Acquired Tufted Angioma of the Lower Lip Mucosa

(Angiome houppé acquis de la muqueuse de la lèvre inférieure)

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Sommaire

L'angiome houppé acquis est une agglomération vasculaire isolée de couleur rouge foncé qui touche généralement la peau, apparaît chez les enfants ou les jeunes adultes et affiche des caractéristiques microscopiques particulières. Cliniquement, cette lésion s'étend à une vitesse variable, se stabilise et peut régresser spontanément. Cet article porte sur une petite lésion vasculaire ancienne de la muqueuse de la lèvre inférieure qui affiche les caractéristiques microscopiques et immunohistochimiques de l'angiome houppé acquis. Le diagnostic différentiel (granulome pyogénique, hémangiome capillaire et hémangiopéricytome) fait l'objet d'une discussion.

Mots clés MeSH : *case report; hemangioma/pathology; skin neoplasms/pathology*

Cet article a fait l'objet d'une révision par des pairs.

In 1976, Jones described an unusual vascular proliferation characterized by progressive clinical enlargement of dusky red plaques of the skin.[1] Subsequently, he and Orkin[2] described 20 cases, while other authors described individuals[3-10] or small groups[11-14] of patients with this rare condition. In the Japanese literature, the lesion has been reported under the designation "angioblastoma" and was first described by Nakagawa in 1949.[12] It was first described in the English literature by Macmillan and Champion[15] in 1971 under the term "progressive capillary haemangioma".

Although a few cases are congenital,[2,6,10] most develop in childhood.[2,8,9,11,12,14] Occasionally, older individuals are affected.[2,4,5,7,13] There is no significant gender predilection. Skin at any site can be affected, but there is a predilection for the neck, shoulder and thorax region.[2,4-7,10-13] Lesions have been reported as macules or plaques, which progressively enlarge at a variable rate and then become more or less stable.[2,6,8] Occasionally, the lesions start[11] or persist[13] as small violaceous papules. Cases have been reported to occur during pregnancy[4] with spontaneous resolution after childbirth, and one case, which ultimately regressed spontaneously, developed after a liver transplant.[5] Miyamoto and others[7] reported a case that exhibited partial spontaneous regression. Since these lesions usually become stable, no treatment is recommended. Steroid treatment was reported to be successful by Munn and others[9] in a 3-month-old girl but to have no effect in an 11-year-old boy, as reported by Padilla and others.[11] Other forms of treatment that have been attempted include surgical excision,[2,7,11,12] cryotherapy[2] and radiation therapy.[2] Recurrence has been reported.[2,3]

Microscopically, the lesions are composed of multifocal, tightly packed knots or tufts of spindle and polygonal cells associated with endothelial cells. The tufts may form capillaries. The lesions appear to arise directly from the normal vasculature of the dermis.[2] Occasionally, the tufts occur close together and coalesce to form larger bundles, but the multifocal lesions are characteristically discontinuous microscopically in individual tissue sections. Mitoses are rare, and there is no significant associated inflammation. The dermal collagen appears normal. Typically, there is a zone of collagen separating the nodule from the overlying epithelium. Many cells are surrounded by a reticulin sheath. Ultrastructural studies confirm the presence of endothelial cells, including the presence of Weibel-Palade bodies,[7,11,12] but the surrounding spindle and polygonal cells may show few cytofilaments[12] or focal condensations of microfilaments.[11]

Immunohistochemical studies have shown consistently strong positive staining of endothelial cells with *Ulex europaeus* agglutinin 1 (UEA1),[2,5,7,11] but less consistent staining for factor VIII-related antigen (VIII-RA).[2,5,7] Based on positive staining for actin in the surrounding spindle and polygonal cells, Chu and LeBoit[5] and Padilla and others[11] interpreted them to be pericytes.

Case Report

A 20-year-old woman presented with a reddish-brown papule about 1 mm in diameter on the left lower lip mucosa near the vermilion at the commissure. The lesion had been present for at least 10 years without change in size. There was no pulsation, bleeding or pain. The patient could not recall her age at the time of onset, but she was aware of a congenital

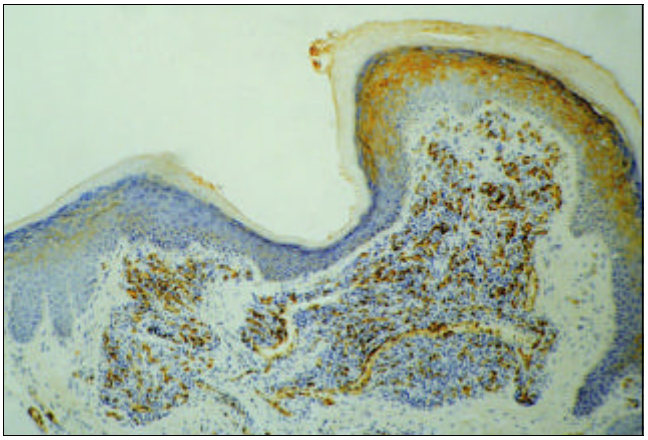

Figure 1: *The acquired tufted angioma consists of separate knots of cells, some of which form capillary lumina, arising from normal subepithelial vessels, which traverse the lesion (hematoxylin, UEA1 immunostaining, magnification x100).*

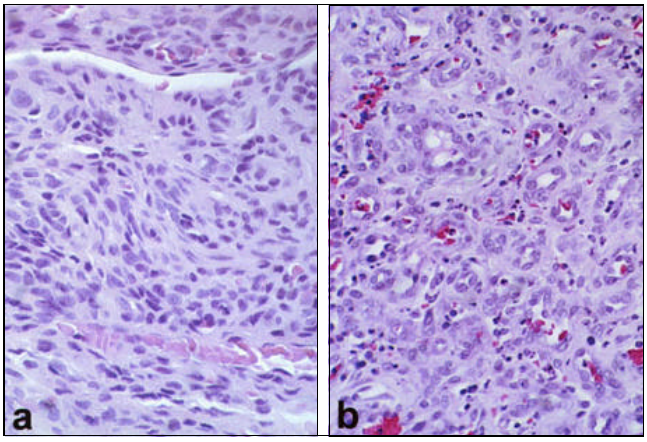

Figure 2: *The acquired tufted angioma is dominated by spindle and polygonal cells, interpreted to be pericytes, which surround fine, endothelium-lined spaces (a). The pyogenic granuloma in the proliferative stage exhibits a proliferation of endothelial cells, most of which line capillaries, often forming back-to-back channels and without significant pericyte proliferation (b) (hematoxylin-eosin, magnification x250).*

port-wine stain on the skin of her back. No other vascular lesions were clinically visible in the head and neck region. An excisional biopsy was performed, and routine hematoxylin-eosin stained tissue sections were examined. Subsequently, additional tissue sections were stained for reticulin, and immunohistochemical staining by UEA1 and for VIII-RA, S-100 and actin were obtained using standard techniques.

Formalin-fixed, paraffin-embedded tissue sections stained with hematoxylin-eosin exhibited two separated, tight knots of spindle and polygonal cells clustered around mature blood vessels lined by thin endothelial cells. The nuclei of the lesional cells were polygonal to spindle-shaped with variable, diffuse chromasia. Cytoplasmic borders were indistinct, but the crowding of the nuclei indicates that there was little cytoplasm (Figs. 1 and 2). Mitoses were not seen, nor was there inflammation or ulceration. UEA1 immunohistochemical staining of endothelial cells was strongly positive (Figs. 1 and 3), but weak and less extensive staining was found with antibodies to VIII-RA. Stains for S-100 and actin were negative in all cells of the lesion. The lesional tissue was interpreted to be consistent with an acquired tufted angioma.

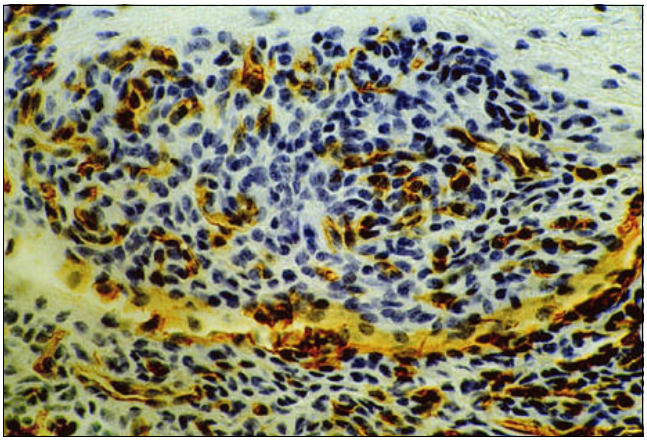

Figure 3: *UEA1 immunostaining clearly separates stained endothelial cells from the abundant unstained pericyte-like cells. A normal vessel from which the tuft appears to arise is seen cutting across the lower part of the illustration (hematoxylin, UEA1 immunostaining, magnification x400).*

Discussion

The most common vascular proliferation of the oral mucosa is the pyogenic granuloma. This is a reactive lesion that develops rapidly, bleeds easily and is usually associated with inflammation and ulceration. Clinically, it is often lobulated, pedunculated and red to purple and it may be hormone-sensitive.[16] In time, the lesion undergoes maturation with increased sclerosis and decreased bleeding. Microscopically, the lesion may show variable composition, reflecting its stage of development. An early, very active lesion shows an intense proliferation of capillaries and endothelial cells packed in a lobular manner into loose or fibromyxoid stroma[17] (Fig. 2b). The active lesion shows numerous mitoses. Secondary inflammation and edema are usual. The present case, however, has neither clinical nor microscopic features of a pyogenic granuloma. Its duration without significant change for 10 years is inconsistent with the progressive changes seen within weeks in a pyogenic granuloma. Microscopically, the localized tufts with extensive proliferation of non-endothelial spindle and polygonal cells surrounding vascular channels is distinctly different from the lobular, obviously capillary histology of the pyogenic granuloma (Fig. 2).

Hemangiomas are often congenital or develop in the neonatal period.[18] They usually cover a large area, may be macular or raised and usually resolve progressively in childhood.[18] The patient in this case report had a congenital vascular lesion of the skin of the back, thought to be a hemangioma, but there were no similar lesions of the head and neck. Clinically, the reported lesion differed from a hemangioma by its papular presentation and persistence. The small size of the lesion is unusual for both the hemangioma and the tufted angioma.

Microscopically, the capillary hemangioma exhibits a progression from a densely cellular proliferation of endothelial cells in the early stages to a lobular mass of well-formed capillaries in the mature phase, often resembling the pyogenic granuloma without the inflammatory features.[17] The microscopic appearance of the lesion in this case, exhibiting an abundant spindle and polygonal cell population, is unlike the lobular, capillary proliferation of a mature hemangioma.

The nature of the spindle and polygonal cells associated with the acquired tufted angioma is unclear. Studies in which they exhibit prominent actin staining suggest that the cells are pericytes.[5,11] However, cells in this case did not stain for actin, and Kumakiri and others,[12] who did not do immunohistochemical studies, were unable to find significant numbers of cytoplasmic microfilaments by electron microscopy. These results suggest that the perivascular cells in some cases may be immature and have not yet expressed sufficient actin to be detected. If the interpretation is that the cells are pericytes, then the differential diagnosis of hemangiopericytoma must be considered. However, the present case has shown no progression in at least a decade, ruling out this possibility on clinical grounds alone. Furthermore, the distribution of tufts arising from mature vasculature in discrete nodules is different from the diffuse growth pattern of the hemangiopericytoma, despite similarities at the cellular level.

Other vascular anomalies — including targetoid hemosiderotic hemangioma, verrucous hemangioma and cherry (senile) angioma — have clinical and histologic features that are not consistent with acquired tufted angioma.[17]

The present case has microscopic and immunohistochemical features more consistent with acquired tufted angioma than with other vascular lesions, but its small clinical size, its site and its lack of progression at some point in its clinical history are atypical when compared to many of the reported skin lesions. However, the clinical features of reported cases of acquired tufted angioma on skin are variable and are not in themselves diagnostic. Furthermore, the clinical presentation of this lesion is not characteristic of other well-defined vascular lesions either. Since this lesion has not previously been reported in the oral cavity, there are no standards for comparison of clinical features. Consequently, diagnosis is based on microscopic and immunohistochemical findings. ◆

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