

Adult-Onset Tufted Angioma: A Case Report and Review of the Literature

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Tufted angiomas (TAs) are benign vascular tumors, primarily occurring on the trunk and extremities of children younger than 5 years. Few cases occurring on the oral mucosa and in adults have been reported. Although they are typically isolated lesions, TA has been associated with port-wine stains. We describe a case of adult-onset TA on the lower lip mucosa.

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Case Report

A 36-year-old, healthy, nongravid woman presented with a 3-month history of a slowly enlarging asymptomatic growth on her lower lip. There was no history of trauma or similar appearing lesions. On physical examination, she had a 5×8-mm raised, red, nontender papule on the left lower lip (Figure 1). A shave biopsy of the lesion revealed a lobular pattern of small vessels with plump endothelial cells in the upper dermis with an edematous fibromyxoid stroma (Figure 2). At 2-week follow-up, surgical excision with narrow margins was performed. Interestingly, histopathologic examination demonstrated scattered round and ovoid dermal lobules in a “cannonball” distribution. These tufts primarily were composed of plump endothelial cells with interspersed narrow lymphaticlike channels (Figure 3). These features were consistent with a tufted angioma (TA). The procedure was well-tolerated, with minimal scarring and no recurrence at 3-month follow-up.

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Comment

TA, a benign vascular tumor, was first described by Nakagawa¹ in 1949 as an angioblastoma and then by Jones and Orkin² in 1989 as TA.²⁻¹⁰ While TA primarily affects children younger than 5 years, with no apparent gender predilection, there have been a few reports of adult-onset TA (Table). Associations with pregnancy, sites of trauma, and post-solid organ transplant are notable. However, so far, the varied morphology and clinical behavior of adult-onset TA have demonstrated no clear differences from childhood onset.^{2,4-15,20,21}

Clinically, TAs typically present as mottled red macules or as deep red to red-brown subcutaneous plaques or nodules varying in size from 2 to 5 cm². They usually are asymptomatic, but painful lesions have been reported. The clinical differential diagnosis for TA includes pyogenic granuloma, hemangioma of infancy, port-wine stain, vascular malformation, bacillary angiomatosis, lymphangioma, hemangiopericytoma, Kaposi sarcoma, and kaposiform hemangioendothelioma.^{2,7,20} TAs occur most frequently on the neck, trunk, and shoulders. They also have been demonstrated on the limbs, lower face, and, rarely, the oral mucosa.^{2,3,6,7}

There have been 3 previously reported cases of TA presenting on the oral mucosa. Daley⁶ reported on a 20-year-old woman with a reddish-brown 1-mm papule on her left lower lip of more than 10 years' duration. Kleinegger et al⁷ reported 2 other cases. One case involved a 34-year-old man with a purple-red 0.3-cm papule on the upper labial mucosa. The other case was a 52-year-old man who had a 0.2×0.2-cm blue, nonulcerated, submucosal nodule of the anterior floor of the mouth. This lesion was noted on follow-up evaluation after endosseous implants were placed in his right posterior maxillary region.⁷ Similar to our patient, all 3 cases of mucosal TA were painless, occurred beyond childhood, and were successfully treated by surgical excision.

Tufted Angioma



Figure 1. Asymptomatic raised, red, nontender papule on the left lower lip.

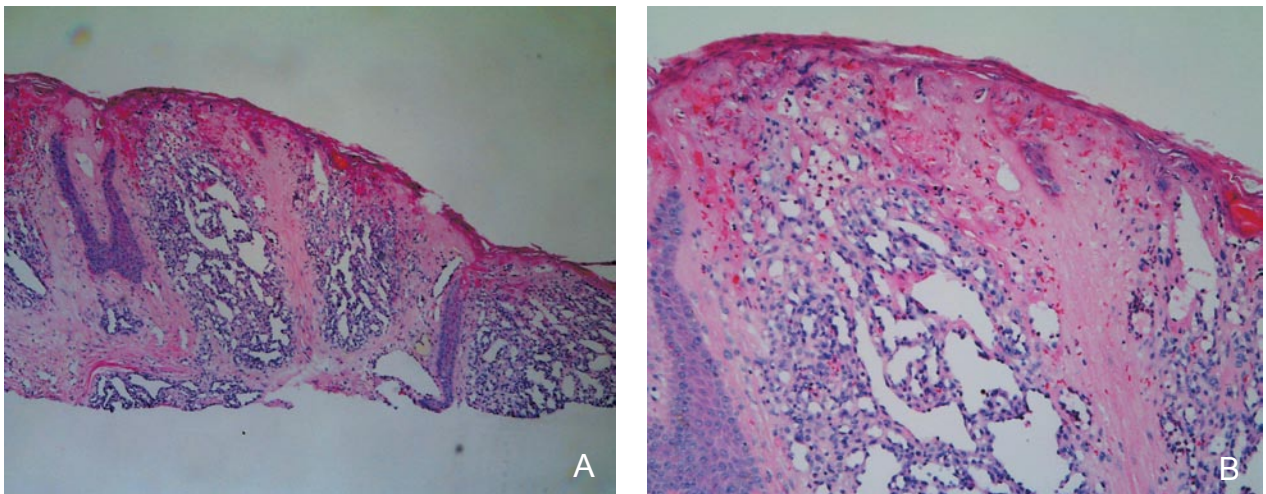


Figure 2. Initial shave biopsy of the lesion revealed a lobular pattern of small vessels with plump endothelial cells in the upper dermis with an edematous fibromyxoid stroma present (H&E, original magnifications $\times 10$ and 40)(A and B, respectively).

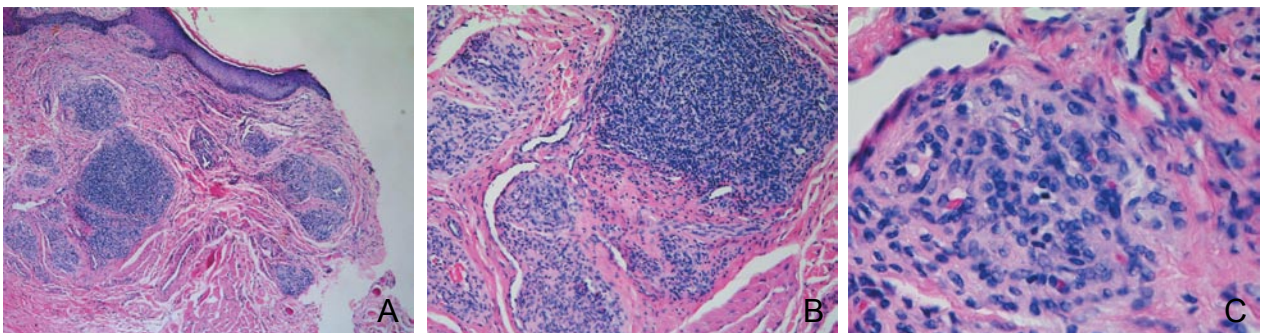


Figure 3. At 2-week follow-up, surgical excision was performed. Scattered round and ovoid dermal lobules in a “cannonball” distribution were present. These tufts primarily were composed of plump endothelial cells with interspersed narrow lymphaticlike channels (H&E; original magnifications $\times 10$, 40 , and 100)(A, B, and C, respectively).

Cases of Adult-onset Tufted Angioma*

Case Report (publication year)	Sex (age, y)	Pain	Anatomic Location	Duration	Other Clinical Characteristics	Treatment
Okada et al ³ (2000)	M (53)	+	Shoulder	2 y	NR	Surgical excision; F/U 1 y, no recurrence
Jones and Orkin ² (1989)	F (48)	–	Neck	2 y	Independent port- wine stain on arm	No reported treatment
	M (56)	–	Axilla	Few wk	NR	No reported treatment
	F (60)	–	Neck	2.5 y	NR	No reported treatment
Kleinegger et al ⁷ (2000)	M (34)	–	Upper labial mucosa	Unknown	NR	Surgical excision
	M (52)	–	Oral mucosa	10 d	Recent place- ment of endos- seous implants	Surgical excision; F/U 3 mo, no recurrence
Miyamoto et al ¹⁰ (1992)	F (62)	+	Upper chest and neck	12 mo	Transient spontaneous regression	Surgical excision
Romano et al ¹¹ (2004)	F (28)	+	Neck	2 y	Actively growing lesion	Intralesional interferon alfa-2a; F/U 1 y, no pain, lesions unchanged
Chu and LeBoit ¹² (1992)	M (59)	+	Axilla and arm	1d	10 d post- liver transplant	Spontaneous involution in 2 mo
Kim et al ¹³ (1999)	F (26)	–	Neck and chest	4 mo	Pregnant; had similar lesions with previous pregnancy	Spontaneous involution by 6-mo F/U
Sumitra and Yesudian ¹⁴ (1994)	F (40)	+	Back	2 y	Occurred at site of healed human bite 6 mo prior	Surgical excision; recurrence at 10 mo

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Case Report (publication year)	Sex (age, y)	Pain	Anatomic Location	Duration	Other Clinical Characteristics	Treatment
Pietroletti et al ¹⁵ (2002)	F (33)	+	Anus	Few wk	Pregnant; lesion resembled anal fissure	Surgical excision; F/U 21 mo, no recurrence
Frenk et al ¹⁶ (1990)	F (25)	–	Back	5 y	Increased in size and number during pregnancy	No reported treatment
Ban et al ¹⁷ (2000)	M (58)	–	Chest	5 y	NR	Evidence of central regression
Dewerd et al ¹⁸ (1998)	F (81)	–	Neck and shoulder	2 y	NR	Pulsed dye laser was unsuccessful
Hebeda et al ¹⁹ (1993)	F (66)	+	Neck	1 y	Positive hepatitis B and C antibodies	No reported treatment

*M indicates male; F, female; +, present; –, absent; NR, none reported; F/U, follow-up.

The histopathologic features of TA include focal arrangements of tightly packed capillary vessels surrounded by connective tissue distributed in the dermis and subcutaneous tissue.² The focal arrangements typically have been described as having a cannonball appearance. Cellular atypia and mitotic activity are absent. The histologic differential diagnosis includes pyogenic granuloma, Kaposi sarcoma, kaposiform hemangioendothelioma with cutaneous involvement, and angiosarcoma.^{2,4,5}

Therapeutic management of TA varies. Monitoring lesions is an acceptable approach of treating asymptomatic lesions. There have been reports of spontaneous remission of TAs^{4,9,12,20}; however, some lesions have persisted for more than 50 years.² Surgical excision has been performed successfully on smaller lesions; however, recurrences have been reported.² High-dose prednisone and interferon alfa-2a have been shown to reduce lesion size, prevent lateral spread, and offer pain relief in some lesions. Other modalities such as

cryotherapy, radiotherapy, and laser therapy have been used with variable results.¹¹

Conclusion

Our case of adult-onset TA occurring on the lip is the fourth reported case of TA involving the oral mucosa. TA should not be overlooked in the differential diagnosis of vascular neoplasms affecting the oral mucosa in both children and adults.

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