

CASE REPORT

Solitary angiokeratoma of the tonsillar pillar of the oral cavity

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Abstract

Solitary angiokeratoma has rarely been described in oral mucosa, mainly in the tongue, where the main concern is either aesthetical or due to bleeding problems. We present a case of solitary angiokeratoma of the tonsillar pillar in a 68-year-old man. Histologically, the morphology was typical of angiokeratoma. It showed an immunohistochemical pattern in consonance with a blood vessel origin, with expression of CD31, CD34, and von Willebrand factor. The lesion did not express D2-40. No other malformation or metabolic disorder was found in the patient. The lesion was surgically removed and due to the disproportionate post-surgery bleeding, the patient was studied by the Hematology Service, and she was diagnosed as an inhibitor of Factor VIII carrier.

Keywords: angiokeratoma, Fabry's disease, Fordyce, Mibelli.

□ Introduction

Angiokeratoma (AK) is a rare muco-cutaneous lesion with a distinctive morphologic pattern. From a clinical point of view, it can show either a generalized and diffuse form of presentation, or a localized one. In the latter, four clinical forms are included, which are as follows: (a) papular lesions limited to the legs; (b) bilateral lesions on the dorsum of toes and fingers (Mibelly type); (c) lesions limited to the scrotum or vulva (Fordyce type); and (d) a congenital form in which multiple lesions on the legs are usually evidenced [1, 2].

AK involving the oral cavity is frequent in a context of the diffuse form of presentation, usually in the context of a metabolic disorder, such as Fabry's disease or fucosidosis [3, 4]. Nevertheless, oral AK in a solitary form of presentation is rare and not many cases have been reported in literature [1, 2, 5–7].

The solitary form has been mainly located on the tongue [1, 2, 6–9], and to the best of our knowledge, this is the first case reported of AK of the tonsillar pillar. In that latter location, a differential diagnosis with the most common tonsillar lymphangiomatous polyp should be done.

□ Patient, Methods and Results

A 68-year-old non-smoker white Caucasian man came to the emergency area of the Hospital due to oral bleeding. Three years before, a supraglottic laryngectomy had been performed on him, due to a squamous cell carcinoma of larynx.

The exploration showed a verrucous lesion at the left tonsillar posterior pillar (1 cm in diameter). The lesion was removed and morphologically studied with Hematoxylin-Eosin stain. As Figure 1 shows, wide

blood vessels were evidenced contacting the surface epithelium. The latter appeared hyperplastic, acanthotic and focally ulcerated. Neither the epithelium, nor the endothelial lining of the vessels showed any dysplasia. No vacuolation or swelling was evidenced in the endothelial cells.

We also performed an immunohistochemical study with the following primary antibodies: CD31 (DakoCytomation, monoclonal mouse anti-human, clone JC70A, isotype IgG1, kappa, code M0823), CD34 (DakoCytomation, monoclonal mouse anti-human, clone QBEnd 10, isotype IgG1, kappa, code 7165), Factor VIII (DakoCytomation, monoclonal mouse anti-human, von Willebrand factor, clone F8/86, isotype IgG1, kappa, code M0616), and D2-40 (DakoCytomation monoclonal mouse anti-human; clone D2-40, IgG1, kappa, code M3619).

The endothelium of the lesion expressed CD31, CD34 and Factor VIII. It was negative for D2-40 (Figure 2). The lesion was therefore diagnosed as angiokeratoma.

Due to an abundant amount of post-biopsy bleeding, the patient was examined by the Service of Hematology, and was diagnosed as carrying an acquired inhibitor of Factor VIII.

No other systemic or metabolic diseases were found and no other oral or nasopharyngeal lesions were evidenced. A skin examination was performed, but no lesions suspicious of angiokeratoma were found.

Owing to the oncological history of the patient, a cervico-thoracic and abdominal scanner was performed and the only evidenced lesion was an adrenal adenoma of 2 cm in diameter. Therefore, since the oral location was the only one involved by the AK in this patient, the final diagnosis was solitary oral angiokeratoma.

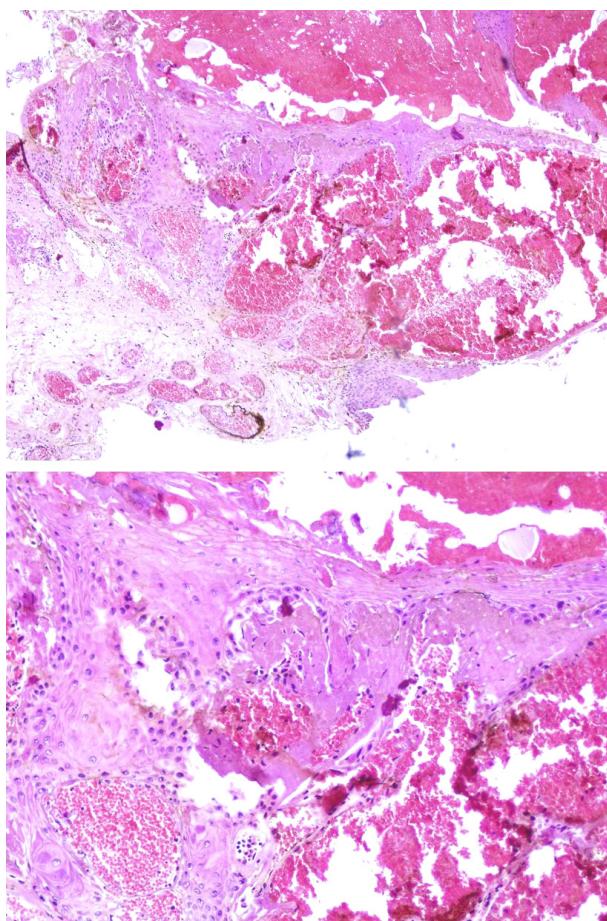


Figure 1 – The removed lesion showed a hyperplastic epithelium, with hyperkeratosis and acanthosis (top). A closer view shows how the dilated vessels were closely attached to the epithelium (bottom). No endothelial atypia was evidenced.

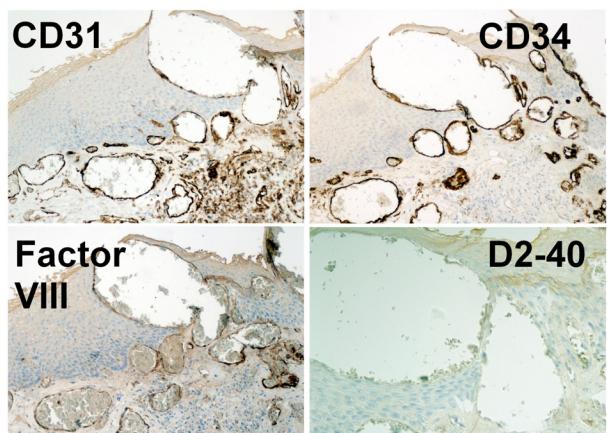


Figure 2 – Immunohistochemical profile of the lesion, with expression of CD31, CD34 and Factor VIII by the vessels. No expression of D2-40 was seen.

Discussion

Angiokeratoma (AK) has rarely been described in the oral cavity [2, 5, 8–10], sometimes in the context of angiokeratoma corporis diffusum (Fabry's disease) [11]. AK of the tongue has occasionally been described associated to AK of the scrotum, in the Fordyce type [12, 13], or in association with AKs of scrotum plus jejunum [14]. AK has hardly ever been described in oral

cavity as a solitary lesion, out of a context of a metabolic disorder [1, 2, 5–7, 15]. Nevertheless, as some authors admit, the lesion is probably more frequent than reported, due to its innocuous character [15].

Due to the low frequency of such a form of presentation, the clinical diagnosis is not always straightforward and the differential diagnosis must be established not only with other vascular lesions, but also with melanocytic lesions of the oral cavity [3, 15].

In the current report, we studied the expression of podoplanin by AK. Podoplanin (marked by the antibody D2-40) has been considered as a good marker for lymphatic differentiation [16–18]. Therefore, it is expected that tumors derived from blood vessels do not express the marker. Some tumors, which traditionally were considered as originating from blood vessels, are now considered as lymphatic in their origin, due to an expression of D2-40. That is the case of angiosarcomas [17, 19, 20], or hobnail hemangiomas [21]. On the other hand, a certain expression of D2-40 is still admitted in other tumors of alleged blood-vessel origin, such as tufted angioma [22].

The current case did not show any expression of podoplanin, as is expected for a lesion of alleged origin from blood vessels. This fact was also important in distinguishing the current case from the more common tonsillar lymphangiomatous polyp, which is made of dilated lymphatic channels [23], the latter expressing D2-40. Some other features can also be of help in distinguishing AK from the tonsillar lymphangiomatous polyp. In the latter, for instance, the lymphatic channels are packed with lymphocytes many times [23]. To the best of our knowledge, the expression of D2-40 by solitary AK of the oral mucosa has never been studied before.

The fact that our patient presented an acquired inhibitor of Factor VIII seems to be a casual finding, since no relation has been described between AK and such an alteration in blood clotting.

Our case is also illustrative in another sense: such a well vascularized lesion can sometimes be a source of either recurrent or important bleeding, moreover if the patient has an unknown clotting defect, as it was with our case. Nevertheless, it is admitted that life-threatening bleeding is usually not a concern [15]. In that respect, it is interesting to remember how, although the first therapeutic option for KA, is surgery [2], an alternative treatment with diode laser has also been presented in literature [1]. Many times, the removal of the lesion is many times not needed and it is only done either for cosmetic or diagnostic reasons [15].

Conclusions

We present the first case of AK of the tonsillar pillar to the best of our knowledge. Therefore, such a diagnosis, although rare, should be included in the differential when evaluating any lesion in this location. This is important not only because the high number of vessels of AK can be a source of important post-surgical bleeding, but because metabolic and systemic disorders can be studied and therefore, properly ruled out.

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