

# Nevus of Ota/Oculodermal Melanocytosis: A Rare Report of an Oral Mucosal Lesion Involving the Hard Palate

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To the Editor:

Nevus of Ota, also known as oculodermal melanocytosis or nevus fuscoceruleus ophthalmomaxillaris, is a hamartoma of dermal melanocytes that is characterized by a unilateral or bilateral blue-brown, speckled patch usually involving the malar, periorbital, temple, and/or forehead regions of the face.<sup>1</sup> It also may affect the sclera, conjunctiva, retinas, corneas, ocular muscles, periosteum, and retrobulbar fat corresponding to the distribution of the ophthalmic (V1) and maxillary (V2) divisions of the trigeminal nerve.

Examination of the oral cavity in the setting of nevus of Ota is imperative, as it can present as a developmental lesion of the oral mucosa.<sup>2</sup> Involvement of the hard palate is rare but has been observed.<sup>3-5</sup> We present a case of blue-pigmented macules in the upper right periorbital region with involvement of the hard palate that were diagnosed as nevus of Ota.

A 34-year-old Indian man presented with progressive, asymptomatic, ashy blue macules in the upper right periorbital region that had been present since

birth. The pigmented macules had gradually increased to cover the infraorbital, maxillary, and temporal regions of the right side of the face with involvement of the conjunctiva and sclera (Figure 1).

Examination of the mucous membrane of the hard palate revealed several blue-pigmented macules with ill-defined borders merging into the surrounding mucosa (Figure 2). Ocular tension was normal and slit-lamp examination of the right eye did not reveal any abnormalities. Hematoxylin and eosin-stained sections prepared from a biopsy of the oral mucosa on the hard palate showed numerous elongated, fusiform, dendritic melanocytes in small aggregates scattered widely between the bundles of collagen in the papillary to midreticular dermis (Figure 3). On histology, the melanocytes stained positive for S100 protein (Figure 4) and human melanoma black 45. No evidence indicative of malignancy was found. The stratified squamous epithelium was unremarkable except for the presence of mild perivascular lymphocytic infiltrate in the subepithelial tissue. A diagnosis of nevus of Ota with involvement of the hard palate was made.

Cutaneous macules may enlarge slowly, become deeper in color, and persist throughout the patient's life. Its pathogenesis is not known, but it is speculated that nevus of Ota is caused by faulty migration of melanoblasts from the neural crest to the skin. Nevus of Ito also is a dermal melanocytic aberration that exclusively affects the shoulders and often occurs in association with nevus of Ota.<sup>1</sup>

Ashy or slate-blue pigmentation in individuals with skin of color (eg, Fitzpatrick skin type V) is uncommon, as this discoloration usually is seen in fair-skinned individuals (eg, Fitzpatrick skin type II).<sup>6</sup> Occasionally, blue-pigmented lesions of the oral mucosa may be seen in nevus of Ota (as in our patient) and are considered developmental; therefore, examination of the oral cavity is suggested

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The authors report no conflict of interest.

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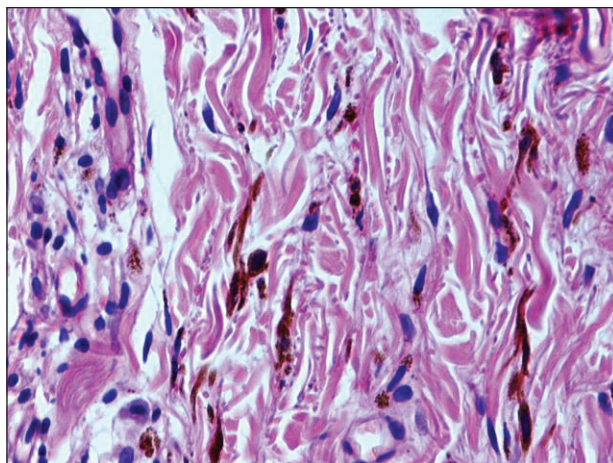
**Figure 1.** Progressive, asymptomatic, ashy blue pigmentation of the upper right periorbital region with involvement of the conjunctiva and sclera.



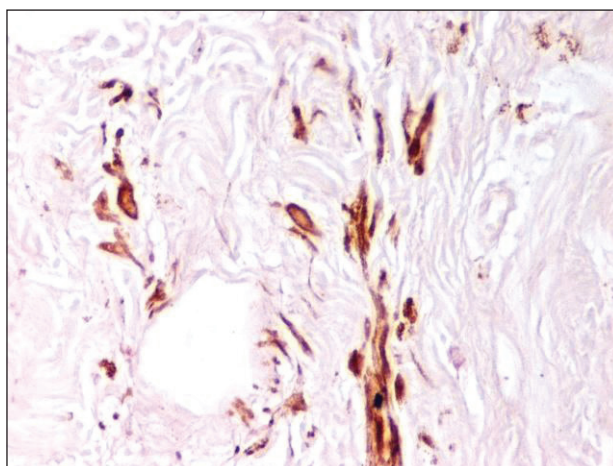
**Figure 2.** Blue macules on the hard palate.

when patients present with blue-pigmented lesions in the facial region. Although this finding is rare, several other cases of blue-pigmented macules on the palatal mucosa have been reported.<sup>3-5</sup>

The diagnosis of nevus of Ota should be confirmed by histopathology and can be classified into 5 types according to the distribution of melanocytes,



**Figure 3.** Numerous elongated, fusiform, dendritic melanocytes in small aggregates (H&E, original magnification  $\times 40$ ).



**Figure 4.** Melanocytes stained positive for S100 protein (original magnification  $\times 40$ ).

including (1) superficial, (2) superficial dominant, (3) diffuse, (4) deep dominant, and (5) deep.<sup>7</sup> The diagnosis of nevus of Ota can be made based on its characteristic morphology; however, nevus of Ito, Mongolian spots, melanoma, fixed drug eruptions,<sup>8</sup> and lichen planus pigmentosus should also be ruled out.<sup>9</sup>

Nevus of Ota is a well-established entity that should be considered when ashy or slate-blue pigmentation is noted along the branches of the ophthalmic and maxillary divisions of the trigeminal nerve. Diagnosis is largely clinical, but should be confirmed on histopathology and immunohistochemistry. Possible concomitant involvement of the

buccal mucosa and/or the hard palate warrants a thorough examination of the oral cavity in the setting of nevus of Ota to identify oral mucosal lesions. Histopathology is essential to confirm its status as well as to exclude melanoma.

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