Vander Woude’s syndrome: The rarest of the rare

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Abstract

One of the most common developmental defects seen in south India is cleft lip and palate. Among them a few are associated with lip pits and termed as Vander Woude’s syndrome. The early diagnosis of this rare syndrome is very necessary followed by a multidisciplinary approach. It is also necessary to differentiate this syndrome from the other syndromes which may present similar features. A case report of the same is presented here requiring a multidisciplinary approach for a functional and esthetically pleasing outcome.

Keywords: Cleft lip and palate, lip pits, Vander Woude’s syndrome

Introduction

Congenital lip pits are malformations that occur on the paramedial portion of the vermilion border of the lip. This may or may not occur along with cleft lip and palate. If it occurs along with cleft lip and palate, it is termed as Vander Woude’s syndrome. It is caused by an autosomal dominant trait. The other anomalies which may or may not be associated with syndrome are hypodontia, hypoplasia, ankyloglossia, high arched palate, limb anomalies, congenital heart defects, etc. Most cases have been associated with a deletion of chromosome 1q32-q41, but an extra chromosomal locus at 1p34 has been identified. The syndrome affects about 1 in 1,00,000–2,00,000 people. There is no definite sex predilection. Lip pits may result due to notching of the lip at an early stage of development with fixation of the tissues at the base of the notch or it may result from a failure of complete union of embryonic lateral sulci of the lip.

The surface opening of the lip may present as a circular or transverse slit or be located at the apex of nipple-like elevations. Its diameter may be upto 3 mm and its depth can range from 1 to 15 mm. The problem associated with lip pits is exudation of mucous on the lower labial skin which is a source of embarrassment for the patient.

Case Report

A 7-year-old male child reported to the Department of Pedodontics and Preventive dentistry with a chief complaint of carious teeth. Natal history revealed that he was delivered through a caesarean section. Maternal history relieved no medications or illness during pregnancy. Family history was not significant. Patient had a history of nasal twang. On detailed general examination, it was seen that the patient was a treated case of unilateral cleft lip and palate, and two lip pits were present on the lower lip [Figures 1 and 2]. When the lip was compressed there was mucous secretion. Intraoral examination revealed carious teeth with 16, 26, 36, and 46 and root stumps with 55, 64, and 65 [Figure 3]. A deep bite was also evident [Figure 4]. A corrected palatal scar was noticeable. An OPG of the patient was taken [Figure 5].

The treatment planned was restoration of carious teeth followed by extraction of root stumps [Figure 6]. The patient was also advised to undergo orthodontic treatment and a cosmetic lip correction for the lip pits.

Discussion

The occurrence of forms of Vander Woude’s syndrome should be kept in mind. It is important to diagnose these syndromes from oro facial digital syndrome and popliteal syndrome which manifests similar clinical features. These entities are allelic variants of the same condition. The other features which help to differentiate are as follows: oro facial digital syndrome shows cleft lobular tongue, digital malformation and mental retardation, whereas popliteal syndrome shows syndactyly.
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digital reduction, and syngathia. Because the symptoms of these children are limited and because the affected individuals have normal intelligence, this disorder should not be confused with non-syndromic cleft lip and palate.

The treatment of such cases should be surgical excision of the labial and commissural pits if esthetics is a major concern. It should include total removal of the minor salivary glands that exude secretions at the base of pits to prevent formations...
of cysts and mucoceles. The treatment should be carried out in collaboration with plastic surgery, oral maxillofacial surgeons, and orthodontists.\textsuperscript{[1,4]} Other treatments like cross bite corrections, maxillary expansions, restorations, and extractions should also be carried out. A multidisciplinary approach is very necessary to carry out the treatment thus improving the self-esteem of the patient at an early age.

**Conclusion**

A very rare case of Vander Woude’s syndrome has been presented here. An early and proper diagnosis followed by a multidisciplinary approach is necessary to improve the self-esteem of the patient.

**References**

1. Newman MA, Narre NO, Nyako EA. Vander Woude’s Syndrome: Report of a case. Ghana Med J 2005; 39: 58-70.
2. Lacombe D, Pedesparn J M, Phontan D, Chateil JF, Verloes A. Phenotypic variability in vander Woudes syndrome: Genetic Couns 1995;6(3): 221-6.
3. Bacain M, Walker AP. Lip pits and deletion 1q 32-41. Am J Med Genet 1987; 26: 436-43.
4. Arangannal P, Muthu MS, Nirmal L. Vander Woude’s syndrome: A case report. J Indian Soc Pedod Prev Dent 2002; 20:102-3.
5. Wideman HR, Kunze J. Clinical syndromes. . 4th edition. Mosby Wolfe Co; London 1997 p. 70-71.
6. Pinkham JR. Pediatric Dentistry. . 3rd edition. WB Saunders Co; Elsevier publications, Canada 2005 p. 44-47
7. Shafer WG. Textbook of Oral pathology. . 4th edition. WB Saunders Co; Elsevier publication, India 2002 p. 10-1.

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