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CASE REPORT

A case of solitary angiokeratoma circumscriptum of the tongue

Dilde soliter anjiyokeratom sirkumskriptum: Olgu sunumu

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Angiokeratoma circumscriptum is an uncommon vascular entity. The lesion is present at birth and usually involves the lower extremities. A 9-year-old girl presented with multiple pink-colored, small-raised lesions in the dorsal part of her tongue, which had been present since birth. There was no history of trauma, cold injury, or similar lesions elsewhere in her body. She complained of pain and rare bleeding from the lesions after eating hard foods. Physical examination showed multiple, grouped, erythematous, shiny papules mostly in the dorsal aspect of the tongue. There were no other lesions in other parts of the oral mucosa. After an incisional biopsy of the lesion, the diagnosis was made as solitary angiokeratoma circumscriptum.

Key Words: Angiokeratoma; tongue neoplasms.

Anjiyokeratom sirkumskriptum nadir görülen bir vasküler bozukluktur. Lezyonlar doğum sırasında vardır ve genellikle alt ekstremiteyi etkiler. Dokuz yaşındaki bir kız çocuğu, dilin dorsal bölümünde pembe renkli, küçük yükseltiler oluşturan lezyonlarla başvurdu. Bu durumun doğumdan beri var olduğu öğrenildi. Hastanın öyküsünde geçirilmiş travma, şiddetli soğuk etkisi veya vücudun başka yerlerinde benzer lezyon oluşumu yoktu. Sert yiyeceklerden sonra ağrı ve nadir olarak lezyondan kan gelmesi gibi yakınmaları vardı. Fizik muayenede, dilin dorsal yüzünde gruplar halinde, eritematöz, parlak papüller görüldü. Ağız mukozasında başka lezyon yoktu. Lezyondan alınan insizyonel biyopsiden sonra soliter anjiyokeratom sirkumskriptum tanısı kondu.

Anahtar Sözcükler: Anjiyokeratom; dil neoplazileri.

Angiokeratomas are a group of vascular lesions that involve the papillary dermis and produce hyperkeratosis of the dermis.[1] Different types of angiokeratomas have been described.[2] Angiokeratoma circumscriptum described by Fabry in 1915 is a rare variant of this disorder.[2] In most of the cases, these lesions are present at birth. There is a peculiar sex ratio of three females to one male. Histopathologically, there are varying degrees of hyperkeratosis, papillomatosis, and irregular acanthosis. Greatly dilated capillary spaces are seen partly or completely enclosed by elongated rete ridges in the papillomatous epidermis. The acanthotic epidermis encircles the vascular spaces (blood cysts) where, occasionally, organized thrombi may be found.[3] The lesions are mostly found in the lower extremities, and also may be observed in the trunk. Oral involvement by angiokeratoma circumscriptum is uncommon. It is usually seen in Fabry's disease and fucosidosis as a component of more generalized cutaneous disease.[4] In this report, we describe solitary angiokeratoma circumscriptum in the dorsal aspect of the tongue.

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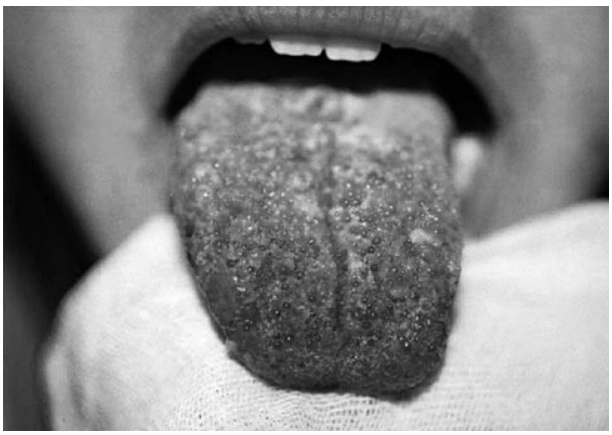

Fig. 1. *Multiple, grouped, erythematous, shiny papules on the tongue.*

CASE REPORT

A 9-year-old girl was referred to the otorhinolaryngology clinic for an evaluation of multiple pink-colored, small-raised lesions in the dorsal part of her tongue, which had been present since birth. There was no history of trauma, cold injury, or similar familial lesions elsewhere in her body. It started as a single raised lesion on the tip of the tongue and then gradually increased in number and extended onto the dorsal surface of the tongue. She complained of pain and rare bleeding from the lesions after eating hard foods.

On physical examination, multiple, grouped, erythematous, shiny papules were observed, some of which had keratotic top seen mostly in the dorsal aspect of the anterior two-thirds of the tongue (Fig. 1). There were no other lesions in other parts of the oral mucosa. After an incisional biopsy of the lesion, the diagnosis of solitary angiokeratoma circumscriptum was made.

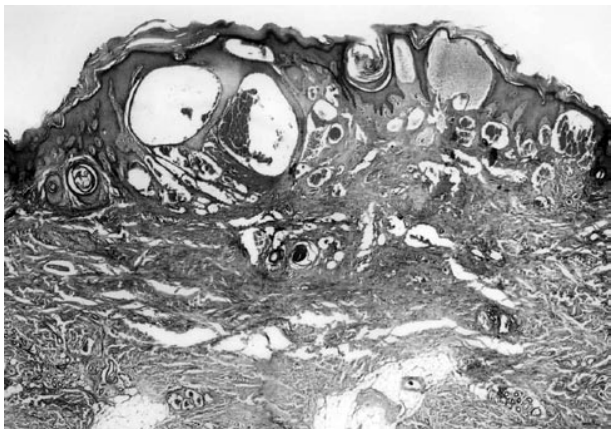

Fig. 2. *Large dilated capillary spaces are seen in the hyperkeratotic and irregular acanthotic epidermis, filled with erythrocytes and organizing thrombi (H-E x 40).*

Histopathologic examination of the lesion showed marked hyperkeratosis, irregular acanthosis, and papillomatosis with large, dilated capillary spaces lined by normally appearing endothelium and filled with erythrocytes and organizing thrombi (Fig. 2, 3).

DISCUSSION

Angiokeratomas are a set of dermatoses, defined histologically by dilated blood vessels in the upper part of the dermis, with the epidermal growth as a secondary reactive phenomenon. Different entities causing vessel ectasia lead to many clinical variants of angiokeratoma. Current classification distinguishes between widespread forms (angiokeratoma corporis diffusum), which is usually associated with an inborn error of metabolism, and localized forms, which include solitary angiokeratoma, Fordyce's angiokeratoma, angiokeratoma circumscriptum naeviforme

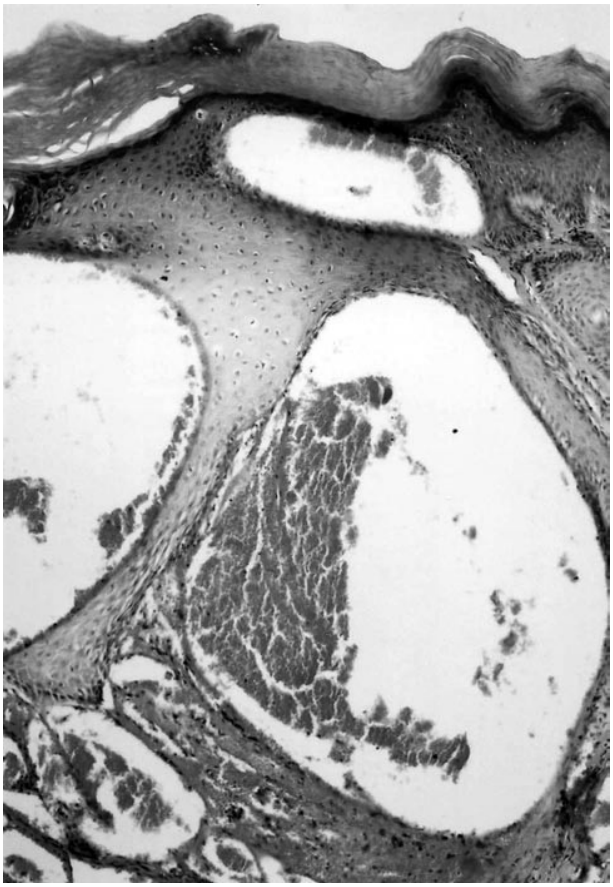

Fig. 3. *High magnification of Fig. 2. Subepidermal dilated capillary lumina containing erythrocytes (H-E x 100).*

and angiokeratoma of Mibelli.[5] Angiokeratoma circumscriptum is a rare type, characterized by aggregates of hyperkeratotic erythematous papules and nodules. In our case, the diagnosis of angiokeratoma circumscriptum was made on the basis of histologic features and the gross appearance of the lesion.

The etiology is not clear in angiokeratoma circumscriptum. It is thought to be a telangiectatic lesion arising from local injury to papillary capillaries, either from trauma or venous hypertension, with secondary epithelial proliferation causing acanthosis and hyperkeratosis. Despite its presence at birth, there is no obvious hereditary pattern. It occurs predominantly in the lower extremities, but other sites include the prepuce, shaft, and glans of the penis, vulva and clitoris. Involvement of the oral mucosa by angiokeratoma is uncommon.[6] It usually arises as pink to red papules, 1 to 3 mm in size. Initially, it is soft and compressible, it gradually enlarges, later becoming darker in color with a keratotic and warty surface. Eventually, the lesion is unilateral, circumscribed, and keratinized.[3,6] Angiokeratoma circumscriptum of the tongue was first described by Leung and Jordan[6] and solitary involvement of the tongue is rare in the literature.[7] Most commonly, it is a component of generalized systemic disorder in Fabry's disease or fucosidosis, where multiple angiokeratomas are present in the skin and oral mucosa. However, in our case the lesion was limited to the tongue.

Angiokeratoma circumscriptum must be differentiated from malignant melanoma, lymphangioma circumscriptum, capillary aneurysms, and verrucous hemangioma. Although angiokeratoma involves the superficial papillary dermis, verrucous hemangioma involves all levels of the dermis and subcutaneous tissue. In addition, the blood cysts that lie within the acanthotic epidermis in angiokeratoma are not observed in verrucous hemangioma. The differentiation of angiokeratoma circumscriptum from verrucous hemangioma is histologically confirmed by the lack of vessel proliferation in the hypodermis.[3,8] Episodes of bleeding and infection following trauma or scratching are frequent symptoms associated with angiokeratomas.[9] In our patient, there were no predisposing factors for angiokeratoma. There was no systemic disease nor evidence for a local factor associated with the condition.

Treatment mostly depends on the severity of symptoms. Because they are often too extensive to perform surgical excision, superficial ablative therapies using carbon dioxide or argon laser, cryotherapy, and electrocautery are known alternatives for treating these lesions.[10]

REFERENCES

1. Dolph JL, Demuth RJ, Miller SH. Angiokeratoma circumscriptum of the index finger in a child. Plast Reconstr Surg 1981;67:221-3.
2. Lever WF, Schaumburg-Lever G, editors. Tumors of vascular tissue. In: Histopathology of the skin. 6th ed. Philadelphia: J. B. Lippincott; 1990. p. 692-3.
3. Pasyk KA, Argenta LC, Schelbert EB. Angiokeratoma circumscriptum: successful treatment with the argon laser. Ann Plast Surg 1988;20:183-90.
4. Prindiville DE, Stern D. Oral lesions in fucosidosis. J Oral Surg 1976;34:603-8.
5. Somasundaram V, Premalatha S, Rao NR, Razack EM, Zahra A. Hemangiectatic hypertrophy with angiokeratoma circumscriptum. Int J Dermatol 1988;27:45-6.
6. Leung CS, Jordan RC. Solitary angiokeratoma of the oral cavity. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1997;84:51-3.
7. Vijaikumar M, Thappa DM, Karthikeyan K, Jayanthi S. Angiokeratoma circumscriptum of the tongue. Pediatr Dermatol 2003;20:180-2.
8. Sardana K, Koranne RV, Sharma RC, Mahajan S. Angiokeratoma circumscriptum naeviforme: rare presentation on the neck. Australas J Dermatol 2001;42:294-5.
9. Yang CH, Ohara K. Successful surgical treatment of verrucous hemangioma: a combined approach. Dermatol Surg 2002;28:913-19.
10. del Pozo J, Fonseca E. Angiokeratoma circumscriptum naeviforme: successful treatment with carbon-dioxide laser vaporization. Dermatol Surg 2005;31:232-6.