




Destructive (Erosive) Tufted Angioma/Kaposiform Hemangioendothelioma of the Eyelid

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Abstract

BACKGROUND: Tufted angioma/kaposiform hemangioendothelioma represents a spectrum of rare benign skin vascular neoplasms that differ in the extent and prognosis. Tufted angioma affects primarily children, is localized to skin, shows no tendency for ulceration or tissue invasion and has a favorable prognosis.

CASE REPORT: In this case report, tufted angioma involved the lower eyelid resulting in its destruction and compromised its function, hence necessitating rapid surgical excision to preserve the function of eyelid.

CONCLUSION: TA/KHE is a rare vascular benign neoplasm usually asymptomatic but may affect delicate structures, such as the eyelid resulting in destruction and hence may require rapid surgical intervention. corresponding eye.

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Introduction

Tufted angioma (TA), also known as angioblastoma/angioblastoma of Nakagawa, is a rare benign vascular tumor frequently developing in infancy and early childhood. The condition is very slowly progressive or stationary and occasionally self-remitting. During the whole course of TA, the lesion remains asymptomatic, the overlying epidermis remains intact and the prognosis is favorable [1]. Because of its considerable clinical, pathological, and immunohistochemical overlap with kaposiform hemangioendothelioma (KHE), both tumors are considered to be a spectrum of the same disease [2]. The concept of TA/KHE spectrum and therefore of a single entity is also adopted by the authors of this rarely destructive TA/KHE entity.

Case Report

A 12-year-old male developed several small multiple red lesions on the left inner canthus and lower

eyelid that started 12 months before presentation and were gradually and slowly increasing in size and number. There were no history of trauma, previous facial or eye surgery, or similar lesion elsewhere on the body. The patient received topical combination of antibiotics and steroids for several weeks without improvement. The condition remained asymptomatic and the child was otherwise healthy. Examination revealed multiple red papules and small non-compressible nodules that partially eroded the left lower eyelid margin and compromised the closure of the eye so that the medial canthus (commissure), lacrimal caruncle, eroded inferior lacrimal punctum, and sclera were clearly visible even with tight closure of the eyelid (Figure 1). There was no affection of the visual acuity and regional lymph nodes were not palpable. Likewise, general examination revealed no abnormality and complete blood count was within normal range. Histopathology revealed cannonball pattern in the form of small circumscribed angiomatous tufts and lobules scattered in the dermis; hence, the diagnosis of TA or TA/KHE was made (Figure 2a and b). Due to the evident eyelid tissue erosion and destruction, surgical excision under general anesthesia was performed and no evidence of recurrence was observed during a 1-year follow-up period.



Figure 1: Multiple erythematous papules and nodules in the left canthus and lower eyelid with partial erosion of its margin resulting in incomplete closure of the left eye with destruction of the medial margin and inferior lacrimal punctum and exposure of the underlying lacrimal caruncle and sclera

Comment

TA/KHE can be either congenital or acquired of unknown etiology, though trauma, high hormonal levels during pregnancy or puberty, as in the present case, and underlying vascular malformation such as port-wine stains has infrequently been reported. The tumor appears as ill defined, dull red macules with a mottled appearance varying from 1 to 10 cm in size. Some show clusters of smaller angiomatic papules superimposed on the main macular areas or as an exophytic, firm, red or violaceous, cutaneous solitary or multiple papules, and nodules as in the present case. The lesion progresses slowly over months to years and the size of the lesion can be variable, though rarely it may undergo spontaneous regression [1], [2].

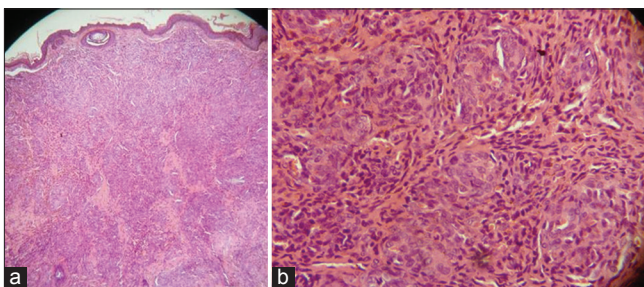


Figure 2: Histopathologic features showing (a) the characteristic vascular tufts "cannonball" dispersed in the superficial and deep dermis. (H&E, original magnification $\times 100$). (b) Multiple and narrow capillary spaces particularly at the periphery of the tufts in a glomeruloid arrangement (H&E, original magnification $\times 400$)

Congenital TA/KHE should be expected when a poorly defined congenital infiltrating vascular tumor with (out) overlying hirsutism appears over the mandibular area, where about three-fourth of all facial congenital TA/KHE develop [3].

Although very few cases involving the eyelids were reported, none was associated with tissue destruction, ulceration, or erosion [4], [5]. The pathogenesis of the destruction of the eyelid margin in

the present case is not known, though it may be due to a compromised blood supply by the newly formed blood vessels and the nature of vasculature of the eyelid margin rather than tumor related as it has been observed only in eyelid margin lesions. Other serious complication includes Kasabach–Merritt phenomenon (KMP) which may develop in about 10% of congenital cases of TA and up to 70% in KHE and is manifested by widespread ecchymoses and petechiae. KPM is characterized by a severe thrombocytopenia with mild to moderate coagulopathy resulting from intralesional platelet trapping. Occasionally, it may be associated with tenderness, hyperhidrosis, and hypertrichosis [1], [6].

No specific laboratory examinations are required for the diagnosis or treatment of TA/KHE. Complete blood cell count and full disseminated intravascular coagulation profile are indicated in the presence of manifestations suggestive of KMP [7]. TA has a specific histologic pattern in the form of lobular arrangement of densely cellular capillaries "cannonball pattern" throughout the dermis. The vascular tufts consist of tightly packed hypertrophic endothelial cells with scanty cytoplasm and nuclei that are round, ovoid, or fusiform. Mitoses are infrequent and hemosiderin deposits may be observed within the endothelial cells. Capillary spaces are narrow and elongated, and more noticeable in the periphery of the lobules, where they have a characteristic half-moon shape. These findings were evident in the present case, hence favoring the TA of the spectrum (Figure 2a and b) [2], [7], [8].

Positive staining for vascular markers CD 31, CD 34, alpha-smooth muscle actin (rare), and the lymphatic marker D2-40 and PROX1 and negative staining of GLUT-1 denote both the existence of two cellular components in TA/KHE and the immaturity of endothelial cells [8]. Immunohistochemistry is confusing and inconclusive in differentiating TA from the more locally aggressive KHE and, in practice, it is usually deferred as in this case [2].

Differential diagnosis of the TA/KHE includes pyogenic granuloma, benign neoplasms of endothelial origin, and Kaposi sarcoma. However, the histopathological and immunohistochemical findings are specific and these lesions exhibit individual characteristics by which they can be distinguished from TA/KHE [2], [9].

The prognosis is favorable; hence, observation is the best course of action. Cryosurgery, radiation therapy, electrocoagulation, and pulsed dye laser are successful to varying degrees, whereas local injection of interferon-alpha is a non-surgical option in growing lesions [10]. For patients who develop KMP, the first-line therapy consists of vincristine plus prednisone or sirolimus (rapamycin). Aspirin may also help to control the platelet interaction, pain, and growth of TA/KHE [11], [12]. Topical timolol maleate is an option for limited lesions [13]. Surgical excision allows permanent removal of the TA and

was preferred in the present case to preserve the function and structure of the partially damaged eyelid and hence the eye.

Conclusion

TA/KHE is a rare vascular benign neoplasm usually asymptomatic but may affect delicate structures, such as the eyelid resulting in destruction and hence may require rapid surgical intervention.

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