Angiokeratoma of Fordyce: A diagnostic enigma

Dr. Anuradha Panchal, Dr. Nida Khan, Dr. Manish Kumar, Dr. Abhishek Mahadik, Dr. Dipesh Goel and Dr. Shweta R Verma

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Abstract
Angiokeratomas are rare, benign ectasias of scrotal dermal vessels with a male preponderance. They are usually asymptomatic, although rarely they may present as an acute bleed. We present a case report of a 27 year old male with multiple large papules over scrotum associated with intense pruritis. Excision of scrotal skin was performed. Patient was followed up for one year, without any recurrence.

Keywords: Angiokeratoma, diagnostic enigma, male preponderance

Introduction
Angiokeratoma of Fordyce is a benign condition presenting as red to purple papules over scrotum. It is due to anomalous ectatic veins [1]. It may present as multiple painless papules or as scrotal bleed [2]. It commonly affects scrotum, though penis may rarely be involved, even rare if the affliction of vulva [3]. Treatment options include electrocoagulation, yttrium laser, electrocoagulation and excision [4].

Case Report
A 22 year old male patient presented to surgical OPD with multiple papules over scrotum since 2 years. The papules were reddish in colour, few millimeters in size and had increased in number over the course of 2 years. Patient had developed itching over the papules over the past 6 months which was causing significant discomfort and had brought him to the OPD. He had no co morbidities. On examination, patient had multiple reddish brown papules, 2-6mm in size, over anterior surface of right scrotum, without any tenderness or active bleed. General and Systemic examinations were normal. Biopsy from the lesion confirmed diagnosis of angiokeratoma of fordyce. Patient was explained various treatment options and potential complications. Patient opted for surgical excision. Under spinal anesthesia, excision of a 3 x 5cm area of right scrotal skin with the angiokeratomas and primary closure was perfomed. Patient was discharged on post operative day 2. Patient was followed up for one year without any recurrence of the angiokeratoma.

Discussion
Fordyce first described Angiokeratoma in 1896 [5]. They are superficial, circumscribed lesions with vascular ectasia and hyperkeratotic surface [6]. It is commoner in older individuals. It has a male preponderance and occur commonly on the scrotum, although it can also affect the vulva in females. Rarely they are seen on the shaft of penis or glans. It presents with multiple papules, 1 to 6mm in diameter, that may be asymptomatic or bleed due to trauma or during sexual intercourse [7]. The exact etiology is unknown. They may be isolated lesions or a part of Fabry’s disease, urinary tract tumours, intra abdominal tumours and varicoceles [8]. Vulval angiokeratomas are associated with use of oral contraceptive pills, localised venous hypertension, phlebectasia, obesity [9]. Biopsy is essential to rule out other differentials including melanoma, pyogenic granuloma and angiosarcoma [10]. They are mostly asymptomatic or may present with pruritis or as an acute bleed. Histopathology shows marked dilatation of superficial dermal blood vessels with collarette at the periphery of the lesion. Treatment options include electrofulguration, cryotherapy, laser ablation, shave excision and surgical excision. Laser used is Nd:YAG or 532 nm KTP [11, 12]. Surgical excision of affected scrotal skin can be performed in case of extensive disease or large angiokeratomas. Recurrence is common.
Conclusion
Angiokeratoma of Fordyce is a rare condition that is often misdiagnosed. It is usually asymptomatic but may present with torrential bleed. Treatment is either for cosmesis, or symptomatic angiokeratomas, with laser ablation, cryotherapy or surgical excision.

Fig 1: Preop image of angiokeratoma of fordyce

Fig 2: Excised scrotal skin with angiokeratoma

References