Angiokeratoma of Fordyce - A Rare Cause of Scrotal Papules

Nga CN[1], Akinkugbe AO[1], Ayanlowo OO[1], Otrofanowei E[1], Akinde R[2], Adekanmbi A[1].

[1]Department of Medicine, Lagos University Teaching Hospital/College of Medicine, University of Lagos
[2]Department of Pathology, Lagos University Teaching Hospital/College of Medicine, University of Lagos
Correspondence :
Dr. Nga, Charles Nnamdi
Department of Medicine
Lagos University Teaching Hospital
Idiaraba, Lagos.
Email: charlesnga2001@yahoo.com

ABSTRACT

Introduction : Angiokeratoma of the Fordyce is a rare condition comprising small painless, dark red to purple papules with a rough scaly and hyperkeratotic surface. They are composed of surface dilated capillaries likened to scrotal 'pimples'. They are a benign condition that can cause great cosmetic and psychosocial disturbance to the patient. Surgical excision, LASER, cryotherapy, electrocautery are some of the modalities of treatment as medical therapy is not encouraging.

Keywords: Angiokeratoma of Fordyce, Angiokeratoma circumscriptum, scrotum, benign papule

INTRODUCTION

Angiokeratoma of Fordyce, also known as angiokeratoma circumscriptum of the genitalia, is a rare cause of benign papular eruptions of the scrotum especially in blacks.[1,2]

The exact aetiology is unknown, but postulations in the elderly may be due to a degenerative disorder secondary to local venous hypertension.[2-4,8] They may present as multiple bright red papules on the scrotum/ labia majora. In blacks they may appear as hyperpigmented papules on the genitalia. They are usually asymptomatic but a few of the patients may itch, bleed or express anxiety because of misdiagnosis.[2-8]

It is believed that males are more affected than females but the exact male to female ratio is unknown. It occurs more commonly in the elderly age groups.[2,4,7,8] Histologic sections show hyperkeratosis and superficial dermal vascular dilatation.[2,4,8] If treatment is required, then local destructive methods like laser, electrocautery or cryotherapy may be performed. Fordyce angiokeratoma could be very distressing in some patients because they may be confused as sexually transmitted diseases (STDs) or some form of cancers. [1-8]

The aim of the case report is to document the presence of this rare condition and to increase awareness and index of suspicion among clinicians in order to adequately manage potential cases.

CASE

A 35 year old trader presented to the dermatology clinic with complaints of “pimple- like” eruptions on the scrotum lasting for 4 years. This was associated with generalised itching of the body which is most marked at the scrotal sac. He had visited several peripheral clinics and pharmacies where he was given antibiotics, anti-histamines (topical and systemic) as well as herbal mixtures. He was also subjected to various laboratory investigations including venereal disease screenings. These did not improve the symptoms, hence his referral to the dermatology clinic of the Lagos University Teaching Hospital.

There was no history of penile discharge, dysuria or other urinary symptoms. There was no fever or weight loss. He is married with two children. There was no similar skin condition with the wife. He had no background medical problems.

Examination findings revealed a normal male phallus, testicles and scrotal sac. Multiple hyperpigmented papules, more than seven in number, measuring 2-3mm in diameter, were seen

on the scrotal sac with scratch marks (Figure 1). Examination of the other systems was essentially normal. At presentation, differential diagnoses entertained were pilar cysts, steatocystoma multiplex and epidermal inclusion cysts. Results of Full blood count, erythrocyte sedimentation rate, electrolyte, urea, creatinine, liver function test, retroviral screening, hepatitis B and C screening were either normal or negative.

A punch biopsy was performed and the histologic section of the tissue showed acanthosis and hyperkeratosis of the epidermis with elongation of the rete ridges. The underlying dermis contained numerous dilated thin-walled congested capillaries some of which are thrombosed; while few showed recanalization (Figures 2 and 3). A histologic diagnosis of Angiokeratoma circumscriptum of the scrotum (Angiokeratoma of Fordyce) was made. He was placed on topical 1% hydrocortisone cream to be applied lightly on the scrotum for 2 weeks initially, and subsequently on alternate days for 1 week; and tablets loratadine 10mg daily. He was also counselled on other management modalities which included surgical excision, LASER, electrocautery and cryotherapy. Lesions disappeared after some days of commencing treatment only to reappear whenever the topical agent was discontinued.

He was counselled on the benign nature of the condition; and the fact that it was not a sexually transmitted infection. However, he was lost to follow up after some months.

DISCUSSION

Angiokeratoma of Fordyce is a rare condition[1][3]. First described in 1896 by an American dermatologist: John Addison Fordyce, who reported the case of a 60-year-old man who had angiokeratoma that was associated with bilateral varicoceles[1]. They are small, painless, red to purplish papules 1-3mm in diameter in Caucasians and hyperpigmented papules in Blacks. They are composed of surface blood vessels (dilated capillaries) and hyperkeratosis of the overlying epidermis. They may be likened to pimples of the scrotum.

The papules are usually located on the scrotum, shaft of penis, labia majora, inner thigh, or lower abdomen. Precise data on their frequency and distribution are lacking [2]. The principal morbidity comes from bleeding, anxiety, and overtreatment due to misdiagnosis by physicians[2]. Anxiety over this skin condition usually arises because of the cosmetic concern about what their partners may think, fear that they may be a sexually transmitted disease like genital warts or that the individual may have some form of genital cancer such as melanoma. It is predominantly a condition of the Whites and Japanese with few cases reported in Blacks [2]. Males are believed to be far more affected than females though this is yet to be correlated and there may be under- reporting of female cases. Direct figures for comparing sex prevalence do not exist.[2] It is most prevalent in the fifth decade of life though it may occur in all ages.

Other types of angiokeratomas include Sporadic angiokeratoma which is a solitary lesion seen in those above the age of 40 years.[4] Angiokeratoma corporis diffusum, which is a serious inherited enzyme defect with widespread lesions concentrated on the lower limbs and groin, may present with fever and organ failure. Mibelli Angiokeratoma are referred to as telangiectatic warts located on the toes and fingers.[7]

Angiokeratoma of Fordyce is most commonly found on the scrotum. It can also be seen on the shaft of the penis, labia majora of the vulva, inner thigh and lower abdomen. It is most prevalent in those over 40 years. The prevalence is reported to increase with age, from 0.6% in 16-year-old males to about 17% in those older than 70 years[2]. Men are more often affected than women. It may be a single lesion or multiple lesions of over 100 in number. The lesions are small, red and less scaly in younger patients whilst they tend to be larger, blue/black and with overlying scales in older individuals. They are usually symptomless and may only be noticed when they bleed after scratching or after sexual intercourse.

The exact cause of Fordyce angiokeratoma is unknown however some experts regard the lesion as a degenerative disorder. Local venous hypertension might play a causative role given that the condition is more common in patients with coexisting varicocele, hydrocele and haemorrhoids.[6][8]

Angiokeratoma of Fordyce may be confused with genital warts, melanomas, cherry hemangiomas, squamous cell carcinoma or melanocytic nevi. Angiokeratoma of Fordyce must also be differentiated from angiokeratoma corporis diffusum (Fabry's syndrome), which is the only other type that can involve the genitalia[5]. Fabry's syndrome is a rare serious inherited disorder caused by a deficiency of an alpha-galactosidase enzyme, ceramide trihexosidase and the angiokeratomas may

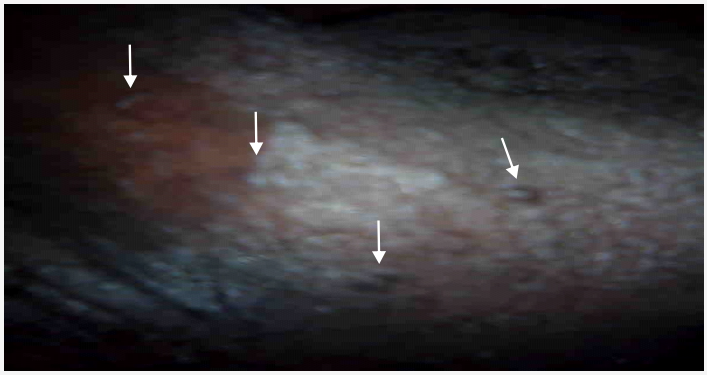
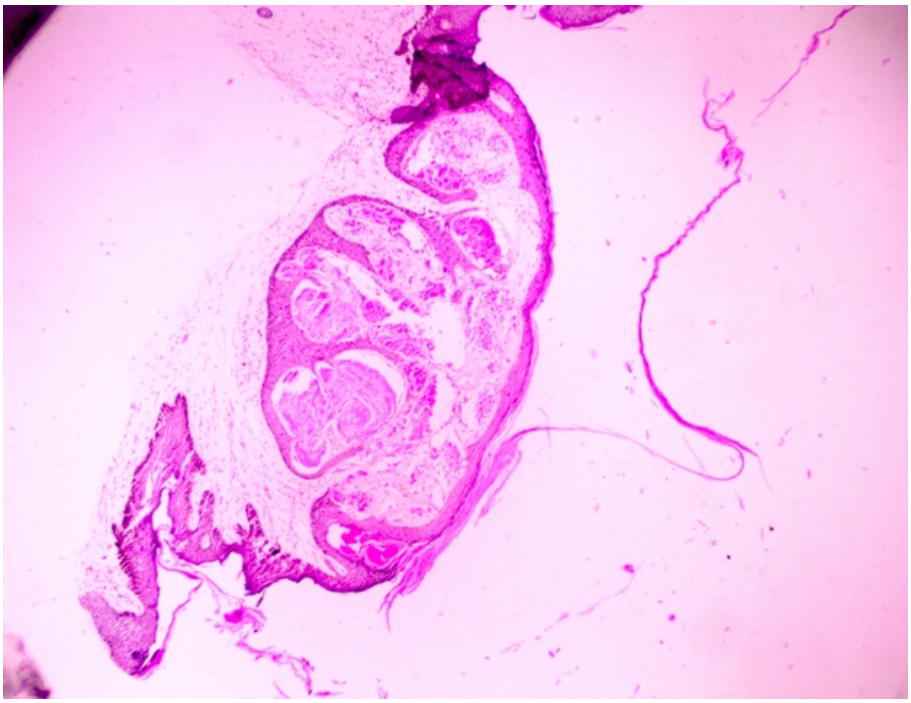
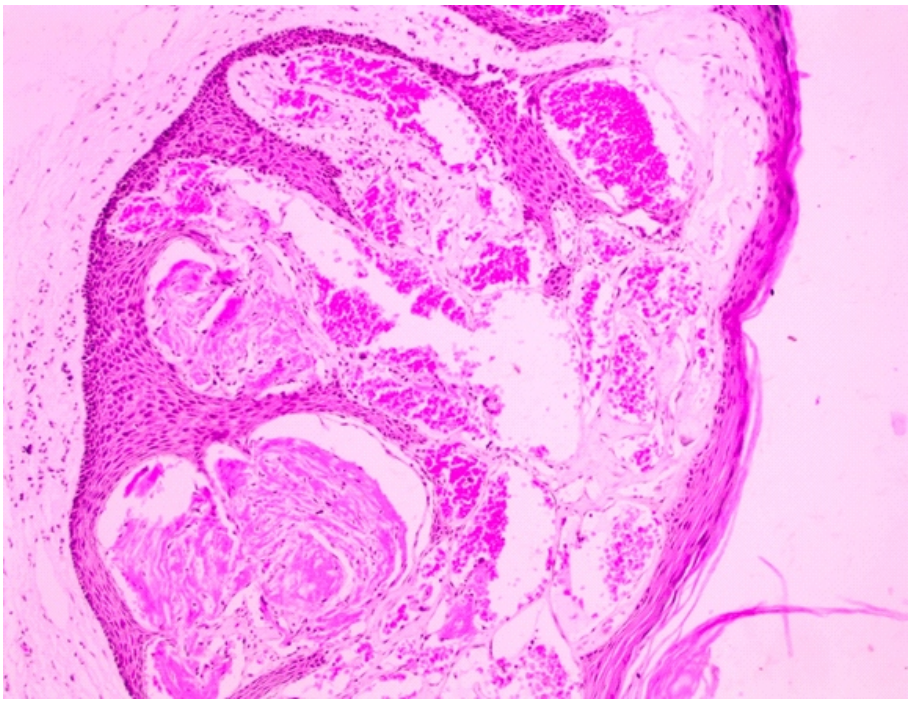


Figure 1: Hyperpigmented papules on the scrotal sac (see arrows)





Figures 2 & 3: Histology of the case

affect other parts of the body like the lower trunk and groin. There may be fever with associated internal organ failure eg kidneys and heart.[4]

A high index of suspicion is imperative as clinical diagnosis and histology are sufficient for diagnosis as such this condition does not warrant imaging studies [2].

Treatment involves counselling and reassurance regarding its benign nature. If cosmetic or bleeding concerns arise, then other treatment modalities may be employed because drug therapy is not encouraging. These modalities include surgical excision, LASER, cryotherapy, electrocautery and sclerotherapy[1][2][3][4][5][6][7][8].

In conclusion, angiokeratoma of Fordyce is a benign scrotal lesion; and this report aims to document the occurrence of this rare condition as a differential of scrotal papules in order to increase the awareness and heighten the index of suspicion amongst physicians and dermatologists.

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