Acquired Tufted Angioma of Upper Lip: Case Report and Review of the Literature

Amarpreet Sabharwal · Alfredo Aguirre ·
Talal M. Zahid · Germain Jean-Charles ·
Michael N. Hatton

Received: 31 January 2013 / Accepted: 13 March 2013 / Published online: 23 March 2013
© Springer Science+Business Media New York 2013

Abstract Acquired tufted angioma is a rare, cutaneous and slow growing benign vascular neoplasm that usually affects pediatric populations. Only six cases of oral mucosal acquired tufted angioma have been reported in the English language literature. We present in this report the seventh case and discuss its clinical and pathological features.

Keywords Acquired · Tufted · Angioma · Upper lip · Vascular · Neoplasm

Introduction

Acquired tufted angioma (ATA) was first identified and later characterized by Jones in 1976 and 1989, respectively [1, 2]. Alternative terms such as tufted hemangioma, progressive capillary hemangioma and angioblastoma of Nakagawa have been widely used synonymously in the scientific literature [3–5]. The World Health Organization

(WHO) classification recognizes ATA as a distinct vascular entity and defines it as “an unusual, acquired, benign vascular neoplasm characterized by slow, indolent growth” [3]. While most cases of ATA are seen in children less than 5 years of age, sporadic cases in adults have been documented in the literature [3, 6, 7]. Most cases of ATA involve the skin of the trunk and limbs [6, 7]. Mucosal involvement by ATA is extremely rare. The first oral case was described as recently as 2000 [8] and only six *bona fide* cases have been reported in the English language literature thus far (Table 1) [8–12].

We present a case of ATA of the maxillary vermilion border of the lip and provide a comparison of mucosal ATA with ATA of cutaneous distribution.

Case Report

A 47 years old Caucasian male presented at the Postdoctoral Dental Clinic, University at Buffalo for routine periodontal assessment. During the initial examination, a slowly enlarging, painless, dull red papule (0.5×0.5 cm across) was noted on the left vermilion border of the upper lip (Fig. 1). The lesion had a slight central depression. The patient reported that he noticed the lesion 8 weeks prior to consultation. His medical history was unremarkable and no previous trauma to the site was reported. However, the patient admitted to occasional irritation resulting from his maxillary anterior teeth. At a two-weeks follow-up, the lesion was clinically unchanged and thus, we decided to excise it.

Gross examination of the specimen showed a wedge of oral mucosa with a white color, measuring $0.7 \times 0.4 \times 0.4$ cm. The base of the specimen was inked and then the tissue was trisected and submitted *in toto* for embedding.

A. Sabharwal · A. Aguirre (✉) · M. N. Hatton
Department of Oral Diagnostic Sciences, Advanced Oral and
Maxillofacial Pathology Program, School of Dental Medicine,
University at Buffalo, 355 Squire Hall, Buffalo, NY 14214, USA
e-mail: aguirr@buffalo.edu

T. M. Zahid
University at Buffalo, Buffalo, NY, USA

T. M. Zahid
Dr. Sulaiman Al Habib Medical Center Group,
Riyadh, Saudi Arabia

G. Jean-Charles
Department of Medicine and Director of Dental Clinic Division
of HIV Medicine, Albany Medical Center, Albany, NY, USA

Table 1 Acquired tufted angioma of the oral mucosa and lip vermillion

Case	Sex/ age	Location	Size	Duration	Treatment	Follow-up
Sabharwal et al. 2013 (present case) (n = 1)	M/47	Upper lip vermillion	0.5 × 0.5 cm	8 weeks	Excision	3 years (no recurrence)
Chaudhari [10] (n = 1)	M/40	Anterior dorsal tongue	0.5 × 0.5 cm	4–5 months	Excision	Not provided
Chaves et al. [9] (n = 1)	M/12	Upper lip vermillion	0.8 cm diameter	2 months	No treatment	Not available
Lee et al. [11] (n = 1)	F/36	Lower lip	0.5 × 0.8 cm	3 months	Excision	3 months (no recurrence)
Daley et al. [12] (n = 1)	F/20	Lower lip mucosa	0.1 cm diameter	10 years	Excision	Not provided
Kleinegger et al. [8] (n = 2)	M/34	Upper lip mucosa	0.3 cm diameter	Unknown	Excision	Not provided
	M/52	Anterior floor of mouth	0.2 × 0.2 cm	10 days	Excision	3 months (no recurrence)

Age is in years

M male, F female

Fig. 1 The left vermillion border of the upper lip shows a red papule with a soft consistency

Microscopic examination of the excised specimen revealed a stratified squamous epithelium with hyperkeratosis, hypergranulosis and irregular acanthosis. The superficial lamina propria shows ectatic capillary channels and discrete, densely cellular capillary lobules in a “cannonball” pattern (Figs. 2, 3). Immunohistochemical stains for CD34 and vimentin were diffusely positive in the cellular capillary lobules. The microscopic and immunohistochemical findings supported a diagnosis of acquired tufted angioma. The two-weeks postsurgical follow-up showed a satisfactory uneventful healing (Fig. 4). No recurrence was seen at a three-years follow-up.

Fig. 2 Low magnification showing a hyperkeratinized stratified squamous epithelium. The subjacent lamina propria shows dilated capillaries and discrete cellular lobules separated by dense fibrous connective tissue (Hematoxylin and eosin; original magnification ×20) **Fig. 3** Higher magnification showing distinct clusters of spindle and polygonal cells with interspersed slit-like blood vessels lined by endothelial cells (“cannonball nests”) (Hematoxylin and eosin; original magnification ×200)

Fig. 4 Clinical presentation 2 weeks after surgery showing an uneventful healing of the biopsy site

Discussion

Acquired tufted angioma is a benign vascular neoplasm that usually affects the trunk and extremities of children [3, 6, 7]. Although cutaneous ATA occurs over a wide spectrum of age, most cases are diagnosed before the age of 5, typically during the first year of life [6, 7]. The adult onset ATA of the skin represents the minority of cases and shows a wide age range with relatively more cases seen between 3rd and 6th decades of life [6, 11]. In contrast, ATA of the oral mucosa and vermilion border is more common in adults [8, 10–12] and only one case has been documented in a 12 years old child [9].

While most cases of ATA are sporadic, some show a familial predisposition suggesting an autosomal dominant inheritance pattern [13]. Overall, cutaneous ATA shows no gender predilection [2, 6]. However, adult onset ATA of the skin shows a slight female predilection [6, 11]. This finding is in contradistinction to mucosal and vermilion ATA, where males are affected twice as much as females [8–12].

Some authors have divided cutaneous ATA into solitary lesions and lesions associated with coagulopathies/syndrome. Solitary cutaneous ATA is typically a lesion with poorly defined borders, ranging from few to many centimeters in size. They range in color from dusky red to violaceous plaques, with, bluish or reddish hue. Their surface may show occasional nodularities, may be warm to touch and tender on palpation. Pediatric and adult onset solitary ATAs do not differ substantially in their clinical presentation [11, 14–16]. Solitary cutaneous ATA is an asymptomatic lesion with excellent prognosis and cosmetic outcome. In contrast, cutaneous ATA associated with chronic coagulopathy, thrombocytopenia (TCP) or, more importantly, Kasabach-Merritt syndrome (KMS) are poor symptomatic, show an infiltrative pattern and have a poor

cosmetic outcome [14]. Occasionally, ATA of the skin has been reported to be associated with port-wine stains, pregnancy, and Crohn's disease [17–19]. Cutaneous ATA lesions associated with KMS and TCP may show induration, tenderness and pain. In addition, cutaneous atrophy, and joint/muscle pathoses resulting in restricted mobility after resolution or regression, may be observed [14].

Mucosal ATA has a consistent clinical presentation. These lesions display a papular to nodular configuration and typically measure <1 cm in their largest dimension. They are asymptomatic and show an intact red-brown to purple-blue surface [8, 10–12]. One case with ulcerated surface is however documented in the literature [9]. Most mucosal ATAs are solitary and only one case has been reported to be associated with a port-wine stain [12].

Clinical differential diagnosis of a lesion presenting in the maxillary vermilion border should include reactive salivary gland lesions (such as a mucocele and a sialolith) and a salivary gland neoplasm (i.e., canalicular adenoma).

Microscopically, a typical ATA shows multilobular vascular proliferation occupying the supportive connective tissue stroma of skin or mucosa. The so called “cannonball” nests consists of discrete lobular aggregates of spindle and polygonal cells with interspersed endothelial cells that line delicate slit-like blood capillaries [3, 8–12, 20]. The presence of endothelial cells has been confirmed by ultrastructural and immunohistochemical studies [12]. The spindle and polygonal cells contain cytofilaments and microfilaments. Dense fibrous connective tissue separates these lobules of cells [12]. ATA shows overlapping histological features with Kaposiform hemangioendothelioma (KHE) that include the presence of glomeruloid structures and a lymphatic network. Although distinguishing KHE from ATA may not be possible at all times, the presence of a conspicuous and prominent lymphatic network surrounding the cellular tumor lobules favors a diagnosis of KHE [20, 21]. In any event, it is believed that KHE and ATA may share a common biologic ancestry as indicated by reported histological and immunohistochemical similarities. These findings suggest that KHE and ATA may represent a spectrum of the same lesion [22–25]. The microscopic differential diagnosis of ATA may also include juvenile capillary hemangioma. However, capillary hemangioma does not show the presence of cannonball nests and glomeruloid structures.

Spontaneous regression of ATA is a fortunate but rare occurrence that has been amply documented in the literature [26–28]. However, a persistent ATA requires treatment. Medical treatments such as corticosteroids, interferon- α and laser therapy have all shown variable treatment outcomes [14, 29–33]. While, treatment guidelines have not been laid out as yet [14], the small size of mucosal ATA yields well to surgical excision [8–12].

Further reports on mucosal ATA may contribute to create a robust database that advances an accurate understanding of the biological behavior and treatment outcomes for this lesion.

References

1. Jones EW. Malignant vascular tumours. *Clin Exp Dermatol*. 1976;1:287–312.
2. Jones EW, Orkin M. Tufted angioma (angioblastoma): a benign progressive angioma, not to be confused with Kaposi's sarcoma or low-grade angiosarcoma. *J Am Acad Dermatol*. 1989;20:214–25.
3. LeBoit PE, Burg G, Weedon D, Sarasin A, editors. World Health Organization classification of tumors pathology and genetics of skin tumours. Lyon: AIRCPress; 2006. p. 239–40.
4. Macmillan A, Champion RH. Progressive capillary haemangioma. *Br J Dermatol*. 1971;85:492.
5. Cho KH. Tufted angioma: is it the same as angioblastoma (nakagawa)? *Arch Dermatol*. 1997;133:789.
6. Okada E, Tamura A, Ishikawa O, et al. Tufted angioma (angioblastoma): case report and review of 41 cases in the Japanese literature. *Clin Exp Dermatol*. 2000;25:627–30.
7. Ward KA, Kennedy CTC, Ashworth MT. Acquired tufted angioma frequently develops at sites other than the neck and upper trunk. *Clin Exp Dermatol*. 1996;21:80.
8. Kleinegger CL, Hammond HL, Vincent SD, et al. Acquired tufted angioma: a unique vascular lesion not previously reported in the oral mucosa. *Br J Dermatol*. 2000;142:794–9.
9. Chaves ACM, de Oliveira MG, Rados PV, et al. Tufted angioma in the upper lip: a report of a case. *International journal of pediatric otorhinolaryngology extra*. 2009;4:173–6.
10. Chaudhary RB. Acquired tufted angioma of the tongue-A case report. *JIDA*. 2011;5:561–2.
11. Lee B, Chiu M, Soriano T, et al. Adult-onset tufted angioma: a case report and review of the literature. *Cutis*. 2006;78:341–5.
12. Daley T. Acquired tufted angioma of the lower lip mucosa. *J Can Dent Assoc*. 2000;66:137.
13. Tille J-C, Morris MA, Bründler M-A, et al. Familial predisposition of tufted angioma: identification of blood and lymphatic vascular components. *Clin Genet*. 2003;63:393–9.
14. Osio A, Fraitag S, Hadj-Rabia S, et al. Clinical spectrum of tufted angiomas in childhood. A report of 13 cases and a review of the literature. *Arch Dermatol*. 2010;146:758–63.
15. Heron MD, Coffin CM, Vanderhooft SL. Tufted angiomas: variability of the clinical morphology. *Pediatr Dermatol*. 2002;19:394–401.
16. Reddy IS, Anuradha SVN, Swarnlata G. Congenital giant tufted angioma. *Indian J Dermatol Venereol Leprol*. 2009;75:639.
17. Al-Zaabi AM, Ghazarian D, Greenberg GR, et al. Eruptive tufted angiomas in a patient with Crohn's disease. *J Clin Pathol*. 2005;58:214–6.
18. Kim Y-K, Kim H-J, Lee K-G. Acquired tufted angioma associated with pregnancy. *Clin Exp Dermatol*. 1992;17:458–9.
19. Kim T-H, Choi EH, Ahn SK, et al. Vascular tumors arising in port-wine stains: two cases of pyogenic granuloma and a case of acquired tufted angioma. *J Dermatol*. 1999;26:813–6.
20. Weiss SW, Goldblum JR (eds) Enzinger and Weiss's soft tissue tumors. Mosby. 2007; 654–5.
21. Weiss SW, Goldblum JR (eds) Enzinger and Weiss's soft tissue tumors. Mosby. 2007; 689.
22. Chu C-Y, Hsiao C-H, Chiu H-C. Transformation between kaposiform hemangioendothelioma and tufted angioma. *Dermatology*. 2003;206:334–7.
23. Brasanac D, Janic D, Boricic I, et al. Retroperitoneal kaposiform hemangioendothelioma with tufted angioma-like features in an infant with Kasabach-Merritt syndrome. *Pathol Int*. 2003;53:627–31.
24. Huu ARL, Jokinen CH, Ruben BP, et al. Expression of prox1, lymphatic endothelial nuclear transcription factor, in kaposiform hemangioendothelioma and tufted angioma. *Am J Surg Pathol*. 2010;34:1563–73.
25. Arai E, Kuramochi A, Tsuchida T, Tsuneyoshi M, et al. Usefulness of D2–40 immunohistochemistry for differentiation between kaposiform hemangioendothelioma and tufted angioma. *J Cutan Pathol*. 2006;33:492–7.
26. Browning J, Frieden I, Baselga E, et al. Congenital, self-regressing tufted angioma. *Arch Dermatol*. 2006;142:749–51.
27. Ishikawa K, Hatano Y, Ichikawa H, et al. The spontaneous regression of tufted angioma. A case of regression after two recurrences and a review of 27 cases reported in the literature. *Dermatology*. 2005;210:346–8.
28. Miyamoto T, Mihara M, Mishima E, et al. Acquired tufted angioma showing spontaneous regression. *Br J Dermatol*. 1992;127:645–8.
29. Na J-I, Cho K-H, Kim Y-G, et al. Angioblastoma showing aggravation after treatment with long-pulsed Nd: YAG laser (1,064 nm). *Pediatr Dermatol*. 2007;24:397–400.
30. Chiu C-S, Yang L-C, Hong H-S, et al. Treatment of tufted angioma with intensified pulsed light. *J Dermatol Treat*. 2007;18:109–11.
31. Wollina U. Interferon for tufted angioma. *Pediatr Dermatol*. 1999;16:338.
32. Robenzadeh A, Don PC, Weinberg JM. Treatment of tufted angioma with interferon alfa: role of bFGF. *Pediatr Dermatol*. 1998;15:482.
33. Munn SE, Jackson JE, Jones RR. Tufted haemangioma responding to high-dose systemic steroids: a case report and review of the literature. *Clin Exp Dermatol*. 1994;19:511–4.