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Case Report

Unusual Bleeding From Scrotal Skin - A Case Report with Review of Literature

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ABSTRACT

Angiokeratomas include diverse conditions characterized by hyperkeratosis and superficial dermal vascular ectasia on histopathology. They are commonly asymptomatic and present with 1- to 6-mm, blue-to-red papules on a scaly background. They have been described on the scrotum, shaft of penis, vulva, inner thigh and lower abdomen. Angiokeratoma of scrotum (Fordyce's spots) is a very rare entity. An association with increased venous pressure as in cases of varicocele has been noted in males. Morbidity in such a presentation is due to bleeding, anxiety and misdiagnosis leading to overtreatment. At times they can present as episodes of recurrent bleeding or as emergencies in the form of excessive bleeding. In general no treatment is required, however if required, local destructive procedures are employed like excision, electro coagulation, cryotherapy, or laser therapy. We describe here a case of 85 year old male who presented to us with complaints of numerous reddish spots over scrotal skin and painless bleeding from the same site. On local examination there were red maculopapular patches over the scrotal skin. No ulcer or growth was present. Rest of the perineal area and groin were normal. Systemic examination was normal except that patient was hypertensive. No immediate diagnosis was possible due to the rarity of presentation. Literature review helped us to make a presumptive diagnosis of angiokeratoma of scrotum. Patient undergone excision of involved skin with primary closure. The patient recovered well and is being followed up on out-patient basis with no recurrence or complication even after 1 year.

Keywords: Scrotal bleeding, angiokeratoma of scrotum, maculopapular, ectatic vessels, epidermal hyperplasia.

INTRODUCTION

Angiokeratomas include diverse conditions characterized by hyperkeratosis and superficial dermal vascular ectasia on histopathological examination (HPE). [1] They are mostly asymptomatic, 1- to 6-mm, blue-to-red papules present on a scaly background located on the scrotum, shaft of penis, vulva, inner thigh, or lower abdomen. Angiokeratomas can be divided into localized or systemic varieties. Angiokeratoma of scrotum is a very rare type of localized form of angiokeratoma first described by John Addison Fordyce in 1896. [2] Hence, it is also referred to as Fordyce's spots. We describe here a case of 85 year old male who presented to us with recurrent painless bleeding from scrotal skin.

CASE REPORT

An 85 year old male presented to us with several reddish spots on the scrotum for last 2 years. He also gave history of 2

episodes of spontaneous profuse painless oozing of blood from scrotal skin with the most recent episode 2 days back. Patient applied pressure on the bleeding area for prolonged time to stop the bleeding on both occasions. He had no other complaints. There was no history suggestive of trauma to scrotum, or any ulcer or mass in scrotum. He was neither sexually active nor had any prior medical history except that he was a known hypertensive for last 5 years and his blood pressure was well controlled on tablet amlodipine 5 mg once a day. There was no similar history in any of the family members. He had no present or past addiction. On physical examination his general condition was fair and his vitals were within normal limits for his age with blood pressure on higher side of normal for age. Systemic examination revealed no abnormality. On local examination, there were multiple (>100), 1-3mm, black to red, maculopapular, hyperkeratotic, erythematous, non-tender lesions over the surface of scrotum on both sides (Figure 1). Bilateral testis and epididymis were normal. No growth or any ulcer was present. Rests of the perineum and groin area were also within normal limits with no regional lymphadenopathy. Urinalysis detected no abnormality. Patient's coagulation profile and platelet were in normal range. An ultrasonography (USG) of the scrotum and abdomen revealed no abnormality. No immediate diagnosis could be made. We did a literature review and found that the presentation closely resembled that of angiokeratoma of scrotum. With this provisional diagnosis, a dermatology opinion was sought. The dermatologist initially managed the patient with topical corticosteroid cream applied locally, to which the patient responded partially in the form of decreased number and size of spots. However after 2 months on treatment, patient again had an episode of profuse bleeding from scrotal skin. Due to non-availability of laser and cryotherapy facilities at our center, the patient was referred back to us for definitive intervention. A decision to excise the involved skin was taken. Surgical excision of involved part of scrotal skin along with presumably involved subcutaneous tissue and capillaries was done with maintenance of a free margin with primary closure of skin flap (scrotoplasty). Biopsy confirmed the presence of angiokeratoma due to the presence of epithelial hyperplasia, acanthosis and hyperkeratosis, and ectatic thin-walled superficial blood vessels (Figure 3). Patient is being followed on out-patient basis and has no recurrence or complication even after 1 year.

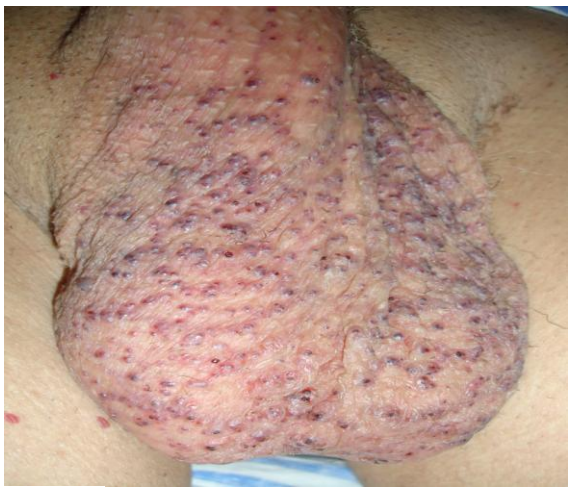

Figure 1: Multiple, 1-3mm, black to red, maculo-papular, hyperkeratotic, erythematous, non-tender lesions on a scaly background over scrotal skin extending up to ventral aspect of root of penis.

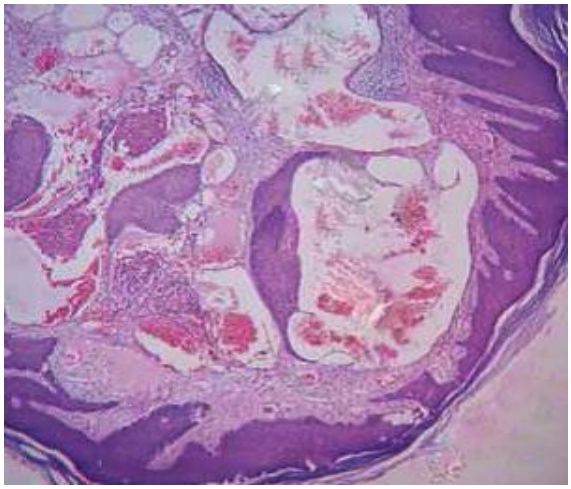

Figure 2: Epidermis is showing irregular acanthosis, hyperkeratosis. Thin-walled, ectatic vessels are seen in the papillary dermis. (H&E stain, magnification ×40)

DISCUSSION

Angiokeratomas are a group of vascular ectasias with variable

morphological forms. [1] They may be localized or systemic. Localized angiokeratomas include solitary papular angiokeratoma (usually occur over legs); angiokeratoma of the scrotum and vulva (Fordyce type); angiokeratoma circumscriptum naviforme (congenital form which presents unilaterally on the lower limb and buttock); and Mibelli type bilateral angiokeratomas (occurring on the dorsum of fingers and toes). [3] Generalized systemic angiokeratomas include angiokeratoma corporis diffusum (associated with Fabry disease and fucosidosis). [3] Histologic features are similar in all forms of angiokeratoma. [1,4]

Exact incidence of angiokeratomas of scrotum is unknown, but the incidence is reported to increase with age. [1,5,6] Literature review shows involvement of all age groups including neonates [2,5-7] however it is found to be more prevalent in people older than 40 years. [6] Our patient was an 85 year old male. The pathophysiology of angiokeratomas largely remains elusive, although increased venous pressure has been proposed to be a contributory factor. [8] Varicocele and conditions associated with increased venous pressure (e.g., hernias, epididymal tumors, intra-abdominal masses mostly urinary system malignancies, trauma, and thrombophlebitis) have been described in many studies as precipitating factors. [3] Imperial et al. [5] in their case series found precipitating factors in up to two thirds of patients. There are many cases and case series with no evidence of increased venous pressure as an inciting factor. [6,9] Angiokeratomas of Fordyce have also been reported in association with nevus lipomatosus, oral mucosal angiokeratomas, papular xanthoma and congenital lymphangiectasia-lymphedema. [3] There was no precipitating factor of evident cause in our case. No fatalities from this condition have been reported till date. [3] Patients are usually asymptomatic and may not be aware of the lesions. Bleeding is the most common presentation which may be spontaneous, after scratching or intercourse. [1,3,5,6] Pain or itching may be present but are uncommon. Painless bleeding is the major cause of morbidity and anxiety to patient in this condition. [3] Patient may present with the concern that the asymptomatic lesion over genitals is a form of sexually transmitted disease [10] or a malignancy. [11] Angiokeratoma of scrotum present as blue-to-red, papules or macules, 1-6 mm in diameter, with a mean of 3 mm with slight scales in the background. [3-5] Lesions are reported to be smaller, more erythematous, and less hyperkeratotic (scaly) in young patients as compared to older patients. [3,5] Numbers of lesions are variable but there can be hundreds of them. [3] Lesions have been described on the scrotum, shaft of penis, vulva, inner thigh and lower abdomen. [3] Scrotum is the most common site of angiokeratoma. [3] Differential diagnosis include - Angiokeratoma Corporis Diffusum (Fabry Syndrome), granuloma pyogenicum, cherry hemangioma, malignant melanoma, melanocytic nevi, squamous cell carcinoma and genital warts.

The diagnosis of angiokeratoma of scrotum is largely clinical and no imaging study is required though dermoscopy aids in diagnosis. Skin biopsy is required in doubtful cases. Biopsy characteristics include numerous dilated, thin-walled vessels located in the papillary dermis or superficial submucosa, with elongated rete ridges, overlying acanthotic epidermis with overlying parakeratosis and hyperkeratosis in epithelial lining. [4] Vascular spaces commonly get thrombosed and then re-canalize leading to the pathologic pattern known as papillary endothelial hyperplasia (Masson lesion). [12]

Incidental and asymptomatic cases are reassured regarding the benignity of lesions. In cases of recurrent bleeding or cosmetic concerns, several surgical options are at hand. Angiokeratomas of scrotum is a benign neoplasm and are reported to be not amenable to medical therapy. However in our case, partial response in the form of

decreased number and size of lesions was noted with the use of topical corticosteroid cream which was prescribed by the dermatologist. Sole use of local corticosteroid for treatment of scrotal angiokeratoma cannot be recommended on the basis of our single experience and its role need to be studied in a larger cohort of patients. Surgical excision under local, spinal or general anesthesia has been traditionally used for management of this condition and yields good cosmetic result owing to laxity of scrotal skin. [1,4,6, 12] Negativity of excised margins makes recurrences unlikely. [3] Various treatment modalities like cryotherapy using liquid nitrogen, [3] light electrocoagulation, [3] 578-nm copper laser, [13] pulsed-dye laser, [14] long-pulse 1064-nm Nd:YAG laser, [15] repeated local injections of 0.5% ethanolamine oleate or 0.25% sodium tetradecyl sulfate [16] have been successfully used in treatment of angiokeratoma of Fordyce with minimal and temporary adverse effects like mild pain, epithelial sloughing varying degree of scarring. One advantage of primary surgical excision scrotoplasty over other modalities is the histopathological confirmation of diagnosis, however it may be associated with prolonged hospital stay, wound infection and more pain; though our patient had none of these complaints. Follow-up is required after surgical intervention to see the cosmesis and to check any recurrence.

CONCLUSION

Angiokeratoma of scrotum is a rare clinical entity. Recurrent bleeding, fear of contracting a sexually transmitted disease, malignancy and cosmesis are the driving factors that bring the patient to a medical care provider. Management of this condition is largely surgical with primary surgical excision, lasers, intra-lesional chemical agents providing adequate relief of symptoms with minimal pain, scarring and rare recurrences. Primary surgical excision with negative margins is a valid treatment modality in this condition especially in those settings where more costly treatment options like lasers are not available and it further helps in histopathological confirmation of diagnosis.

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