Intraoral Manifestations in a Patient with Epidermal Nevus Syndrome

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ABSTRACT

Aim: To report rare findings of oral and periodontal manifestations in a patient with Epidermal nevus syndrome (ENS).

Background: The ENS describes the rare association of an epidermal nevus with abnormalities of central nervous system, ocular and skeletal abnormalities. Reports of oral involvement have been few. Also, most of the intraoral lesions have been reported in patients with nevi that do not fulfill the criteria for the diagnosis of ENS.

Case description: This report describes a case of ENS that, in addition to cutaneous manifestations showed skeletal involvement and intraoral manifestations such as the extension of the nevi on the face intraorally involving the labial mucosa, hypoplasia, hypodontia of teeth and severe periodontal destruction.

Conclusion: Patients with extensive epidermal nevi and systemic abnormalities should be suspected of having the ENS. Evaluation and management of patients with ENS requires a multidisciplinary team approach involving the dermatologist, pediatrician, ophthalmologist, neurologist, genetist, plastic surgeon and orthopedic services. Although uncommonly described in association with ENS, significant intraoral lesions do occur. Periodontal manifestations as in our patient, which to our knowledge has not been described in association with ENS so far, may also be present.

Clinical relevance: Alteration of the response of periodontal tissues to dental plaque in the presence of certain systemic diseases has been reported, but not in association with ENS. Severe periodontal destruction due to exaggerated response to dental plaque was seen in the present case. Hence, emphasis on oral hygiene maintenance in such patients is essential. Patients with ENS must be evaluated periodically as they show a persistent predisposition for the development of tumors.

Keywords: Facial nevi, Oral mucosal lesions, Periodontal manifestations, Hypoplasia, Case report.


Source of support: Nil

Conflict of interest: None declared

INTRODUCTION

The epidermal nevus syndromes (ENSs) represent a group of distinct disorders that comprise a clinically variable group of mosaic skin disorders, mainly characterized by extensive/atypical epidermal nevus with additional cutaneous and extracutaneous features. ENSs may be distinguished by the type of associated epidermal nevus and by the criterion of presence or absence of heritability.1

The most common oral manifestations reported are localized or diffuse verrucous growths on the lips, palate, gingiva, buccal mucosa and tongue.2 Other findings include hypertrophy of the tongue, cleft palate, high arched palate, bifid uvula3 and various dental anomalies, such as hypodontia, unerupted teeth, malformed teeth and odontodysplasia.2

Linear epidermal nevus (LEN), also known as verrucous epidermal nevus or linear verrucous epidermal nevus, presents clinically as tan or brown verrucous papules, arranged in a linear configuration. LEN are found at any cutaneous site, but tend to occur along the long axis of an extremity or across the trunk.4 We report a case of LEN which in addition to cutaneous manifestations, showed extension of the epidermal nevi intraorally, with mucosal changes, hypodontia, hypoplasia and severe periodontal destruction.

CASE DESCRIPTION

A female patient aged 17 years reported with a chief complaint of redness of the gums since 5 to 6 months. She presented with multiple, verrucous and linearly arranged pigmented lesions on the right side of the body involving the face, the neck and arm which were demarcated in the midline. She was the 2nd child of a healthy unrelated couple without family history of bone or skin disease. She was the product of an uncomplicated term pregnancy and born by vaginal delivery.

A large pigmented skin lesion was found on the right cheek extending to the ala of the nose. In addition, multiple
streaky verrucous plaques were seen involving the right side of the face, ear, postauricularly extending to the occipital region and the neck (Fig. 1). The lesions were present at birth and became more verrucous and darker since then.

Satellite lesions were found on the forehead, lower lip, and the philtrum which extended intraorally, involving the labial mucosa. Hypoplasia of maxillary right canine and mandibular right central incisor and areas of intense desquamation extending up to the alveolar mucosa were found (Fig. 2). Generalized inflammatory enlargement was seen, the findings being marked in the upper anterior region (Fig. 2). Panoramic radiograph showed generalized horizontal bone loss with greater destruction on the right side, and missing right third molars (Fig. 3). In addition, early exfoliation of mandibular right second molar owing to periodontitis was reported.

Apart from cutaneous and oral findings, a 2 cm shortening of the right lower limb was also seen. A biopsy specimen from the preauricular area on the right side of the face showed papillomatosis, acanthosis, hyperkeratosis and presence of sebaceous glands consistent with the diagnosis of ENS (Fig. 4). Based on the characteristic cutaneous findings associated with skeletal involvement and the histopathologic findings, a diagnosis of linear ENS was made. Since, the patient did not want any intervention for the nevi close follow-up was advised.

**DISCUSSION**

By definition, all ENSs reflect mosaicism and to date, nine well defined ENSs have been described which can be distinguished by clinical, histopathologic and genetic criteria. Nevi associated with ENSs follow the lines of Blaschko, a notable exception to which is the CHILD syndrome, in which lateralization pattern reflects the functional origin of the postzygotic modulation of the inherited mutation. Nevus sebaceous occurring as a part of nevus sebaceous syndrome may be differentiated histologically, by the presence of hyperplastic sebaceous glands. Coexistence of nevus sebaceous and nevus philus may be seen in phacomatosis pigmentokeratotica. Ipsilateral cataract is the diagnostic clue in nevus comedonicus syndrome, whereas linear nevus covered with soft white hair distinguishes Angora hair nevus from other epidermal nevi (EN). The Becker nevus, in contrast to other EN, does not follow the lines of Blaschko but are instead arranged in a checkerboard pattern. Proteus syndrome can be differentiated by the presence of nevi which are soft, velvety, and flat presenting with cerebroplantar hyperplasia and asymmetrical macrodactyly.

Many inherited and acquired diseases of the skin or mucosa including inflammatory linear verrucous nevus, incontinentia pigmenti, lichen striatus, linear lichen planus, and linear lupus erythematosus may also manifest themselves in a linear configuration, and hence differentiation from these diseases has to be done.
Among the osseous abnormalities, cranial and facial bone deformities have been observed in as many as one-third of patients with ENS. Our patient showed focal bony destruction of the mandible just beneath the nevi on the face. Accordingly, it has been proposed that such bony defects reflect a postzygotic, intercellular paracrine effect from the skin lesions.

To date, reports describing intraoral anomalies associated with ENS are few. Historically, since the first review of Brown and Gorlin in 1960, who reported 24 patients with oral and cutaneous LEN, only 11 patients presented with oral involvement. Another recent review reported five cases of LEN, with intraoral involvement, but none of these cases fulfilled the criteria for the diagnosis of ENS. The most frequently affected intraoral sites were found to be the lips, tongue and the palate. In addition, these two reviews reported occurrence of dental abnormalities which included enamel hypoplasia and hypodontia, affecting around 20% of the reported cases.

Hypoplasia of the maxillary right canine was seen in our patient. Two other case reports describe intraoral manifestations involving the canine and showing oral mucosal lesions contiguous to facial nevus, as in the present case, but in association with nevus sebaceous syndrome. It has been suggested that the lines of Blaschko of the external skin are connected to lines extending onto the oral mucosa and therefore, cells from the cutaneous and intraoral regions of the linear nevi, such as ours, represent the same genetic mosaicism.

Enlargement of the gingiva is a common feature of gingival disease causing an aesthetic problem and creates inaccessible areas for the patient to deliver proper oral hygiene. Magnification of an existing inflammation initiated by the presence of dental plaque by the systemic condition of the patient has been reported. This exaggerated response of the gingiva to dental plaque may have resulted in areas of intense erythema and marked enlargement with the extent and severity being greater on the right side, which is also the side affected by the nevus.

CONCLUSION
Patients with extensive epidermal nevi and systemic abnormalities should be suspected of having the ENS. Evaluation and management of patients with ENS requires a multidisciplinary team approach involving the dermatologist, pediatrician, ophthalmologist, neurologist, genetist, plastic surgeon and orthopedic services. Although uncommonly described in association with ENS, significant intraoral lesions do occur. Periodontal manifestations as in our patient, which to our knowledge has not been described in association with ENS so far, may also be present.

CLINICAL SIGNIFICANCE
Alteration of the response of periodontal tissues to dental plaque in the presence of certain systemic diseases has been reported, but not in association with ENS. Severe periodontal destruction due to exaggerated response to dental plaque was seen in the present case. Hence, emphasis on oral hygiene maintenance in such patients is essential. Patients with ENS must be evaluated periodically as they show a persistent predisposition for the development of tumors.

ACKNOWLEDGMENT
We thank Dr Rudolf Happle, Department of Dermatology, University of Marburg, Germany for his analysis of the case.

REFERENCES

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