Case Report

Inflammatory linear verrucous epidermal nevus and regional odontodysplasia: A rare sorority

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Abstract

Epidermal nevi are hamartomatous lesion and its association with other developmental defects particularly of the central nervous system, eye and skeletal system are well recognized. We report a rare case of inflammatory linear verrucous epidermal nevus syndrome along with regional odontodysplasia; and to the best of our knowledge this is the second case reported in the literature.

Key words: Epidermal nevi, neuroectodermal disorder, regional odontodysplasia

INTRODUCTION

Epidermal nevus syndrome (ENS) is a group of neuro-ectodermic disorder characterized by epidermal nevi in association with deformities and dysplasia of the skin, eyes, brain, skeleton, and heart. Inflammatory linear verrucous epidermal nevus (ILVEN) is a rare variant of this syndrome. It is characterized by verrucous papules and plaques that are distributed in a linear pattern following Blaschko's lines, skin tension lines (purported embryonic lines of ectodermal cleavage).[1] Epidermal nevi can be classified into other variants according to the predominant epidermal structure involved. Additionally, they are also classified according to their clinical appearance, distribution, and the extent of involvement.[2] Based on the extent of the involvement they are either classified as localized or diffuse. Localized lesions that are confined unilaterally called as nevus unius lateralis whereas extensive bilateral involvement called as ichthyosis hystrix. Other epidermal nevus variants distinguished by the epidermal structure involved include nevus sebaceus, nevus comedonicus, eccrine nevus, Becker's nevus, and white sponge nevus.[3] In addition, they are associated with central nervous system and skeletal abnormalities but its association with oral mucosal lesions has been rarely reported.[1]

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We report a rare case of ILVEN syndrome along with regional odontodysplasia (ROD); and to the best of our knowledge this is the second case reported in the literature.

CASE REPORT

An 8-year-old female patient reported to the Department of Oral Medicine and Radiology with the complaint of discolored teeth in the upper right and left region back teeth region since 6 months. The past medical and family history of the patient was not contributory. The general examination of the patient was normal, and the review of systems was negative. She presented with dark brown papules arranged in a linear configuration, limited to the left upper part of her body, involving neck, axilla, chest, back, shoulder, and left arm on which the lesion extended [Figure 1a]. Lesions present on the nape of the neck [Figure 1b] were thick plaque like in appearance while on axilla, chest, and back were dark and mossy in appearance. On the face, verrucous papules were present on the left side, involving the forehead, external ear, and preauricular region and extending to the right cheek [Figure 1c]. Examination of the right and left eye revealed prominent vascular markings along with tan colored nevi in bulbar conjunctiva [Figure 2].

On intra oral examination, the maxillary right and left deciduous second molar exhibited yellowish brown

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color with severely malformed crowns and with evidence of caries [Figure 3]. The soft tissues showed normal color and texture. On intra oral periapical radiographs [Figure 4], the affected teeth showed hypoplastic and hypomineralized enamel along with enlarged pulp chamber and root canals, in comparison to the rest of the dentition. In addition, no radiographic distinction between the enamel and dentin could be appreciated.

Panoramic radiograph [Figure 5] showed the ghostly appearance of the maxillary right and left deciduous second molar. The radiodensity of both the enamel and dentin was not discernible with enlarged pulp chambers. These alterations were compatible with ROD.

On the basis of unilateral verrucous plaque distributed on the upper extremities and the ghost teeth involving the primary maxillary molars, a diagnosis of ILVEN with ROD was given.

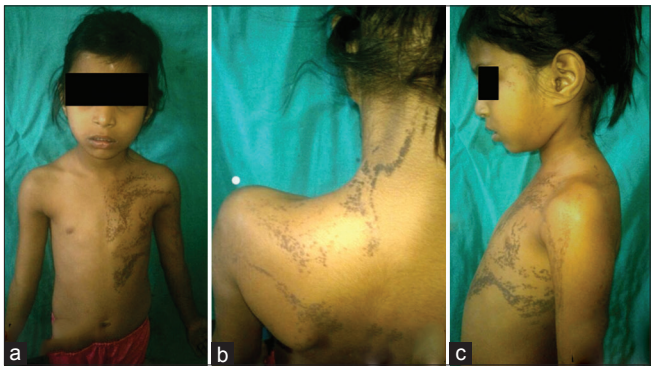

Figure 1: (a) Frontal view of the patient showing dark brown papules limited to the left upper extremity of the body, (b) Nape of the neck showing thick plaque like lesions, (c) Lateral view of the patient presented with thick verrucous papules distributed on the face

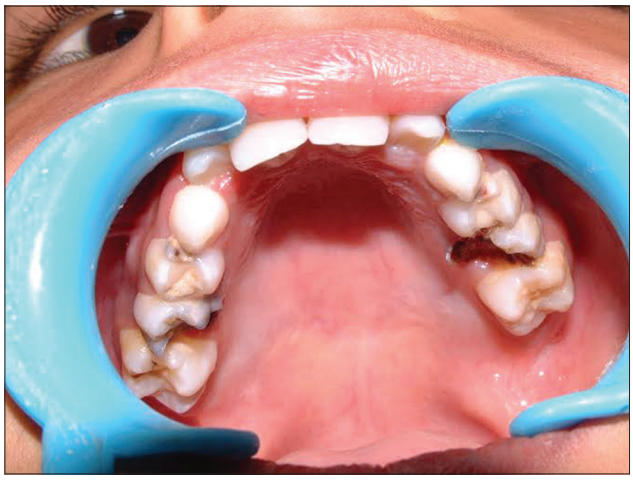

Figure 3: Intraoral photograph showing yellowish brown color with severely malformed crowns of the maxillary right and left deciduous second molar

DISCUSSION

Inflammatory linear verrucous epidermal nevus is a variant of verrucous epidermal nevus and was originally described by Altman and Mehregan.[4] In the review by Rogers,[2] in 233 cases the prevalence of ILVEN was around 6%. He found that the distribution was almost exclusively on the lower half of the body, with the buttock being the most frequently involved area. Altman and Mehregan[4] also showed that the distribution of ILVEN was predominantly on the lower extremities. Only 16% of lesions were found on the upper half of the body, including the axillae, arm, and hand. While in our case, thick verrucous plaques predominantly present on the upper half of the body.

Clinically, the lesions in these patients have been described as unilateral or midline papules or nodules with a papillary or verrucous surface. These lesions are found on one side of the body, and characteristically they do not cross the midline. The most common location of these lesions is on the lips, tongue, buccal mucosa, hard and soft palate and gingiva.[1] However, in our case no intraoral lesions could be appreciated.

Inflammatory linear verrucous epidermal nevus is caused by somatic mutations that result in genetic mosaicism

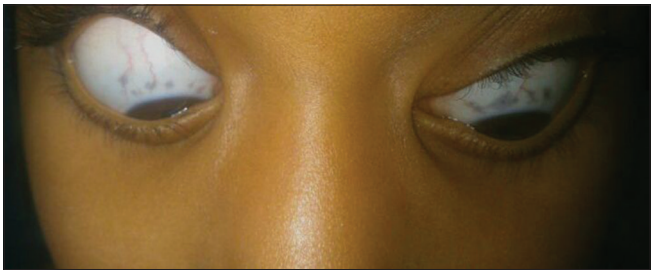

Figure 2: Right and left eye showing prominent vascular markings along with tan colored nevi in bulbar conjunctiva

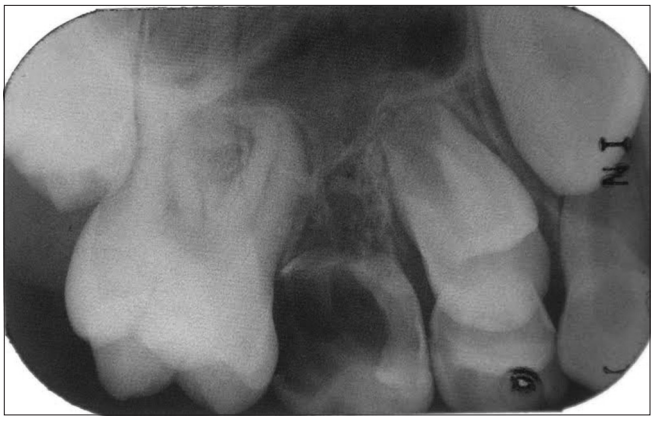

Figure 4: Intraoral periapical radiographs of maxillary right second deciduous molar showing the hypoplastic and hypomineralized crown with wide pulp chamber and root canal

and although its physiopathology is still unclear, it is believed that it may be associated with an increase in the production of interleukins-1 and 6, tumor necrosis factor-alpha, and intercellular adhesion molecule.[1]

Solomon and Esterly[5] provided an extensive account of the extracutaneous findings in patients with epidermal nevi, thus classifying as having ENS. Ophthalmologic anomalies are quite common in such patients. These include extension of nevi to lid and bulbar conjunctiva, lipodermoids, colobomas, corneal opacity, nystagmus and cortical blindness.[6] In our case, prominent vascular markings and nevus on the bulbar conjunctiva could be appreciated in both the eyes.

Skeletal changes associated with ILVEN are incomplete development of various bones, vertebral defects, camptodactyly, clinodactyly, abnormal clavicles, asymmetry of ribs, shortening of limb bones, posterior luxation of ankles, and asymmetry of the skull, hemihypertrophy, kyphoscoliosis, ankle/foot deformities, and Vitamin D resistant rickets.[6,7]

Neurologic involvement is estimated to occur in around 50–70% patients with ILVEN syndrome. The most common abnormalities are mental retardation and seizures. Other neurologic manifestations include hypotonia, hyperkinesia, hemiplegia, hemiparesis, cranial nerve palsies, ventricular abnormalities, cortical lesions, and intracerebral calcification.[8,9] In our case, neurologic and skeletal involvements were not detected.

Intra oral involvement of ILVEN is a rare manifestation. Oral findings in patients with ENS have been summarized by Gorlin *et al*.[9] These consist of nevoid alterations of the oral mucosa, cleft palate, bifid uvula, and dental abnormalities consisting of hypoplasia or odontodysplasia. A case of ROD in ENS was reported by Slootweg and Meuwissen, 1985.[10]

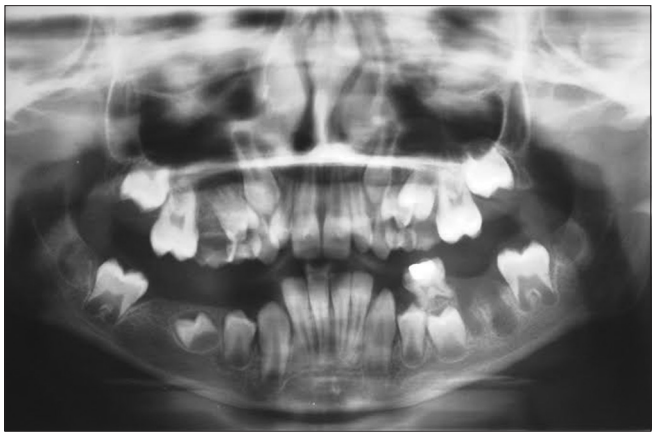

Figure 5: Panoramic radiograph of the patient showing the ghostly appearance of the maxillary right and left deciduous second molar

Table 1 enumerates the various skeletal/dental abnormalities associated with ILVEN that is, reported in the literature.

In an exhaustive review of ROD by Crawford and Aldred,[17] it was stated that females are more often affected than males (1.4:1) which was consistent with our case. These authors enumerated several etiological factors such as local circulatory disorders, viral infections, pharmacotherapy during pregnancy, facial asymmetry, local trauma, metabolic disturbances, somatic and neural mutations, and syndromal involvement. While in our case, the most appropriate etiology that can be thought of is syndromal involvement (ILVEN). It has been postulated that tooth morphogenesis depends on the presence of neural crest cells that are called as ectomesenchyme and these cells also contribute to dermal and epidermal structures. Therefore, it is possible that the co-existence of tooth malformation and nevoid alterations in the overlying skin, as were noted in the present case, may be explained by a local failure in the corresponding area of the neural crest to migrate and differentiate normally.[18]

The diagnostic criteria for ROD are based on distinctive clinical and radiographic findings. The maxilla is affected twice as often as mandible. As seen in our case, deciduous maxillary second molars were affected. Usually, clinically teeth appeared to be small, brown, grooved, and hypoplastic. While in our case, the affected teeth exhibited yellowish brown color with severely malformed crowns and with no evidence of caries.[14]

Radiographically, the enamel and dentin was not discernible, both of which are less radiopaque than unaffected counterparts. In addition, enamel and dentin layers are thin, giving the teeth a "ghost like" appearance.[19] These radiographic findings were favorable in our case. Since the clinical and radiographic features were distinctive of ROD so a straightforward diagnosis was made.

Table 1: Enumerates the various skeletal/dental abnormalities associated with ILVEN as reported in literature

Authors	Year	Skeletal/dental abnormality associated with ILVEN
Kelley *et al*.[11]	1972	Enamel hypoplasia
Muller *et al*.[12]	1980	Facial hemihypertrophy
Baghaei-Rad *et al*.[13]	1982	Compound odontomas
Slootweg and Meuwissen[10]	1985	Regional odotodysplasia
Kaplan *et al*.[14]	1993	Giant cell granuloma
Basopoulou-Kyrkanidou[15]	2000	Ameloblastoma
Kumar *et al*.[16]	2012	Enamel hypoplasia

ILVEN: Inflammatory linear verrucous epidermal nevus

In the present case, the diagnosis of ILVEN along with ROD is thus justified by following salient features that were present in our patient: Female patient, presence of thick verrucous plaque involving the upper extremities, early onset of disease, ghostly appearance of teeth in radiographs and concomitant abnormalities in both eyes and dentition.

This case highlights the importance of meticulous examination of children with epidermal nevi and confederation of neurologic, ophthalmologic, and orthopedic specialties for piloting the management of such patients. To the best of our knowledge and belief, our case was a unique and second case in the literature in which was a simultaneous coalition of ILVEN with ROD.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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