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A case of angiokeratoma in the nose: Rare entity

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Abstract

Background: Angiokeratoma is a benign cutaneous lesion of capillaries characterized by large dilated blood vessels in the superficial dermis with the hyperkeratosis. It is usually presents on the extremities and can be solitary or multiple. This lesion affects male more commonly while comparing to females.

Case Presentation: A 35 years old male presented with asymptomatic pigmented papule on his nose. Its excisional biopsy was done and sent for histopathology where it was found to be Angiokeratoma on the nose which is rare site.

Conclusion: This case highlights the unusual presentation of a rare disease at uncommon site so it needs to be differentiated from other similar lesions.

Keywords

Angiokeratoma; blood vessels; nose

Introduction

Angiokeratoma is a rare benign vascular lesion characterized by dilatation of blood vessels in the papillary dermis associated with epidermal changes in the form of epidermal hyperplasia[1]. It is an asymptomatic lesion present on the exposed parts of the body and usually treated for cosmetic reason [2]. The etiology of this disease is not known, but the common factors thought to be are congenital, pregnancy, trauma, itchy, and painful swelling on hand and foot, tissue asphyxia. These lesions are present in 70% of males and 39% of females who report with pinpoint, dark-red to blue-black, macular and papular lesions of around 4 mm, which do not blanch on pressure [3].

Case Report

This case is about an unusual site of the Angiokeratoma which is nose. A 35 years old male presented

to the dermatologist with asymptomatic pigmented papule on his nose for cosmetic reasons. The excisional biopsy was done and sent for the histopathology. Grossly, it was seen as the elliptical shaped skin covered tissue measuring 2.5 x 2 x 0.7 cm with a hyperpigmented lesion in the center measuring 0.7 x 0.5 x 0.5 cm. After serial slicing the representative sections revealed greyish brown areas and submitted in one block (Figure 1).

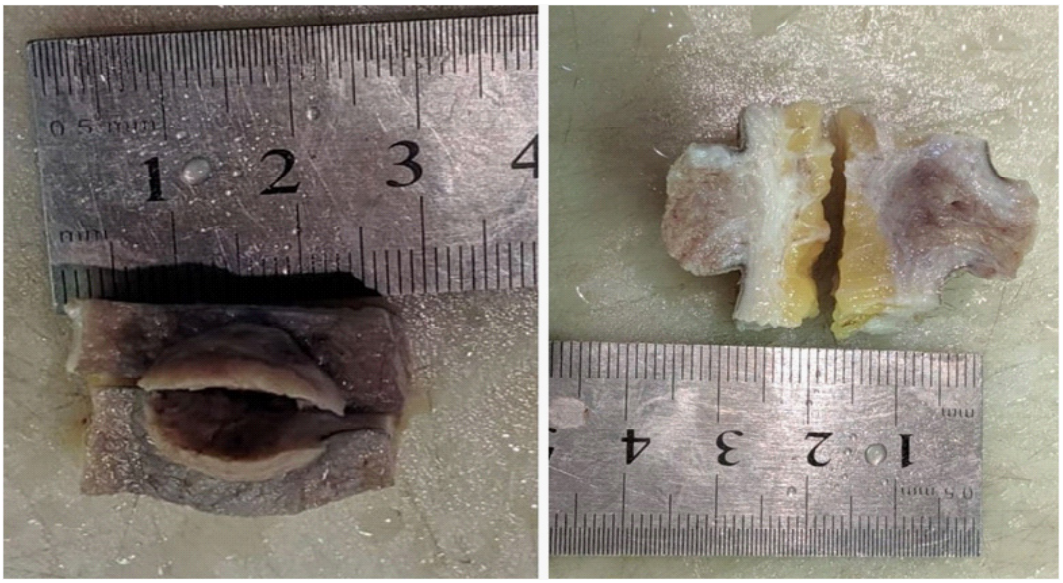

Figure 1: This figure reveals the gross appearance of lesion.

The hematoxylin and eosin stained sections from the submitted tissue showed benign ecstatic superficial dermal blood vessels mostly thrombosed, overlying irregular acanthosis with prominent thick stratum corneum and elongated rete ridges. These vascular spaces were surrounded by epidermis on all sides. Sections also revealed the presence of adnexal tissue with no significant pathology (Figure 2-3). These findings were consistent with the diagnosis of Angiokeratoma.

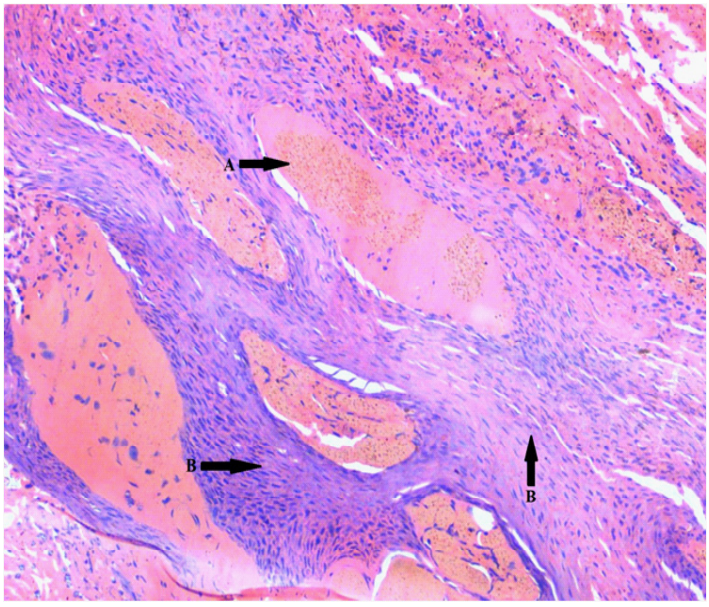

Figure 2: This figure revels (A) Benign ectatic superficial dermal vessels overlying irregular acanthosis with prominent thick stratum corneum (B). (H&E 40Xmagnification)

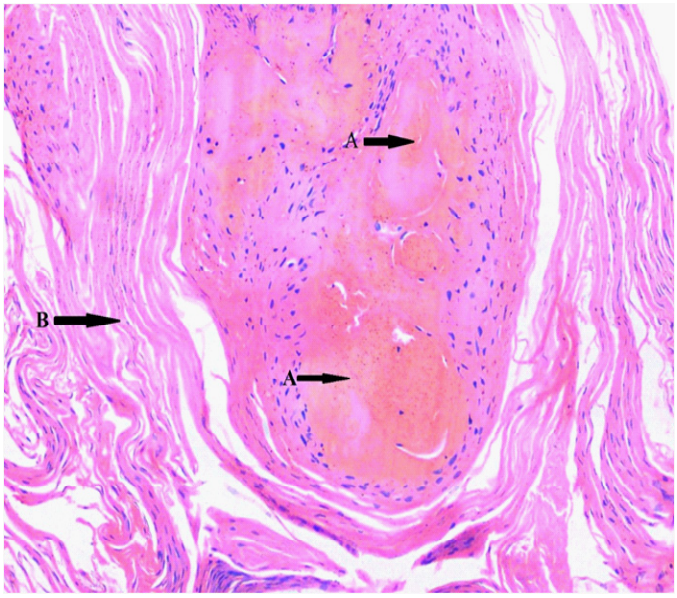

Figure 3: This figure shows (A)Ectatic blood vessels and (B) acanthosis. (H&E 100X magnification)

Discussion

The prevalence of angiokeratoma is not known accurately due to the fact that these lesions are asymptomatic. However in first and second decades it is 0.6% and rises to 16.6% in the 7th decade of life. Males show common predilection than females. Caucasian and Japanese individuals are commonly affected [4]. Clinically these are well-circumscribed, localized or generalized, single or multiple red or black papules, varying in size up to 10-mm in diameter [5]. It usually presents with the solitary or multiple lesions on the extremeties [6]. Solitary angiokeratoma occurs most frequently on the lower extremities, penis, vulva and clitoris and rarely in the head and neck region [5].

It can be classified into Fordyce's angiokeratoma, in elderly arising on the genitals, Mibelli's angiokeratoma on dorsum of toes and fingers in young patients and the congenital type angiokeratoma circumscriptum presents as unilateral large keratotic plaques. In the generalized form, angiokeratoma corporis diffusum, lesions are usually concentrated between the umbilicus and knees which is observed in both persons with various enzyme deficiency disorders as well as healthy individuals [7]. The systemic form is known as angiokeratoma corporis diffusum and is usually linked to some metabolic disorders, mainly Fabry's disease and fucosidosis [8]. As far as aetiopathogenesis of angiokeratoma is concerned, it is uncertain. It however starts with vascular ectasia within the papillary dermis. The epidermal hyperplasia is a reactive change. On electron microscopy angiokeratoma is composed of vascular channels lined by very thin endothelial wall. Their cytoplasm shows microfilaments, pinocytosis vesicles and vacuoles with scarcely electron-opaque content. There are scanty cytoplasmic organelles, such as mitochondria and endoplasmic reticulum [9].

It is important to diagnose angiokeratoma because of its similarity to some other lesions [10]. Thereby other vascular lesions like hemangioma, and lymphangioma should be excluded on help of histopathology. Small capillaries lined by single layer of endothelial cells supported by connective tissue stroma are seen in a hemangioma. While lymphangioma, are characterized by several intertwining lymph vessels present very near to the epithelium and also in papillary connective tissue. And in case of angiokeratoma, blood vessels lie very close to the epithelium [11]. Also the presence of blood-filled spaces lined by endothelium helps to differentiate angiokeratoma from lymphangiomas [11].

Clinically, Angiokeratoma needs to be differentiated from aggressive lesions like malignant melanoma. In malignant melanoma atypical melanocytes arranged in clusters or groups or singly are seen often with prominent nucleoli. This morphology is not seen in case of angiokeratoma [12]. Malignant transformation has been albeit never reported but common surgical excisions show recurrences. Hence among various treatment choices excision, electrodessication, cryotherapy, or laser ablations are the mostly preferred [1].

This case report draws the attention to the fact that although angiokeratoma is very rare in the head and neck area, nose is the least thought site. So any vascular lesion under consideration must rule out the diagnosis of angiokeratoma.

Conclusion

Angiokeratoma in the nose is the least common site even for the least common and rare disease itself yet any vascular lesion in the nose have to be thoroughly studied to rule out angiokeratoma.

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