[Downloaded free from http://www.meajo.org on Tuesday, November 25, 2014, IP: 41.234.27.97] || Click here to download free Android application for this journal

Case Report

Eyelid Angiokeratoma

Ramadan S. Hussein, Hala Kfoury[1], Yasser H. Al-Faky[2]

ABSTRACT

Angiokeratoma is a rare, usually acquired muco-cutaneous wart-like vascular lesion that is frequently reported in the scrotum. Bleeding may occur if angiokeratoma is excoriated or traumatized. We report an exceedingly rare solitary eyelid angiokeratoma in an otherwise normal middle aged male. Our case represents the second case in the English peer reviewed literature since 1966, when the first case of eyelid angiokeratoma was reported.

Key words: Angiokeratoma, Eyelid, Skin

Access this article online

Website: www.meajo.org

DOI: 10.4103/0974-9233.134702

Quick Response Code:

INTRODUCTION

Angiokeratoma is a rare, wart-like vascular skin lesion, which was first described by Mibelli in 1889.[1] It is usually acquired, although, congenital cases have also been reported.[1] It has male predominance and may affect different age groups, but is rarely seen in individuals with pigmented skin.[2] Despite being reported in the extremities, trunk and tongue, it has a predilection for the scrotum.[3] Since 1966, only a single report of eyelid angiokeratoma has been published as we conducted an electronic search of three databases (Medline, Scopus and Scirus) using angiokeratoma and one of the following key words (eyelid,ocular or peri-ocular).[4] We present the second case of solitary eyelid angiokeratoma in an otherwise normal adult male.

CASE REPORT

This was a case of a 28-year-old male patient who was presented with a painless purple-color skin lesion involving the left upper eyelid of 1 year duration without preceding trauma. He also reported spontaneous or traumatic recurrent bleeding whenever scratching the lesion. The lesion was raised, compressible, well-circumscribed and freely mobile over the tarsus, measuring 5 mm × 5 mm. It had a patchy, reddish surface discoloration [Figure 1a and b]. No detectable signs of inflammation, tenderness, regional lymphadenopathy or antedated eyelid lesion were noted. Patient was otherwise normal with no systemic illness and a normal coagulation profile. Ophthalmic examination was unremarkable.

The lesion was completely excised and histopathologic examination revealed skin comprising epidermis and most of the dermis. The epidermis showed acanthosis however, no significant hyperkeratosis was noted. The dermis showed dilated papillary vessels [Figure 2a and b]. Some blood vessels showed thrombosis. At the periphery, palisading of inflammatory cells around eosinophilic collagenous material was seen.

Based on the clinical and histopathologic features, a diagnosis of solitary papular angiokeratoma was made. At this stage, the patient was referred to a Dermatologist for further assessment that revealed neither other mucocutaneous lesions nor nail changes.

DISCUSSION

Angiokeratoma is a broad term for rare muco-cutaneous disorders characterized by hyperkeratosis and superficial dermal vascular malformations.[5] It can present as either acquired (Mibelli, Fordyce, solitary and multiple papular) or congenital lesions (angiokeratoma circumscriptum and angiokeratoma corporis diffusum).[1,6] It is frequently reported in

Departments of Dermatology, [1]Pathology, and [2]Ophthalmology, College of Medicine, King Saud University, Riyadh, Saudi Arabia

Corresponding Author: Dr. Yasser H. Al-Faky, Department of Ophthalmology, College of Medicine, King Saud University, P.O. Box: 245, Riyadh 11411, Saudi Arabia. E-mail: alfaky@gmail.com

[Downloaded free from http://www.meajo.org on Tuesday, November 25, 2014, IP: 41.234.27.97] || Click here to download free Android application for this journal

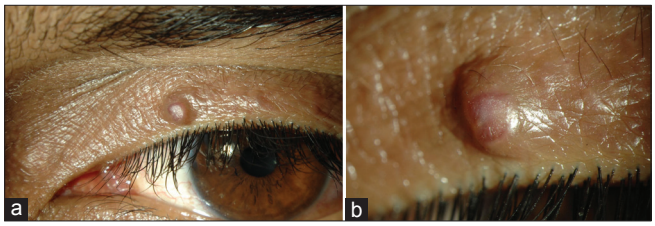

Figure 1: Angiokeratoma of the upper eyelid. (a) Shows a raised, well circumscribed papule measuring 5 mm × 5 mm (b) Shows a close up view to highlight the reddish surface discoloration and circinate border

the scrotum; nevertheless, extremities, trunk, vulva and tongue have been also reported.[3,5] Clinically, angiokeratoma consists of elevated, warty, slightly compressible, dark red to purple papules; yet small nodules or plaques can also be seen.[2,3] The surface and edges of the papules often have rough, hyperkeratotic scales. [3] Intra-epidermal hemorrhage or dermal deposition of hemosiderin pigment frequently causes associated pigmentation. [3] Epithelial erosion and bleeding may occur if angiokeratoma is excoriated or traumatized.[2] Histopathologic features include epidermal hyperkeratosis, acanthosis and papillomatosis with dilated vascular spaces in the papillary dermis that may be associated with organizing thrombi.[1,3] Several differential diagnoses have to be considered including lymphangioma circumscriptum, Fabry syndrome, verrucous hemangioma or even melanoma.[3,7,8] Lymphangioma circumscriptum is consisting of dilated lymphatic vessels within the papillary dermis which are filled with serosanguineous lymph fluid. If hemorrhage occurs within the spaces, it may mimic angiokeratoma microscopically. In such cases, immunohistochemistry is helpful in differentiating between blood vascular endothelium from lymphatic vascular endothelium.[9] Lipid-containing cytoplasmic vacuoles in fibroblasts, endothelial cell and pericytes are helpful in differentiation between Fabry syndrome and angiokeratoma.[7] Verrucous hemangioma is a conventional hemangioma associated with verrucous epidermal changes, which can be identical to angiokeratoma, however, hemangioma extends to involve all levels of the dermis and subcutis.[10]

Our case represents an acquired solitary angiokeratoma. The unusual location and lack of evident hyperkeratosis were challenging features. The history of bleeding raised the possibility of vascular tumor; nevertheless, the rarity of such a lesion in ophthalmology practice delayed diagnosis until histopathologic evaluation. In view of this, angiokeratoma should be added to the armamentarium of differential diagnoses of vascular eyelid tumors.

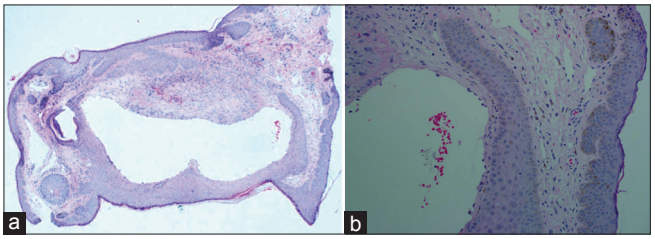

Figure 2: Histopathology study. (a) Photomicrograph showing the elongated rete ridges overlying the dilated blood vessels (H and E, ×25) (b) Higher magnification highlighting the red blood cells in the dilated blood vessels. The overlying "collaret" of the epidermis is partly engulfing the vascular channels (H and E, ×200)

REFERENCES

1. Rodríguez-Serna M, Botella-Estrada R, Chabás A, Coll MJ, Oliver V, Febrer MI, *et al*. Angiokeratoma corporis diffusum associated with beta-mannosidase deficiency. Arch Dermatol 1996;132:1219-22.
2. Gomi H, Eriyama Y, Horikawa E, Miura T. Solitary angiokeratoma. J Dermatol 1988;15:349-50.
3. Schiller PI, Itin PH. Angiokeratomas: An update. Dermatology 1996;193:275-82.
4. Offret G, Dhermy P. Angiokeratoma of the eyelids. Bull Soc Ophtalmol Fr 1966;66:30-4.
5. Haidopoulos DA, Rodolakis AJ, Elsheikh AH, Papaspirou I, Diakomanolis E. Vulvar angiokeratoma following radical hysterectomy and radiotherapy. Acta Obstet Gynecol Scand 2002;81:466-7.
6. Craig LM, Alster TS. Vascular skin lesions in children: A review of laser surgical and medical treatments. Dermatol Surg 2013;39:1137-46.
7. Kwon NH, Kim SY, Kim GM. A case of post-injury angiokeratoma circumscriptum of the chest. Ann Dermatol 2010;22:66-8.
8. Goldman L, Gibson SH, Richfield DF. Thrombotic angiokeratoma circumscriptum simulating melanoma. Arch Dermatol 1981;117:138-9.
9. Haroon S, Hasan SH. Lymphangioma circumscriptum in the scrotum: A case report. J Med Case Rep 2012;6:233.
10. Tennant LB, Mulliken JB, Perez-Atayde AR, Kozakewich HP. Verrucous hemangioma revisited. Pediatr Dermatol 2006;23:208-15.

Cite this article as: Hussein RS, Kfoury H, Al-Faky YH. Eyelid angiokeratoma. Middle East Afr J Ophthalmol 2014;21:287-8.

Source of Support: Nil, **Conflict of Interest:** None declared.