Case Report

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Tooth and Nail Syndrome- A Rare Case Report and Review of Literature.

Abstract:

A uncommon autosomal dominant condition called Witkop's syndrome is marked by irregularities in the teeth and nails. It is an ectodermal dysplasia variation, which is thought to affect 1 in 100,000 live births. As a result, it is extremely difficult to identify in the general population due to its low prevalence and lack of distinguishing characteristics. The case study's objectives are to add to the body of existing knowledge by providing a useful approach to discovering the uncommon.

Key-words: Ectodermal dysplasia, Tooth and Nail Syndrome, Hypodontia

Introduction:

Agenesis of six or more teeth, excluding the third molars, is referred to as oligodontia. With a female-to-male gender ratio of 3:2.2, it has a documented prevalence of 0.3%. Oligodontia may result in ectodermal dysplasia (ED), Rieger's syndrome, oto-palato-digital syndrome, Witkop syndrome, oro-facial-digital syndrome, oculo-facial-cardiodental syndrome, or incontinentia pigmenti are a few examples of syndromes where this condition can occur alone or as a component.[5] Ectodermal Dysplasia (ED) is the name given to a collection of uncommon inherited diseases that are characterised by the dysplasia of tissues with ectodermal ancestry. The Witkop Tooth and Nail syndrome is one of the many types of ED.[1] Teeth, nails, and hair are the main organs affected, while sweat gland function is unaffected. This autosomal dominant condition, which was first described by Witkop in 1965, is reported to affect 1-2 out of every 10,000 newborns.[2] Congenital toothlessness can severely impair the young patient's physical and emotional development, especially during puberty. In the absence of permanent successors, primary teeth must be retained because they maintain the height of the alveolar bone in a region that may eventually have more extensive treatment. The purpose of this paper is to present a case of Witkop's syndrome and contribute to the body of literature because it is difficult to diagnose in the general population due to its low incidence rate and lack of distinguishing characteristics.[3]

Case report

A 15-year-old female patient reported tooutpatient department with a chief complaint of missing teeth in upper front region of mouthsince 8years. History of present illness reveals congenitally missing teeth in maxillary anterior region of mouth since 8 years. Medical history was non-contributory. On clinical examination general examination along with extraoral examination was done. General examination

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revealed normal gait, built, height and weight with normal motor and sensory functions. It also revealed normal hair (both in growth and texture) and skin. However, long thin nail bed seen in all the digits of both hands indicating koilonychia. Extraoral examination revealed no facial asymmetry with well coordinated and well synchronised movements of temporomandibular joint. It also revealed strabismus, hypoplastic maxilla with concave profile and incompetent lips.

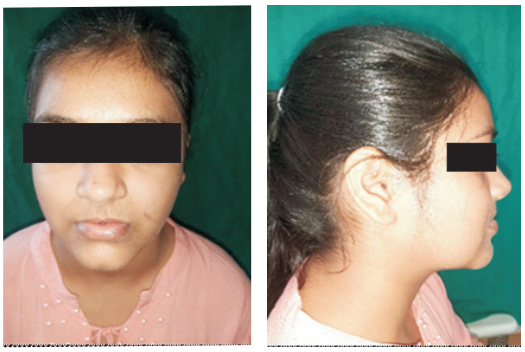

Figure 1: Frontal view showing strabismus and lateral view showing concave profile of the patient.

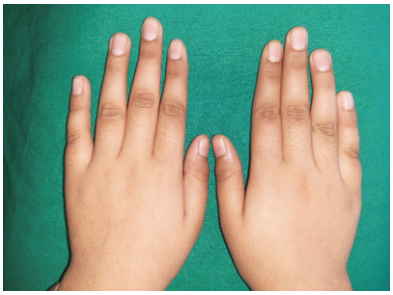

Figure 2: Image of koilonychia in both hands.

Intraoral examination revealed multiple clinically missing teeth including maxillary central incisors and premolars and mandibular canines and premolars. Patient also presented an angle's class III molar relation bilaterally with peg shaped right lateral incisor on the maxillary arch.

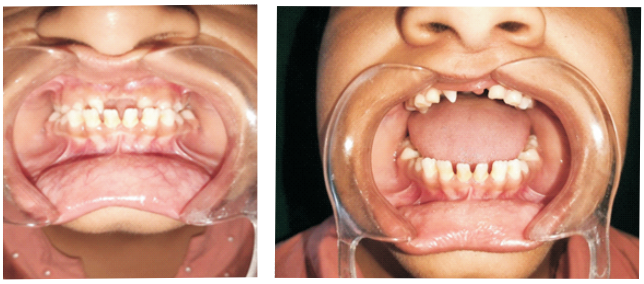

Figure 3: Intraoral images showing multiple missing permanent teeth

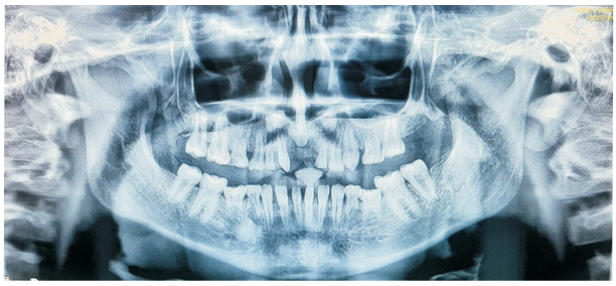

Figure 4**:** Panoramic radiograph showing multiple missing permanent tooth.

Radiographic examination included orthopantomogram (OPG) that revealed multiple missing permanent tooth 11 12 13 14 15 21 22 24 25 in the maxillary arch and 31 32 33 34 35 41 42 43 44 45 in mandibular arch.

Considering the clinical and radiographic findings she was diagnosed to be a patient of Witkop's syndrome (familial hypodontia, fingernail onychorrhexis, and toenail koilonychia).

Discussion:

Witkop syndrome is an uncommon form of ectodermal dysplasia, a category of genetic illnesses characterised by the absence or diminished function of two or more ectoderm-derived structures, such as hairs, nails, teeth, glands, etc. The presence of hypodontia and nail dysplasia, with minimal involvement of the hair and sweat glands, are the primary characteristics of the syndrome. Witkop syndrome is caused by a nonsense mutation in the MSX1 gene, which was identified in 2001. The oral mesenchyme is one embryonic structure that expresses the transcription factor MSX1.[4]

In the first branchial arch's mandibular and maxillary processes as well as the nail bed mesenchyme, it is expressed in neural crest-derived mesenchyme. When compared to the more prevalent x-link recessive hypohidrotic Ectodermal dysplasias, dental defects in the tooth-and-nail syndrome are typically less severe.

Little, widely spaced conical teeth, congenital absence of several permanent teeth, and retention of primary teeth are characteristics of those who are affected. The maxillary incisors, second molars, and maxillary canines are the most frequently missing teeth. There are nails that are dysplastic, such as brittle, spoon-shaped finger and toe nails (koilonychyia), which are thin, slow-growing, and spoon-shaped (koilonychyia). In general, toenails experience more damage than fingernails. With age, the nail alterations are

known to get better. Witkop's syndrome is not explicitly related with hair abnormalities, but some patients may have finely textured or diffusely thin hair without any obvious hypotrichosis or alopecia patches. The affected woman in the current case study is normothermic, has normal hair development and pattern, but has damaged (dysplastic) toenails and fingernails.[5]

An intraoral clinical and radiographic examination revealed that 18 permanent teeth were missing due to birth defects. The extraoral and intraoral findings of the current case report are comparable to those of previously reported research.

Other syndromes that resemble Witkop syndrome in certain ways include Fried TNS and Trichoonychodental (TOD) syndrome. Fried TNS may be an autosomal recessive variant that is markedly characterised by sparseness and thinning of hair, brows, and lips. Although hypohidrosis is a symptom of TOD syndrome, it is also extremely comparable. The absence of palmoplantar keratosis and the prominence of tooth involvement in the current instance assisted to distinguish between Clouston and Witkop syndromes because both conditions are frequently associated with each other.[6]

It might be difficult to diagnose Witkop syndrome in young patients when they exhibit a variety of symptoms. In order to maintain and restore the facial form and function, such patients require orthodontic and prosthodontic treatment considerations. Incorrect functions are brought on by partial or complete anodontia, including impaired speaking, reduced eating ability, and aesthetic issues. Affected people and their family benefit from genetic counselling since it helps them understand the illness. It has been suggested to lubricate nails and keep them short and smooth to prevent fungal infections.
For the functional and aesthetic repair as well as the psychological health of such patients in the early stages of life, a multidisciplinary approach is required.[6]

Conclusion:

As with any kind of ED, where young people present with a variety of symptoms, identifying a patient with Witkop's syndrome can be extremely difficult. Complete or partial anodontia results in functional impairments including a speech abnormality, a problem with the way they chew, and an aesthetic issue. As a result, a comprehensive approach to oral therapy in infancy may offer functional and aesthetic restoration in addition to mental health.[1]

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