Dermatology Practical & Conceptual

Lingual Epidermal Choristoma: Ultrasonography as a Diagnostic Tool

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Introduction

Choristoma is a rare benign congenital malformation consisting of normal growth of cells or tissue that is foreign to that body site [1]. Epidermal choristoma (EC) is ectopic epidermal tissue. It is a rare benign lesion on the tongue that presents as solitary or multiple brown macules or papules [2]. Recent publications suggest clinical follow-up considering its benign course [2]. We report a patient with an EC with a 5-year follow-up and whose diagnosis was supported by high-definition ultrasonography.

Case Presentation

A healthy 2-month-old boy consulted for three light-brown homogeneous macules on the right dorsal aspect of the tongue (Figure 1A). Congenital oral melanotic macules and epidermal choristoma were included in the differential diagnosis. After discussing management alternatives with the patient family, clinical observation was chosen. The macules grew proportionally to the child's development, becoming palpable and with surface changes at age of five (Figure 1B). High frequency and ultra-high frequency color Doppler ultrasound demonstrated submucosal thickening with multiple hypoechogenic oval structures, consistent with sebaceous glands, without hypervascularity (Figure 2, A-C). The findings were consistent with lingual EC.

Conclusions

The presence of sebaceous glands in the oral mucosa is not uncommon, but on the tongue is exceptional and is considered an EC [3]. Most cases present on the tongue in males before the age of 2 [2]. It is clinically characterized by one or multiple macules or papules unilaterally or in the midline dorsal tongue. Color varies from light to dark brown; therefore, it is usually misdiagnosed as a congenital melanotic macule [1,2]. The histopathological study shows the

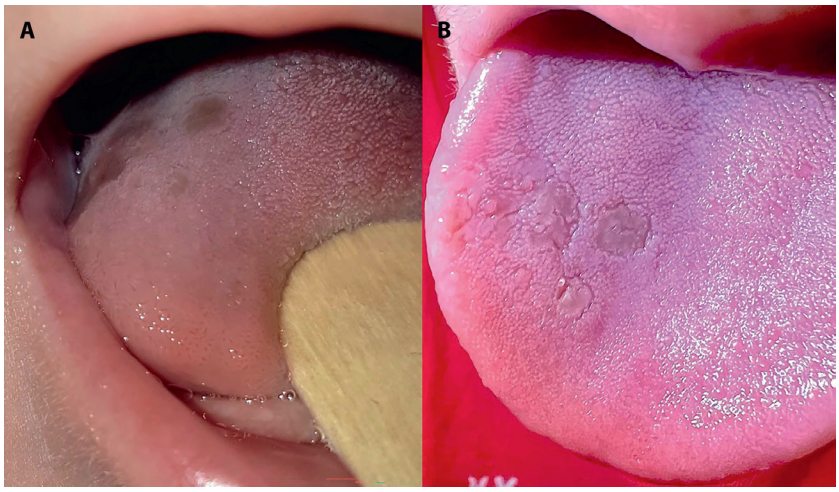

Figure 1. (A,B) (A) Three light-brown homogeneous macules on the dorsum of the tongue when the patient was 2 months old. (B) At the age of 5, 3 light-brown homogeneous now palpable lesions on the right dorsum of the tongue.

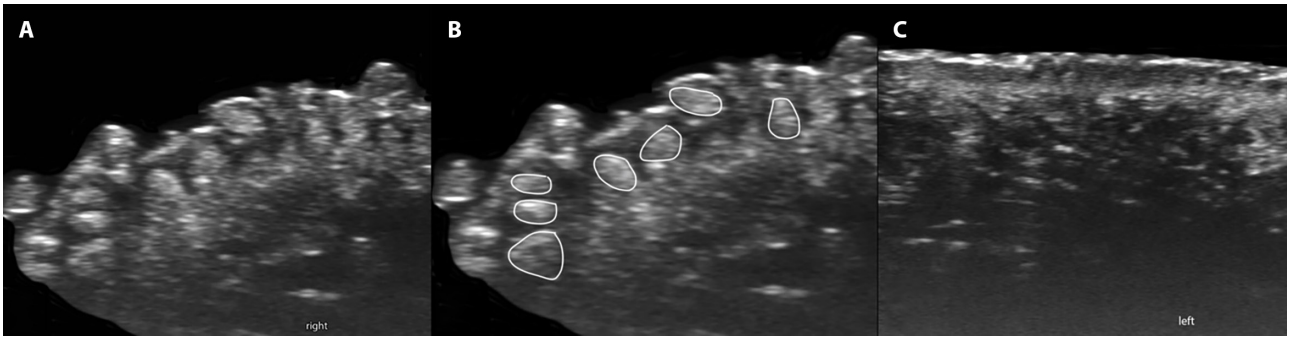

Figure 2. (A-C) Ultrasound at 70 MHz (Vevo MD; Visualsonics Fujifilm) of the tongue (transverse views) demonstrates multiple sebaceous glands as hyperechoic oval-shaped submucosal structures on the right side of the tongue (A) that were delineated with a white color (B). Notice the difference with the left side of the tongue (C) that does not present sebaceous glands.

replacement of normal oral mucosal epithelium by stratified squamous epithelium, hyperkeratosis, pigmentation of the basal layer, abortive hair follicles, and sebaceous glands [1].

In the case of congenital exclusively lingual pigmentation, most relevant differential diagnoses are congenital lingual melanotic macule (CLMM) and EC. Clinical clues to differentiate EC from CLMM are that EC usually becomes a palpable lesion over time, whereas CLMM usually presents as highly pigmented macules in dark skinned patients [4].

High and ultra-high frequency color Doppler ultrasound have gained relevance in skin and mucosal conditions and can be useful in children where an oral cavity biopsy procedure is complex. The axial spatial resolution of ultra-high frequency ultrasound at 70 MHz goes up to 30 microns, closer to the lower magnification of histology. The ultrasound pattern of sebaceous glands has been reported previously as oval-shaped hyperechoic structures that may be or are not attached to the hair follicles, as our case (Figure 2, A-C) [5].

The gold standard for the diagnosis of EC is histopathology. However, a tongue biopsy in a child usually requires sedation, with all the risks it implies, including 0.4%-17% [6] of acute complications and yet unknown long-term neurological side effects. For this reason, we propose that ultrasound and particularly ultra-high frequency ultrasound, may be an alternative for the diagnosis and monitoring of EC in cases where families and physicians decide to perform active clinical observation, especially considering that oral congenital melanoma has not been reported yet, the difficulty of oral biopsy in a neonate and that sedation under three years of age may carry risks.

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