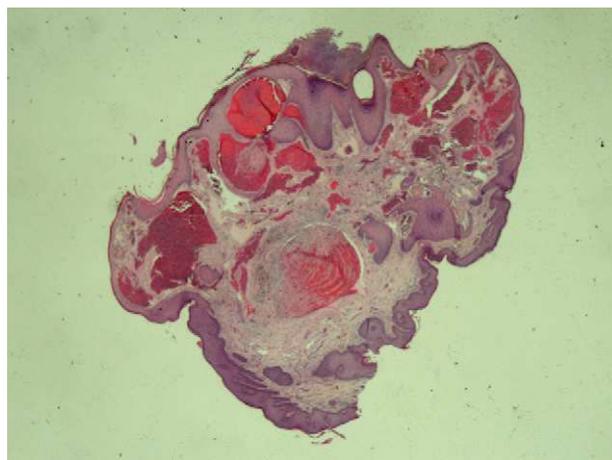


Figure 2 Histological appearance of a lesion, showing a pedunculated papule with grossly dilated blood vessels. Signs of thrombosis and recanalization can be seen. Acanthosis and hyperkeratosis are present in the epidermis. Some vessels appear to be included within the epidermis. Hematoxylin-eosin, original magnification $\times 40$.

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Table 1 Cases of Unilateral Angiokeratomas of Fordyce.

Reference	Age, y/Sex	Duration	Side Affected	Varicocele/Examinations	Treatment
9	21/Male	6 y	Left	Subclinical/Diagnosed by echo-Doppler	Carbon dioxide laser and Nd:YAG laser
8	32/Male	2 y	Left	No/Physical examination	Electrocautery
Present case	66/Male	8 mo	Left	Grade 2 varicocele/Ultrasound	Shaving of the larger lesions

Abbreviation: Nd:YAG, neodymium:yttrium aluminum garnet.

known as the internal spermatic vein) follows an oblique trajectory to drain directly into the inferior vena cava, the vein from the left testicle ascends vertically to the left renal vein, which means it is 8 to 10 cm longer than the vein on the right side. This leads to greater resistance to venous drainage on the left side. Varicocele is associated with testicular dysfunction, and sterility can develop due to the associated testicular hyperthermia.⁵ Another factor that could be important is retrograde blood flow from the left renal vein,^{5,6} although the results of relevant studies are contradictory.^{6,7}

In our patient, higher cortisol levels were detected in the testicular region than in the general circulation. As the testicles do not produce cortisol, these values would support the hypothesis of retrograde flow in this patient. The suprarenal glands secrete various hormones, including estrogens and adrenomedullin, a potent vasodilator.⁶ Retrograde blood flow in cases of varicocele could lead to higher hormone levels in the testicles than in the general circulation; this situation would favor vascular dilatation and, thus, the appearance of angiokeratomas.

Unilateral angiokeratomas are very rare; we have only found 2 reports describing this situation.^{8,9} The details of those cases are shown in Table 1. Although the patient described by Pande et al.⁸ did not present a clinically evident varicocele, it is recognized that up to 60% of varicoceles are not detected on physical examination and require ultrasound study for diagnosis¹⁰; we cannot therefore rule out the possible presence of a subclinical varicocele.

There are anastomoses between the left and right testicular circulations.⁵ If hormone levels do influence the development of angiokeratomas, these connections could explain the appearance of bilateral lesions, while unilateral disease may be due to abnormalities of the anastomoses. Another possible explanation would be the presence of mosaicism, which, in association with other factors such as increased pressure and high hormone levels, would cause angiokeratomas to develop only in genetically susceptible areas.

In summary, we have presented a patient with unilateral angiokeratomas of Fordyce in whom we detected higher cortisol levels in the scrotal circulation than in the systemic

circulation. Studies with a larger number of patients are required to determine whether retrograde blood flow with high concentrations of suprarenal hormones could play a role in the etiology and pathogenesis of these angiokeratomas.

References

1. Imperial R, Helwig EB. Angiokeratoma of the vulva. *Obstet Gynecol.* 1967;29:307–12.
2. Bechara FG, Huesmann M, Stucker M, Altmeyer P, Cansen T. An exceptional localization of angiokeratoma of Fordyce on glans penis. *Dermatology.* 2002;205:187–8.
3. Agger P, Osmundsen E. Angiokeratoma of the scrotum (Fordyce). *Acta Dermatovenereol.* 1970;50:221–4.
4. Orvieto R, Alcalay J. Lack of association between varicocele and angiokeratoma of the scrotum (Fordyce). *Military Med.* 1994;159:523–4.
5. Fretz PC, Snadlow JI. Varicocele: current concepts in pathophysiology, diagnosis and treatment. *Urol Clin N Am.* 2002;29:921–37.
6. Ozbek E, Yurekli M, Soyulu A, Davarci M, Blabay MD. The role of adrenomedullin in varicocele and impotence. *BJU Int.* 2000;86:694–8.
7. Steeno O, Koumans J, De Moor P. Adrenal cortical hormones in the spermatic vein of 95 patients with left varicocele. *Andrologia.* 1976;8:101–4.
8. Pande SY, Kharkar D, Mahajan S. Unilateral angiokeratoma of Fordyce. *Indian J Dermatol Venereol Leprol.* 2004;70:377–9.
9. Bechara FG, Altmeyer P, Jansen T. Unilateral angiokeratoma scroti: a rare manifestation of a vascular tumor. *J Dermatol.* 2004;31:39–41.
10. Pierik FH, Dohle GR, van Muiswinkel JM, Vreeburg JT, Weber RF. Is routine ultrasound advantageous in fertile men. *J Urol.* 1999;162:1618–20.

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