Congenital melanocytic nevus of the oral mucosa: report of a rare pigmented lesion and review of the literature

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Abstract

Oral pigmented lesions are uncommon and congenital melanocytic nevi are especially rare. We report a case of a patient with multiple congenital melanocytic nevi including a palatal lesion. This is reported to add to the scant literature that exists on this subject. Prognosis and management are discussed.

Case Report

A 19-year-old woman was referred for assessment of a pigmented palatal lesion. She was a recent immigrant and was accompanied by her father who provided translation. This was first identified at three-months of age. There had been no pain or dysphonia but a bothersome sensation related to progressive surface roughness that prompted her to seek care. She was otherwise healthy; a small congenital melanocytic nevus affecting the right inner thigh had been excised during early childhood.

On examination, there was no palpable cervical lymphadenopathy. A well-demarcated, circular plaque (1.2×1.1×0.3 cm) with a slight pebbled texture affected the left posterior hard palate. While predominantly erythematous, there was scattered light brown pigmentation (Figure 1). No osseous abnormalities were appreciated on plane radiographic films. Benign and malignant melanocytic neoplasms were considered in the differential diagnosis. An incisional biopsy showed papillomatous squamous epithelium that was otherwise unremarkable. The underlying stroma contained sheets of nevus cells (Figure 2) exhibiting maturation and extending deep into the lamina propria and around minor salivary ducts (Figure 3), nerves and blood vessels. There were scattered nevus giant cells and scant superficial melanin (Figure 4). There was no atypia, necrosis or mitotic activity. Cells diffusely stained with MelanA (Figure 5) and Ki-67 suggested a low proliferative index. Only the superficial cells stained with HMB-45 (Figure 6). Clinical-pathologic correlation was consis...
nevus cells with HMB45.

Figure 6. Scant staining of only the superficial nevus cells with HMB45.

tent with an intraoral congenital melanocytic nevus. An excisional biopsy was performed and the histologic features were similar. The patient failed to return for post-operative assessment.

Discussion

Melanocytic nevi are separated into acquired and congenital subtypes with the latter identified at birth. The palate is not easily visualized, however, which may explain delayed identification. While it is recognized that parental history may not be accurate, there was certainty of identification during early infancy. Most acquired nevi, in contrast, develop throughout childhood and in younger adulthood.

Small congenital melanocytic nevi may be clinically indistinguishable from acquired nevi. The larger size was suggestive of a congenital nevus as acquired nevi are usually less than 6mm in diameter. Congenital nevi may have a papular, pebbly or verrucous surface as well as hypopigmented areas. Histologically, a congenital pattern was characterized by a diffuse infiltration of nevus cells extending around blood vessels, nerves, salivary ducts as well as between collagen bundles.

Nevus cells may exist and proliferation in diverse anatomic locations as evidenced by identification of aggregates in the parenchyma of lymph nodes. Oral melanocytic nevi are uncommon, and to the best of these authors knowledge, only three well-documented cases of intraoral congenital melanocytic nevi have been reported in the English literature,1,4,5 (Table 1). Other mucosal sites are also uncommonly affected with rare reports of conjunctival or genital congenital nevi.6,7

Classification of congenital melanocytic nevi is by size: small (<1.5 cm), medium (1.5 to 19.9 cm) and large or giant (>20 cm). The primary concern is malignant transformation and there is general agreement that the risk increases with the size.8 While a clear risk exists for larger lesions, such an association is controversial with small congenital nevi. Rhodes et al. identified histologic features of congenital nevi in 8.1% of melanoma specimens.9 In another study, a melanoma risk of 2.6-4.9% was estimated for persons with small congenital nevi and it was concluded that small congenital nevi may represent precursors for at least some cases of cutaneous melanoma.10

Diagnostic biopsy of any oral melanocytic lesion is generally warranted to exclude melanoma. There is general agreement that the larger the congenital nevi the more likely the need for excision.4 For small congenital nevi, management may be either excision or observation but there exists no clear consensus or guidelines. Additionally, there is no evidence of recurrence.11 Excision seems reasonable as in most cases one would expect minimal surgical morbidity and excision facilitates comprehensive histologic sampling to exclude melanoma, and presumptively, may prevent malignant transformation. From the patient’s perspective, as was the case with this patient failing to attend post-surgical assessment, excision may potentially mitigate the necessity for long-term observation. Ultimately, decisions require individualization with consideration of the size, anatomic restrictions as well as patient factors such as co-morbidities and ability to attend observation.

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