

An Atlas of Lumps and Bumps, Part 4

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Angiokeratoma of Fordyce

“Angiokeratoma” (derived from 3 Greek words meaning vessels, horn, and tumor, respectively) refers to a benign vascular lesion in the papillary dermis characterized by ectasia of blood vessels often accompanied by overlying hyperkeratosis.¹⁻³ In angiokeratoma of Fordyce, the angiokeratoma is most often situated on the scrotum in men and labia majora in women.³ Angiokeratoma of Fordyce usually develops in individuals aged older than 40 years.^{4,5} The condition is most common in White and Japanese populations.² In a cross-study of 213 White/European American adults (127 men and 86 women), angiokeratomas of Fordyce were detected in 47 (37.0%) men and 17 (19.8%) women.⁶ Presence of at least 1 angiokeratoma of Fordyce was significantly associated with male sex (OR, 2.4; 95% CI, 1.3-4.5; $p < 0.001$) and age older than 50 years (OR, 3.4; 95% CI, 1.7-6.7; $p < 0.008$).⁶ In the older age group, the condition may be associated with conditions

with increased localized venous pressure such as inguinal hernia, thrombophlebitis of the scrotum, and varicocele.^{3,7-9} However, in patients younger than age 20 years, usually no such association exists.⁹ On the other hand, angiokeratoma of Fordyce may be the first sign of Fabry disease, a rare inherited lysosomal storage disorder caused by deficiency of α -galactosidase A.^{7,10}

Clinically, angiokeratoma of Fordyce presents as multiple, well-circumscribed, dome-shaped papules, 2 to 5 mm in diameter mainly on the scrotum in men (Figures 1 and 2) and labia majora in women (Figure 3).^{1,3,11,12} Rarely, the lesions can be found on the prepuce, glans penis, and penile shaft in men and clitoris in women.^{1,3} Lesions on the inguinal folds, lower abdomen, buttocks, and inner thighs have very rarely been reported.^{2-4,10} The lesions are usually bilateral, although unilateral angiokeratomas have rarely been reported.^{4,10,13}



Figure 1. Angiokeratoma of Fordyce presents as multiple, well-circumscribed, dome-shaped papules mainly on the scrotum in men.



Figure 2. Angiokeratoma of Fordyce presents as multiple, well-circumscribed, dome-shaped papules mainly on the scrotum in men.

The color of the lesions may be black, bluish-black, dark red, red, purple, or blue.^{3,7} Newer lesions are often smaller, red, shiny, soft, compressible, and smooth, while longstanding lesions are often larger, darker, firm, noncompressible, keratotic, scaly, and sometimes warty.^{3,6,7} Confluence of telangiectases may lead to a red scrotum.^{3,7} Most lesions are asymptomatic but irritation, pain, burning sensation, pruritus, and bleeding may occasionally occur⁵

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This article is part of a series describing and differentiating dermatologic lumps and bumps. To access previously published articles in the series, visit <https://bit.ly/35J11tv>.



Figure 3. Angiokeratoma of Fordyce presents as multiple, well-circumscribed, dome-shaped papules mainly on the labia majora in women.

The diagnosis is mainly clinical. The use of dermoscopy facilitates visualization of dark or red lacunae, whitish veil, peripheral erythema, and hemorrhagic crusts, which are features of angiokeratomas.^{3,10,15}

Idiopathic Scrotal Calcinosis

Scrotal calcinosis refers to deposits of insoluble calcium salts (in particular, calcium phosphate) in the scrotal skin leading to the formation of papules and nodules within the scrotal skin. Idiopathic scrotal calcinosis is a rare condition; the exact incidence of which is not known, as information on this condition is mostly derived from case reports.¹⁶ The condition typically presents in the third to fourth decades of life.¹⁷⁻¹⁹ Idiopathic scrotal calcinosis is more common in dark-skinned individuals, suggesting an ethnic susceptibility.²⁰⁻²²

Scrotal calcinosis may result from dystrophic calcification of an epidermal inclusion cyst, an eccrine epithelial cyst, or a degenerated dartos muscle of the scrotum.^{18,22,23} Rarely, scrotal calcinosis may result from metastatic calcification secondary to abnormal calcium and



Figure 4. Scrotal calcinosis is characterized by multiple, slow-growing, firm to hard, papules or nodules within the scrotal skin.

phosphate metabolism that predisposes to calcium precipitation, or it may develop de novo.¹⁶ When scrotal calcinosis develops de novo, the condition is referred to as idiopathic scrotal calcinosis. Idiopathic scrotal calcinosis occurs in the absence of known tissue injury or systemic disease.¹⁶ In particular, the serum calcium, phosphate and parathyroid hormone levels are normal in patients with idiopathic scrotal calcinosis.²³ There is no evidence of residual cyst and epithelial lining around the calcified nodule.²³⁻²⁵

Scrotal calcinosis is characterized by multiple, slow-growing, firm to hard, papules or nodules within the scrotal skin (Figures 4 and 5).^{18,24,26} However, lesions may be solitary or pedunculated.¹⁸ The lesions tend to increase in size and number with time.²⁷ They are initially skin-colored but may become whitish to yellowish or brownish over time (Figures 4 and 5).^{26,27} The lesions vary from 2 mm to 2 cm in diameter.¹⁶ Scrotal calcinosis is usually bilateral and asymptomatic.¹⁸ Uncommonly, the lesions may be mildly itchy or painful.^{18,27} Some patients may experience a feeling of heavy or dragging sensation in the scrotum.¹⁷ Lesions may break down



Figure 5. Nodules are initially skin-colored but may become whitish to yellowish or brownish over time.

spontaneously or when compressed to produce a white chalky material.¹⁷

Idiopathic scrotal calcinosis is benign but can be socially embarrassing. Affected individuals may have low self-esteem and fear of sexual dysfunction, which may affect intimate relationships with sexual partners.^{17,20} As such, the condition may have an adverse effect on quality of life.^{18,20} Superimposed infection, inflammation, and ulceration of the lesion may also, albeit seldom, occur.^{22,26,27}

The diagnosis is usually a clinical one. A biopsy of the lesion is usually not necessary unless the diagnosis is in doubt. The condition is benign, and the prognosis is good.

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