Case Report

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Pathology Section

Tufted Angioma of Eyelid in an Adult - A Case Report

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ABSTRACT

Tufted Angiomas also known as angioblastomas /Angioblastoma of Nagakawa are rare vascular neoplasms localised to the skin and subcutaneous tissues with the upper trunk and neck being most common sites. They are mainly seen in children but a few cases in juveniles and adults have been reported. We hereby report this case, a 40-year-old male who presented with a right lower lid, painless, slowly progressive, firm swelling diagnosed as Tufted Angioma on histopathology and immunohistochemistry.

Keywords: Angioblastoma, Immunohistochemistry, Vascular neoplasm

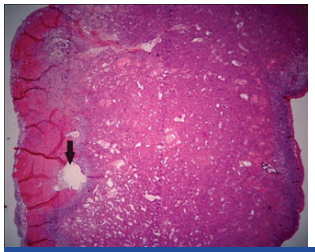
CASE REPORT

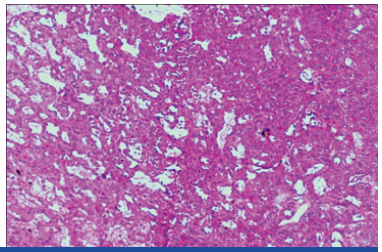
A 40-year-old male presented with a right lower lid painless, gradually progressive swelling for the past 2 years. There was no preceding history of trauma, cutaneous lesion, infection or any drug intake. No physical or systemic abnormality was noted. Patient had normal haemogram, coagulation profile and immune status. Lesion was excised and submitted for histopathological examination.

Biopsy specimen received was a single, irregular, firm, reddish brown piece of tissue measuring 0.5x0.5cm in size and was processed as such.

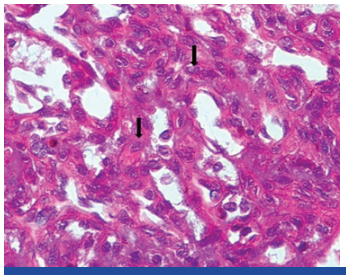
The haematoxylin and eosin – stained sections [Table/Fig-1-3] were examined and showed fragments of stromal tissue of eyelid with relatively circumscribed ovoid foci of closely packed capillaries scattered throughout the stroma with surrounding pericytic proliferation. Capillaries were bloodless and were lined by plump endothelial cells. Dilated lymphatic like vessels were seen in close approximation to capillary aggregates. No well-defined epithelial lining was seen. No significant mitotic figures were identified.

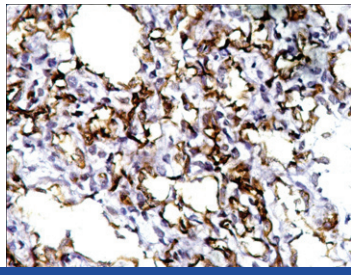
On immunohistochemistry, the endothelial cells lining the capillaries were positive for CD 34 [Table/Fig-4] and the pericytic cells showed diffuse strong positivity for SMA [Table/Fig-5].





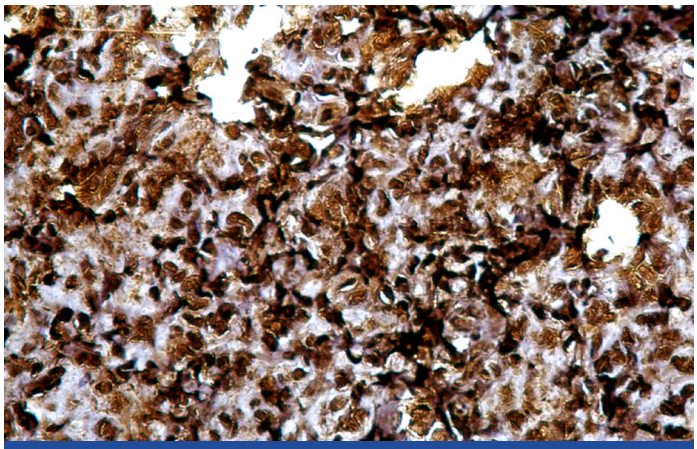
[Table/Fig-1]: Lobular aggregates of capillaries with peripheral dilated lymphatics (arrow) in Tufted Angioma. (H&E x4). **[Table/Fig-2]:** Closely packed proliferating capillaries with Pericytic proliferation. (H&E x200).





[Table/Fig-3]: Pericyte (arrow) proliferation surrounding capillaries lined by plump endothelial cells. (H&E x400). **[Table/Fig-4]:** Endothelial cells lining capillaries showing CD34 positivity. (IHC x400).

Hence the final diagnosis of tufted angioma was established on the basis of clinical presentation, histopathological examination and immunohistochemistry. Post biopsy performance of the patient could not be assessed as he didn't turn up.



[Table/Fig-5]: Pericytic cells showing diffuse strong positivity for SMA. (IHC x400).

DISCUSSION

Tufted Angioma (TA) is a rare, slowly progressive benign vascular tumour with variable clinical presentation. Most cases (60-70%) of TA develop before the age of five years and fewer than 10% of cases with TA occur after the age of 50 years. There is no sex predilection [1]. TA usually presents as a solitary nodule, papule or plaque predominantly over neck, upper thorax and shoulders [2] with face, scalp and proximal extremities being rare sites of involvement. TA may even be multifocal. The lesion progresses slowly over months to years and size of the lesion can be variable [3]. TA is usually asymptomatic but may be found associated with tenderness, hyperhidrosis and hypertrichosis [1,4,5]. Occasionally, TA may be associated with the Kasabach-Merritt syndrome (KMS) [2]. If patient presents with ecchymotic patches or petechiae, clinician should be alert as it may have developed into KMS.

TA are discrete, round to ovoid shaped, vascular tufts of densely packed but poorly canalized capillaries. These capillaries are usually lined by round to oval and sometimes spindled, plump endothelial cells which are randomly dispersed in the dermis in a "cannonball distribution" [6]. Occasionally they may be seen extending into the subcutaneous tissue. Predominant component of TA is formed by the pericytes which surround these capillaries. These pericytic cells are bland looking, have oval to slightly elongated nuclei with scant cytoplasm and indistinct cell boundaries. On IHC, the endothelial

cells are reactive for numerous markers including CD31, CD34 and von Willebrand factor (factor VIII) [7]. These angiomatous lobules are surrounded by dilated, crescent shaped lymphatic spaces. The epidermis and cutaneous appendageal structures of the dermis are usually uninvolved. Oedema and inflammation is absent.

TA needs to be differentiated from various other non-neoplastic as well as neoplastic vascular lesions like capillary haemangioma, Kaposiform haemangioendothelioma, Kaposi's sarcoma and low grade angiosarcoma depending on the age [1,6,8-10] [Table/Fig-6,7].

Till date, only two cases of TA in eyelid in adults have been reported [7]. Our case was similar to them on all basis except for Ki-67 which showed low proliferation index, and which was not done in our study [7].

Cases with clinical suspicion of TA should undergo a complete haemogram (including platelet count), coagulation profile and full screening for disseminated intravascular coagulation to exclude consumptive coagulopathy and Kasabach- Merritt Syndrome (KMS). Currently, the risk of association of KMS in patients presenting with tufted angioma of eyelid is not very well known. Some studies have reported spontaneous regression of TA, albeit rarely [11,12]. Magnetic Resonance Imaging (MRI) studies have proven useful in the evaluation of the depth of invasion and extent of growth of TA [2].

Features	Tufted Angioma	Kaposiform Haemangioen Dothelioma	Capillary Haemangioma
Microscopy	Cannon ball nests, lobular aggregates of spindle and polygonal cells with interspersed endothelial cell lining capillaries. Pericyte rich epithelioid nodules(-)	Cannon ball nests with glomeruloid structures (+) with prominent lymphatic network surrounding tumour lobules. Pericyte rich epithelioid nodules (+)	Cannon ball nests and glomeruloid structures (-)
IHC	(+)	Well formed capillaries (+) Neoplastic spindle cells focally (+) Slit like vessels (vWF -ve)	(+)
CD34, CD31, vWF			
SMA	Pericytes diffusely (+)	Pericytes focally (+)	NA
D2 40	Cannon ball like capillaries (-) Surrounding Dilated vessels (+)	Peripheral KS like proliferating capillaries (+) Surrounding dilated vessels(-)	NA
GLUT 1	NA	(-)	(+)

[Table/Fig-6]: D/D of tufted angioma in children.

Features	Tufted Angioma	Kaposi Sarcoma	Low Grade Angiosarcoma
Endothelial cells	Appear plump to slightly spindled	Spindling is prominent	Cells are atypical and mildly pleomorphic
Vascular Spaces	Bloodless	Blood filled	Sinusoid like spaces
Nuclear Atypia	Not seen	Present in nodular stage	Mild to moderate
Collagen	Absent	NA	Dissected collagen
Mitosis, necrosis, haemorrhage	Absent	May be present	Present
IHC	CD 34,CD31+ SMA+(pericytes)	Factor VIII positive HHV 8 positive	CD 34,CD31, factor VIII+

[Table/Fig-7]: D/D of tufted angioma in adults.

CONCLUSION

This case of Tufted angioma gains special consideration due to its unusual site of involvement and age of presentation. Based on this fact, TA needs differentiation from various other vascular lesions of eyelid, especially in adults. Also, it is essential to get a detailed physical and haematological work up done in all cases diagnosed with eyelid TA, to exclude rare association of Portwine stain and Kasabach-Merritt Syndrome.

ABBREVIATIONS

TA: Tufted Angioma; KMS: Kasabach-Merritt syndrome; D/D: Differential Diagnosis; KS: Kaposi's sarcoma.

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