The Congenital Lingual Melanotic Macule
Magdalene A. Dohil, MD; Glenn Billman, MD; Seth Pransky, MD; Lawrence F. Eichenfield, MD

Background: Circumscribed oral melanotic macules in adults are a well-defined entity. Congenital oral melanotic macules, however, are rare and not well characterized. We report 5 cases of congenital lingual melanotic lesions with consistent clinical and histologic features.

Observations: Congenital lingual melanotic macules presented in all cases at birth, followed by proportional growth. Clinical findings were well-circumscribed, brown, single or multiple macules on the tongue, 2 to 5 mm in size, without history of bleeding, ulceration, or trauma. Family history was negative for similar pigmented lesions. A literature review showed only 2 previous clinical case reports with similar features. Histopathologic examination showed increased melanin pigmentation in the basal epidermal layer with varying degrees of overlying hyperkeratosis and subepidermal pigment-laden macrophages. No appreciable increase in melanocyte number, junctional nests of melanocytes, or cell atypia was noted. Two retrospective histopathology reviews mention 4 corresponding cases but report no clinical data.

Conclusion: The congenital lingual melanotic macule represents a clinically distinct, benign, pigmented oral melanotic lesion, and may be more common than the literature suggests.

Arch Dermatol. 2003;139:767-770

HYPERPIGMENTED lesions involving the oral mucosa have various pathogenetic causes. The most common lesions are oral and labial melanotic macules.1,2 The oral melanotic macule presents in patients older than 40 years as a flat, blue, brown, or black, mostly solitary lesion less than 10 mm in diameter. It may also arise on the gingival, buccal, or palatal mucosa.1 The labial melanotic macule presents at an earlier mean age of 27.5 years, is more common in the female population, and is noted almost exclusively in whites. It usually manifests at the vermilion border of the lower lip near the midline, is mostly solitary, and rarely exceeds 5 mm in diameter. Family history may be positive, and it may occur as part of the Laugier-Hunziker syndrome.4,5

Histologically, both the oral melanotic macule and the labial melanotic macule are characterized by excess melanin in the basal cell layer and lamina propria. Some mild pigment incontinence may be present. However, a significantly increased number of melanocytes, elongated rete ridges as seen in lentigo simplex, and nuclear atypia as observed in malignant melanoma are not seen.6,7

The congenital melanotic macule of the tongue has rarely been reported, and its clinical features are less well defined. We studied 5 cases of lingual melanotic macules that were present at birth. Clinical presentation was recorded and punch biopsies were performed on each patient. Histopathologic changes in each patient were assessed in comparison with age-matched normal lingual tissue. We also present 2 previous clinical case reports and 4 histopathology cases from our review of the literature.

REPORT OF CASES

CASE 1
A 5-month-old white female infant presented with a pigmented lesion on the tongue that had been noted at birth and had grown proportionally with the child. The infant had not received any medication and was otherwise healthy. The family history was negative for melanoma, polyposis, and mucosal pigmentation. On examination, a solitary 3.0 x 3.1-mm black macule on the left anterior tongue surface was noted. The results of the rest of the physical examination were normal ex-

From Pediatric and Adolescent Dermatology (Drs Dohil and Eichenfield), Department of Pathology (Dr Billman), and Pediatric Otolaryngology (Dr Pransky), Children’s Hospital, San Diego, Calif; and Departments of Pediatrics and Medicine, University of California, San Diego, School of Medicine (Drs Dohil and Eichenfield). The authors have no relevant financial interest in this article.
except for a hemangioma on the left posterior part of the scalp, measuring 1.5 × 1.8 cm.

CASE 2

A flat, brown lesion of the tongue was noted at birth in a white female infant. Evaluation at 6 months of age showed a solitary, irregular, flat, hyperpigmented macule, measuring 3.5 × 3.0 mm on the midportion of the tongue. The history indicated proportional growth of the lesion since birth. No bleeding or ulceration was reported, and the family history was unremarkable.

CASE 3

A 2-month-old white male infant was seen for multiple dark-brown–pigmented macules on the dorsal left surface of the tongue that had been present since birth and were slowly increasing in size. The patient was followed up clinically, and proportional growth of the lesions during the next few months was noted. At 6 months of age, the lesions measured between 3.0 and 4.0 mm each, and a biopsy of 1 lesion was performed. Family history was unremarkable.

CASE 4

A 3-week-old Hispanic male newborn was seen for evaluation of multiple bluish black, irregular-shaped macules on the dorsal surface of the tongue that had been present since birth (Figure 1). Family history was unremarkable. At 4 months of age, a biopsy of 1 lesion was performed; further clinical follow-up showed only mild increase in size proportional to the child’s growth.

CASE 5

A 3-week-old Hispanic male newborn was examined because of 6 hyperpigmented, dark-brown, circular lesions on the tongue that had first been noted at birth. The macules measured 3 to 4 mm and were located on the dorsal mid and anterior portion of the tongue. Family history was unremarkable. At 2 months of age, no appreciable increase in size was noted, and 2 biopsy specimens were obtained.

RESULTS

Histopathologic evaluation in all 5 cases (Figure 2) showed increased basal layer melanin pigmentation with some scattered subepidermal pigment-laden macrophages and varying degrees of hyperkeratosis. Melanocyte number was normal. Rete ridges were only mildly elongated in cases 3 and 4. No junctional nests of melanocytes or atypical basilar melanocytes were appreciated. The margins of noninvolved lingual tissue were distinct and clearly delineated. No similar histopathologic changes were identifiable in age-matched normal lingual tissue.

Two previous case reports8,9 of congenital lingual melanocytic lesions that appear consistent with our cases were identified in the review of the literature. One report described a 12-year-old African American girl with 3 pigmented papules on the right dorsum of the tongue. The lesions had been present since birth and had grown proportionally since. On examination they measured 1 to 3 cm and were distinct in appearance from the child’s gingival and buccal physiologic mucosal melanosis. Histologic examination showed basilar hyperpigmentation with normal melanocyte number.8

Figure 1. Clinical appearance of multiple, macular, pigmented lesions present since birth on the tongue of a 4-month-old infant.

Figure 2. Lingual melanotic macule demonstrating diffuse hyperpigmentation of the basal zone, scattered melanophages in the lamina propria, and focal thickening of the granular zone and orthokeratosis (hematoxylin-eosin, original magnification ×40).
The other, more recent case report described a 3-day-old white male newborn with 3 congenital pigmented lesions on the left side of the dorsal aspect of the tongue. Histopathologic examination showed focal increase of melanin in the basal cells without associated findings.

Two extensive retrospective series of oral histopathologic studies included 4 cases of histologically verified melanotic macules with lingual location. No clinical data regarding age at onset, anatomic location, size, color, or growth were provided.

**Comment**

Oral mucocutaneous hyperpigmentation is not uncommon in clinical practice and may represent many conditions and diagnoses. These range from physiologic melanin pigmentation to systemic disease as well as drug-related pigmentation. Other causes include pigmented fungiform papillae, Laugier-Hunziker syndrome, and melanin pigmentation to systemic disease as well as drug-related pigmentation. The oral mucosa, often associated with melanin pigmentation, may be the site of pigment alteration usually fades with time and often includes a history of other dermatoses. Addison disease presents with diffuse pigmentation and evidence of systemic involvement. The pigmentation in Peutz-Jeghers syndrome is more speckled and extensive, with gastrointestinal polyposis manifesting later in life. Laugier-Hunziker syndrome results in acquired pigmentation of the oral mucosa, often associated with melanonychia. In pigmented fungiform papillae, the pigmentation is confined to these papillae. Pigmented lesions such as mucosal melanocytic nevi or melanomas are more difficult to distinguish, and a biopsy is often necessary.

In the past, terminology for oral melanotic macules of the oral mucosa and skin has been a source of confusion. Various terms have been used, such as *ephelide, melanosis, lentigo, labial lentigo, melanotic macule, and oral melanocytosis*. The work of Weathers et al in 1976 on labial melanotic macule and Page et al in 1977 on the oral melanotic macule helped to standardize current terminology characterizing the features of oral and labial melanotic macules, respectively.

We believe that the congenital lingual melanotic macule is a distinct entity. Although the small number of documented cases (7 patients) does not allow any definite conclusions, a clinical diagnosis of congenital lingual melanotic macule should be considered when the following criteria apply: solitary or multiple melanotic lesions on the tongue; presence at birth with subsequent proportional growth; and a negative family history for systemic conditions associated with mucosal pigmentation.

The consistent histologic features include increased basal pigmentation with varying degrees of overlying hyperkeratosis. No or only minimal increase in the amount of melanin in the melanocytes and very mild pigment incontinence can be found. These features should aid in the diagnosis of the melanotic macule. The absence of or only subtle elongation of the rete ridges and the normal number of melanocytes distinguishes it from a lentigo simplex. The lack of nesting of melanocytes, a negative melanosome antibody stain (HMB-45), and a lack of atypical cells aid in the differentiation from melanocytic nevi and melanoma, respectively.

Primary malignant melanoma of the oral cavity accounts for about 0.2% to 8% of all melanomas and shows a distinct predilection for the maxillary alveolar ridge and palate, whereas the tongue represents a rather unusual site. It is more commonly observed in the 40- to 70-year age group. Our literature review disclosed a single case of histologically documented transformation of benign oral melanosis into malignant melanoma. In this description of an adult, the patient originally presented with an ulcerated lesion, which would direct the diagnosis away from an oral melanotic macule. Furthermore, it is not clear that the biopsy was performed of the most atypical portion of the lesion on the original visit. Nevertheless, it illustrates that the distinction from oral melanoma with its extremely poor prognosis can be difficult, and a biopsy should be undertaken to secure the diagnosis should any clinical suspicion exist. We suggest that biopsy of pigmented tongue lesions include adjacent normal tissue, as this allows for an easier comparison of normal and lesional hyperpigmentation.

The cause of the congenital lingual melanotic macule is unclear. There are sporadic reports of acquired oral melanotic macules appearing after trauma, irradiation, or medication. Various hypotheses for localized increased melanin production in these cases have included physiologic genetic variations or viral and immunologic factors, but none has been conclusive yet. It is possible that the congenital lesions may represent a hamartoma of melanocytes with localized functional change in melanin production.

**Conclusions**

We believe that the congenital lingual melanotic macule represents a unique entity of oral and labial melanotic macules. It is distinguished by its appearance at birth, its location on the dorsal surface of the tongue, and its tendency toward proportional growth. The lesion shares the benign histologic features of other oral melanotic macules, and a biopsy is recommended to ascertain this. Encountering 5 cases at our medical center during 2 to 3 years leads us to believe that this condition is more common than the medical literature would suggest.

Accepted for publication September 24, 2002.

Corresponding author and reprints: Lawrence F. Eichenfield, MD, Pediatric and Adolescent Dermatology, 3030 Children’s Way, Suite 408, San Diego, CA 92123 (e-mail: Leichenfield@chsd.org).

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