Tufted angioma (Angioblastoma) of eyelid in adults—report of two cases

Ruchi Mittal1* and Devjyoti Tripathy2

Abstract
Tufted angioma, first recognized in Japanese literature as “Angioblastoma of Nagakawa”, is a rare benign vascular tumour with a variable clinical presentation. It commonly manifests as a macule, papule or nodule in infancy or childhood in the region of the upper trunk and neck. Here in we report two cases of this rare progressive angioma as lesions of the eyelid in adults. Tufted angioma has a classical “cannon ball” like appearance of vascular tufts on histopathology. Immunohistochemical staining with actin highlights the spindly stromal cells surrounding the capillaries. Complete physical examination and haematological work up is recommended in patients with tufted angioma to exclude rare association of port wine stain and Kasabach-Merritt syndrome with this rare entity. To the best of our knowledge, our cases illustrate the first case report of tufted angioma presenting as an eyelid lesion.

Virtual Slides: The virtual slide(s) for this article can be found here: http://www.diagnosticpathology.diagnomx.eu/vs/1230909536950947.

Keywords: Tufted angioma, Angioblastoma, Eyelid, Adults, Kasabach-Merritt syndrome

Background
Tufted angioma, first recognized in Japanese literature as “Angioblastoma of Nagakawa”, is a benign progressive angioma, with a variable clinical presentation [1]. Wilson-Jones and Orkin [2] coined the term “tufted angioma” for this unusual pattern of angiomatous proliferation which was found to have some morphological similarities with strawberry nevi. However, as the name suggests, the lesion was seen as cannon ball like, small circumscribed angiomatous tufts and nodules in the dermis and subcutaneous tissue with characteristic lymphangioma-like vessels [3]. Tufted angioma (TA) can be congenital or acquired, commonly presents in infancy or early childhood, can be present at birth in approximately 25% of cases [4], and few cases of TA have been reported in adults [5]. It commonly presents as a macule, papule or plaque over the upper trunk, neck and proximal part of the limbs [6], however involvement of other locations like face, oral mucosa and lip [5] is also known. A pubmed search of dermatological and ophthalmic literature using key words “Tufted angioma, Angioblastoma, eyelid, adults, Kasabach-Merritt syndrome” did not reveal any case of tufted angioma presenting as an eyelid lesion. To the best of our knowledge, involvement of the eyelid with TA is previously unreported. Herein, we report 2 cases of TA of eyelid in adult patients which were clinically diagnosed as lymphangioma and epidermal cyst. This is a retrospective study, approved by Institutional review board of L.V. Prasad Eye Institute as a retrospective study.

Case presentation
Case 1
A 17-year-old male presented with a history of a gradually progressive, painless swelling of right lower eyelid of 6 years’ duration. There was no history of trauma, previous ocular surgery or presence of similar lesion elsewhere in the body. Clinical examination revealed a right lower eyelid soft tissue mass not fixed to the underlying tarsus. It was non-tender and spongy to firm on palpation. On eversion of the eyelid the lower palpebral conjunctiva was unremarkable. Rest of the ocular examination was normal. A vascular lesion, probably a lymphangioma, was suspected. Systemic examination and complete blood picture was normal and a biopsy was planned. At biopsy under local anesthesia, the mass
appeared to be reddish and ill-defined with intermingling soft and firm areas. Piecemeal excision of the lesion was done and the excised tissue was submitted for histological examination.

Case 2
A 44-year-old female presented with a slowly progressive painless swelling of the right upper eyelid of about 3 years’ duration. There was no other significant ocular or systemic history. Clinical examination revealed a mobile, non-tender, and cystic to firm nodular mass, not fixed to the tarsus. Systemic evaluation and complete blood picture was normal. Clinically, the lesion was thought to be an epidermal cyst. At biopsy under local anesthesia, a nodular, well delineated, non-encapsulated, firm mass about 15 mm in its maximum dimension was excised in Toto and submitted for histological evaluation.

Histopathology sections from case 1 showed multiple fragments of stromal tissue of eyelid with adnexal structures, striated muscle bundles of orbicularis oculi (Figure 1) and relatively circumscribed ovoid foci of closely set capillaries scattered throughout the stroma (Figures 1 and 2). Capillaries were predominantly bloodless, lined by plump endothelial cells and surrounded by prominent oval to slightly spindly cells. Dilated lymphatic-like vessels, some of which were crescent shaped, were seen in close approximation to, or surrounding the capillary aggregates. Cellular atypia or mitoses were absent in multiple serial sections studied.

Histopathology sections from case 2 showed a single fragment of stromal tissue of eyelid with striated muscle bundles of orbicularis oculi and closely set relatively circumscribed ovoid to round angiomatous aggregates. These angiomatous aggregates were composed of bloodless capillaries with slit like or mildly dilated lumina, lined by plump bland endothelial cells (Figure 3). These capillaries were surrounded by short spindly to oval bland cells (Figure 3). Dilated lymphatic like vascular...
spaces were seen in close approximation to the angioma-
tous tufts. No mitotic figure was identified in the sec-
tions studied.

Immunohistochemical staining in both cases showed
similar findings. Staining with actin revealed a promi-
ient pericytic component (Figure 4). CD34 stained the
endothelial cells (Figure 5). Ki-67 showed a very low
proliferation index. A diagnosis of tufted angioma of the
eyelid was made in both the cases based on the above
histopathological and immunohistochemical findings.

Complete physical examination by a dermatologist was
normal. Both cases have not had a recurrence in over
12 months of follow up.

Discussion
Tufted angioma is a rare, slowly progressive benign
vascular tumor which can have variable clinical mor-
phology. It can present as red or bluish to violaceous
papule, plaque or nodule over neck, upper back and
proximal part of limbs typically in childhood and in-
fancy. These lesions may also develop in adults or the
elderly [7]. There is no sex predilection [8]. The lesions
range from few millimeters to few centimeters in size
but may be extensive covering larger areas and can be
multifocal [9]. Most of the lesions are asymptomatic but
may present with tenderness, hypertrichosis and hyper-
hidrosis [8,10,11]. Tenderness, hypertrichosis, and indu-
rating can be useful in differentiating TA from common
hemangioma [12]. Microscopically TA has a classical
morphology. It is found dispersed in the dermis and oc-
casionally extends into the subcutaneous tissue and is
seen as discrete round to ovoid angiomatous aggregates
[8]. These aggregates are composed of relatively bloodless,
poorly canalized capillaries. Capillaries are lined by plump
endothelial cells, which may show slight spindling. The
endothelial cells show reactivity for several markers, in-
cluding CD31, CD34 and von Willebrand factor (factor
VIII). In our case, CD34 was used to highlight the
endothelial cells. Pericytes are seen surrounding the ca-
pillaries and are the predominant cellular component of
TA. These cells have indistinct cell boundaries, scant
cytoplasm, and oval to slightly elongated nuclei with
bland morphology. Dilated crescent shaped lymphatic
like vascular channels are seen surrounding the angio-
matous lobules. These channels are lined by plump to
flattened cells with oval to slightly spindly nuclei.

Tufted angioma in childhood needs differentiation
from strawberry nevi and kaposiform hemangioendo-
thelioma [4]. Angiomatous aggregates of strawberry nevi
are more massive and replace wider planes. Eyelid
lesions tend to have involvement of deeper orbital struc-
tures. Kaposiform hemangioendothelioma is mor-
phologically intermediate between strawberry nevi and
Kaposi sarcoma [8], is more commonly seen in child-
hood and shows capillaries with lobular pattern that
are locally infiltrative. Periphery of the tumor shows mi-
cro thrombi within the capillaries. Angioblastoma of
Nagakawa (TA) should not be confused with the term
giant cell Angioblastoma (GCA), which is a rare hem-
angiomia-like lesion of infancy [13]. GCA is charac-
terized by nodular, linear and plexiform granuloma–like
aggregates of histiocytes–like cells and giant cells, sur-
rounding capillary sized vessels lined by plump endo-
thelial cells. Background is loose, mesenchymal with
mononuclear inflammatory cell infiltrate and mast cells
[13,14]. In adults, TA needs to be differentiated from
Kaposi’s sarcoma and low grade angiosarcoma [2,7].
Endothelial cells in TA appear plump to slightly spindled
and lack the spindling of Kaposi’s sarcoma [8]. Vascular spaces of TA are relatively bloodless when compared to blood filled spaces in Kaposi’s sarcoma. Lack of nuclear atypia excludes angiosarcoma [7]. Other diagnostic hints of angiosarcoma include presence of sinusoid like spaces, dissection of collagen, mitoses, necrosis and haemorrhage [8,15]. Cases with clinical suspicion of TA should be worked up with complete physical examination and complete blood count with red cell morphology to exclude consumptive coagulopathy and Kasabach–Merritt syndrome (KMS) [16,17]. Prothrombin time and activated partial thromboplastin time should be performed in patients with thrombocytopenia. Rare association of Port wine stain with TA is also reported [5,17]. Spontaneous regression is known, but is exceptionally rare and regression may occur over a period varying from months to years [18,19].

Conclusions
As evident by this paper, TA although not described in the eyelid before, should be considered as a differential diagnosis of benign eyelid vascular lesions. TA can have variable clinical presentation and can present at any age. They can be ill defined to circumscribed, and should morphologically be distinguished from other commoner benign and malignant vascular eyelid lesions. A complete physical examination and hematological work up has been recommended in patients with classical TA to exclude rare association of Port wine stain and KMS. Whether a significant risk of the KMS exists in patients of eyelid TA is not currently known. Considering the smaller size of eyelid lesions this risk is less likely. Nonetheless it may be prudent to get a detailed physical and hematological work up done in all cases diagnosed with eyelid TA.

Consent
Written informed consent was obtained from the patient for publication of this Case Report and accompanying images. A copy of the written consent is available for review by the Editor-in-chief of this journal.

Abbreviations
TA: Tufted angioma; KMS: Kasabach–Merritt syndrome.

Competing interests
The authors declare that they have no competing interests.

Authors’ contributions
RM carried out the histological and immunohistochemical work up, compiled the cases and drafted the manuscript. DT carried out the clinical work up, provided the clinical information, participated in designing the manuscript and edited the manuscript. Both the authors read and approved the final manuscript.

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Author details
1. Dalmia Ophthalmic Pathology Services, L.V. Prasad Eye Institute, Bhubaneswar, Orissa, India. 2. Department of Ophthalmic Plastic Surgery, Orbit and Ocular Oncology, L.V. Prasad Eye Institute, Bhubaneswar, Orissa, India.

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References
1. Alexi E, Bertani E, Sala F: Acquired tufted angioma. Am J Dermatopathol 1986, 8(5):426–429.
2. Jones EW, Orkin M: Tufted angioma (angioblastoma): a benign progressive angioma, not to be confused with Kaposi’s sarcoma or low-grade angiosarcoma. Am J Acad Dermatol 1989, 20:214–225.
3. Sadeghpour M, Antaya RJ, Lazova R, Ko CJ: Dilated lymphatic vessels in tufted angioma: a potential source of diagnostic confusion. Am J Dermatopathol 2012, 34(4):400–403.
4. Alberola LT, Bertiuch I, Montero LC, Nentes IJ, Martinez NL, Paz AA: Congenital tufted angioma: a report and review of the literature. Dermatol Online J 2010, 15(6):2.
5. Lee B, Chiu M, Soriano T, Craft N: Adult-onset tufted angioma: a case report and review of the literature. Cutis 2006, 78(5):341–345.
6. Tsang WW, Chan JK, Fletcher CD: Recently characterized vascular tumours of skin and soft tissues. Histopathology 1991, 19(6):489–501.
7. Hebeda C, Scheffer E, Stannik TM: Tufted angioma of late onset. Histopathology 1993, 23(2):191–193.
8. Eduardo C: Vascular tumours: tumors and tumor-like conditions of blood vessels and lymphatics. In Histopathology of skin. 10th edition. Edited by Elder DE, New Delhi: Wolters Kluwer; 2009:1007–1056.
9. Ghosh SK, Bandypadhyay D, Ghosh A, Biswas SK, Barma KD: Acquired multifocal tufted angiomas in an immunocompetent young adult. Indian J Dermatol 2011, 56(4):412–414.
10. Schaffer JV, Fangman W, Bossenbroek NM, Meehan SA, Camino H: Tufted angioma. Dermatol Online J 2008, 14(10):20.
11. Cho KH, Kim SH, Park KC, Lee AY, Song KY, Chi JG, Lee YS, Kim KJ: Congenital angiolipoblastoma (Nakagawa)–is it the same as tufted angioma? Clin Exp Dermatol 1991, 16(2):110–113.
12. Wong SN, Tay YK: Tufted angioma: a report of five cases. Pediatr Dermatol 2002, 19:388–393.
13. Goh SG, Calonje E: Cutaneous vascular tumours: an update. Histopathology 2008, 52(6):661–673.
14. Mao R, Jiang ZM, Zhang HZ, Zhu XK, Zhang QL: Clinical and pathological characteristics of giant cell angiolipoblastoma: a case report. Diagn Pathol 2012, 7:13.
15. Lin XY, Liu Y, Zhang Y, Yu JH, Wang EH: The co-expression of cytokeratin and p63 in epithelioid angiosarcoma of the parotid gland: a diagnostic pitfall. Diagn Pathol 2012, 7:118.
16. Kim T, Roh MR, Cho S, Chung KY: Kasabach-Merritt syndrome arising from tufted angioma successfully treated with systemic corticosteroid. Ann Dermatol 2010, 22(4):436–430.
17. Osio A, Fratagl S, Hadi-Rabia S, Bodemer C, De Prost Y, Hamel-Teillac D: Clinical spectrum of tufted angiomas in childhood: a report of 13 cases and a review of the literature. Arch Dermatol 2010, 146(7):758–763.
18. Browning J, Frieden I, Basilegla E, Wagner A, Metry D: Congenital, self-regressing tufted angioma. Arch Dermatol 2006, 142:749–751.
19. Ishikawa K, Hatanou Y, Ichikawa H, Hashimoto H, Fujiwara S: The spontaneous regression of tufted angioma: a case of regression after two recurrences and a review of 27 cases reported in the literature. Dermatol 2005, 210(4):346–348.

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