

Oral manifestations of Linear Sebaceous Nevus Syndrome: A case report and review of literature

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ABSTRACT

A sebaceous nevus is an unusual kind of birthmark. It frequently involves scalp, but sebaceous nevi may also emerge on the neck, face or forehead. It comprises of the overgrown epidermis (upper skin layer), sebaceous glands, hair follicles, apocrine glands and connective tissue. Linear sebaceous nevus syndrome rarely reveals any oral manifestation. A sebaceous nevus is also called an organoid nevus because it may comprise components of the entire skin. We report a case of linear sebaceous nevus syndrome in a 14-year-old female patient with abnormal oral findings.

KEYWORDS: Sebaceous Nevus, Schimmelpenning Syndrome, Pruritic Papules, Enamel Defect, Nodular Swelling Of tongue

INTRODUCTION

Sebaceous nevi is a variety of epidermal nevi classified under the headings of benign tumor of the hair follicle that consists of the overgrown epidermis (upper skin layer), sebaceous glands, hair follicles, apocrine glands and connective tissue. It is usually found at birth, can appear anytime during childhood and may scarcely appear in adulthood.¹ Sebaceous nevus and Sebaceous nevus syndrome (Schimmelpenning syndrome) are believed to be originated by postzygotic mosaic mutations in the HRAS or KRAS genes. RAS stimulate cell growth through activation of various pathways, embracing the mitogen-activated protein kinase (MAPK) signaling pathway.^{2,3,4} It is most commonly present on the scalp, but sebaceous nevi may also arise on the face, neck or forehead. It normally presents as a solitary or multiple lesions of the skin. The lesion can form large hairless plaques, somewhat elevated, confined, with a soft velvety surface, yellowish in white individuals and hyperpigmented in black individuals.

Linear sebaceous syndrome presenting with oral manifestation is rarely reported case. Oral manifestation of the linear sebaceous syndrome is very sparsely discussed the topic in literature with not much of information available. Here we report a case of linear sebaceous nevus syndrome presenting with multiple oral manifestations in a 14-year-old female patient. Furthermore, there is very less literature on such kind of oral findings in Linear Nevus Sebaceous Syndrome (LNSS), and we, therefore, consider this report to be interesting and informative in this respect.

CASE REPORT

A 14-year-old female patient reported to the department

of oral medicine and radiology with the complaint of poor chewing ability and poor esthetics due to malaligned teeth. No relevant family history was present. The lesion was present since birth and increase in size than before. The patient presented with the history of seizures and was on valproate 300 mg B.D. for the same. The patient had some intellectual disability.

Extraoral examination revealed presence of dark brown pruritic papules involving midline and both sides of the face specified as follows: forehead, bridge of the nose and chin, right side of forehead along the right lateral half of the nose, right nasolabial region, left ear lobule, left side of face along the left rail region, left side if neck and midline of neck (Figure 1 and 2). No such papular lesions were detected at any other site on the body. Cervical lymph nodes were not palpable



Figure 1 Frontal view of patient showing dark black papules having linear distribution limited to the right side forehead region, mid forehead region inner canthus of right eye, bridge of nose, symphysis region and midline neck region.

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Figure 2: Lateral view of patient showing dark black papules having linear distribution in front of left external acoustic meatus runs linearly downward involving left ear lobe and extended to the lateral side of neck on left side.

Intraoral examination revealed three firm nontender nodular swelling of size approx 1 cm in diameter noted on the dorsum surface of tongue in the midline and the left lateral half region. Black pigmentation was noted on the tip of tongue on the right side (figure 3), enamel defect as stained vertical ridges on labial aspect of 21 (figure 4) and root stump in relation to 46., linear mucosal depression seen in the anterior region of the hard palate running from the interdental region of 22 and 23 (figure 5) and poor dental esthetics due to crowding with respect to maxillary and mandibular dental arch, constricted maxillary arch with high arch palate and lingually erupted 12 and 22. Localized gigantism wrt 21 with increased mesiodistal diameter (figure 5).



Figure 3: Intraoral view shows nodular swellings on tongue and melanin pigmentation on tip of tongue on right side.



Figure 4: Showing enamel defect as stained vertical ridges on labial aspect of 21 and gingival inflammation.



Figure 5: Showing lingually erupted 12 and 22 and constricted maxillary arch.

Gingival inflammation, heavy plaque, and calculus were evident.

OPG reveals that missing 12, 32 and 41, a well defined radio-opacity of radiodensity similar to that of tooth squarish in shape size approximately 1cmx2cm is partly overlying the 22 with intermittent linear radiolucent striations running longitudinally seen in the 21 regions, at the coronal level extending into the alveolar bone in linear form. Mucosal thickening with respect to maxillary sinus (figure 6). CBCT revealed blunderbuss canal wrt 21 with the wide open apex (figure 7).



Fig 6

Figure 6: OPG reveals that missing 12, 32 and 41. Well defined radio opacity of radio density similar to that of tooth squarish in shape size approximately 1cmx2cm partly overlying the 22 with intermittent linear radiolucent striations running longitudinally seen in the 21 region, at coronal level extending into the alveolar bone in linear form. Mucosal thickening in respect to maxillary sinus.



Figure 7: CBCT views revealed blunderbuss canal wrt 21 with wide open apex . Figure 7a: Coronal view, Figure 7b: Sagittal view

After looked out at medical history, clinical findings and radiographical examination provisionally diagnosis was given as Linear Nevus Sebaceous Syndrome (LNSS) with differential diagnoses of Becker nevus syndrome, Nevus comedones syndrome, Phakomatosis pigment keratotic, Proteus syndrome and CHILD syndrome.

Biopsy of scalp patch shows hyperkeratosis, acanthosis, proliferated sebaceous glands and hypermelanosis. It confirmed the diagnosis of Linear Sebaceous Nevus.

So on the basis of history of seizure, intellectual disability and histopathologically confirmed linear sebaceous nevus we reached to the confirmatory diagnosis of Linear Sebaceous Nevus Syndrome.

DISCUSSION

Linear nevus sebaceous syndrome is a rare condition with the prevalence of 1:10,000 in the population varies with case reports. The incidence of sebaceous nevi estimated at 1 in 1000 new birth. It mainly affects the eyes, central nervous system, and skin presenting with symptoms like seizures, intellectual disability, congenital hairless plaque that is usually found on the scalp, face, or neck, hemimecephaly, ophthalmologic deviations, skeletal peculiarities, heart flaws, and an increased chances for the development of cancer.⁵

The nodular and papular lesions were reported unilaterally on the labial mucosa, tongue, buccal mucosa, palatal mucosa and also on the gingiva with most of the cases presenting on the left side of the oral cavity.^{6,7} Teeth were mostly presented with hypoplasia, and few of the reported cases had odontodysplasia.

The present case shows classical extra-oral feature of linear sebaceous nevus syndrome like hairless plaque on the face and neck and with rare oral manifestations like nodular growth on the midline and left dorsum of the tongue, constricted maxillary arch, high arch palate, odontodysplasia wrt 21 22, crowding with teeth, submucosal partial cleft of the anterior hard palate, melanotic pigmentation on tip of the tongue (Table-1).

SITE	FINDINGS
Tongue	Three firm non tender nodular swelling of size approx 1 cm in diameter noted on the dorsum surface of tongue in the midline and left lateral half region Black pigmentation was noted on the tip of tongue on the right side
Palatal mucosa	Linear mucosal depression seen on the anterior region of the hard palate running from the interdental region of 22 and 23
Gingiva	Inflammation was present
Maxillary arch	Crowding Constricted narrow V- shape arch
Palate	High arch palate
Tooth	Odontodysplasia irt 21
Occlusion	Malocclusion with anterior deep bite

Table-1 Oral manifestations

While the terms “linear nevus sebaceous syndrome” and “epidermal nevus syndrome” are frequently used conversely, epidermal nevus syndrome present with the different type of epidermal nevi with the involvement of another organ. Patients of LNSS surely have midline nevus, and for that nevus, there are three dermatologic stages. In stage 1, enduring since birth to puberty, small hairless lesion, and may decrease in dimension. Stage 2 occurs throughout puberty, and is identified by enlargement of the gland and the epidermis becoming verrucous. During stage 3, there is the possible malignant

conversion of the lesion, with the most common pathology being basal cell epithelioma. Altman and Mehregan in 1971 suggested classical clinical criteria for the diagnosis of LNSS⁸ and later modified by Morag and Metzker in 1985⁹ namely early age arrival, females predilection, commonly left leg involvement, pruritus, typical psoriasiform presentation, and marked unresponsiveness to therapy

The histopathologic examination of epidermal nevi showed moderate hyperkeratosis, increased thickness of stratum spinosum, warty appearance in the psoriasiform pattern, and neutrophils infiltration of dermis or even Munro's abscesses.¹⁰ All the features makes a clinical picture of a raised, papillary lesion. Occasionally, hypermelanosis in the basal cell layer is also evident. This causes lesions tan or brown-colored clinically. Additionally, on microscopic examination the margins of the lesion can be clearly differentiated from the nearby normal epithelium.^{11, 12} the chances of these lesions to convert into malignancy are approximately 0-22%.

Treatment plan for LNSS require teamwork i.e. multidisciplinary team approach involve different medical and dental specialties viz. dermatologist, pediatrician, ophthalmologist, neurologist, plastic surgeon, orthopedic surgeon, orthodontist and pedodontist.¹³

Many therapies have been tested for the epidermal nevi including topical steroids, topical and systemic retinoids as well as intralesional retinoids, topical 5-fluorouracil, podophyllin, dermabrasion, cryosurgery, and excisional surgery. However, these treatment options cause either partial elimination of the nevus or result in an intolerable scar that can be as unaesthetic as the nevus itself. Recent advancement in CO₂ laser technology developed the modern pulsed and scanned CO₂ laser as an outstanding treatment option for epidermal nevi patients.¹³

The present case was treated for oral findings so as to improve the esthetics and chewing ability of the patient and promote normal remaining maxillofacial skeletal growth. Malformed 21 was extracted followed by replacement of the teeth and orthodontic treatment to correct the malocclusion. Patient was advised for regular follow up for plaques on the face and black pigmentation on the tongue to look for any ominous signs of malignancy if present in future.

CONCLUSION

Sebeaceous nevi syndrome shows varied presentations and manifests with multiple oral findings. Oral manifestations of the condition can result in varied functional, esthetic and speech problems and thus hampering the standard of life of the patient. Oral diagnostician can play a prominent role in early diagnosis and proper referral of the patient.

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