

VAN DER WOUDE SYNDROME – CASE REPORT

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Abstract:

Genes contains the blue prints for human growth and development. Van der Woude syndrome is a developmental disorder characterized by lip pits on lower lip, cleft palate alone or with cleft lip in some cases & missing teeth. The features of VWS are discussed here with a case report. Bilateral lip pits with submucous cleft palate and bifid uvula were appreciated in the case.

Key words: Congenital lip pits, cleft palate, Van der woude syndrome

Introduction and review of literature:

The Van der Woude syndrome is a dominantly inherited developmental disorder that was first described by Anne Van der Woude in 1954. Van der Woude syndrome is a rare autosomal dominant craniofacial disorder characterized by lower lip pits, cleft lip and / or cleft palate. The VWS locus has been mapped to 1q32-q41 but the VWS gene has not been isolated. Frequency in. population is 1: 75,000 – 1:1, 00,000. 70% of lip pits are associated with cleft lip or palate. It may be unilateral or bilateral. Submucous cleft palate is common with hypernasal voice. Missing incisors or premolars is a common finding. Other oral manifestations include sygnathia (congenital adhesion of the jaws), narrow, high arched palate and ankyloglossia.^{1,2,3,4,5,6,7, 8}

Case Report:

We reported a 15 year old son who had chief complaint of teeth stained with tobacco. We also examined that patient had lip pits in lower lip bilaterally as depression and he was aware about that since childhood, but had not taken any treatment (Fig 1). On intraoral examination, he had tobacco stains and submucous cleft palate with bifid uvula (Fig.2). He did not have any significant past medical history. Patient was asked for family history which revealed that his 50- year old father also had lip pits bilaterally as depression on lower lip (Fig 3 & 4). Intraoral examination revealed generalized periodontitis with tobacco stains. Lower left second molar tooth was grade III mobile. Recession of gingiva present in all teeth. . Patient was operated for cleft palate before 34 years. Hypodontia not present in any of patients. No sign of saliva leakage

or sinus present in region of lip pits of lower lip of both patients. Both the patients had hypernasal voice.

We recommended surgical correction for esthetic purpose. However, none of above mentioned patients was ready for that.

Discussion

Congenital lip pits and fistulae are malformations of the lips which often follow a hereditary pattern, may occur alone or in association with other anomalies. Different theories have been postulated to explain the etiology of lip pits. Lip pits can result due to notching of lip at an early stage of development with fixation of tissues of the base of the notch or it may result from a failure of complete union of embryonic lateral sulci of the lip. Lip pits usually present itself as a unilateral/ bilateral depression that occurs on vermilion portion of either lip but commonly on lower lip. Lip pits may be associated with accessory salivary glands. Sometimes lip pits may be the only manifestation of this syndrome. The treatment of pits is surgical excision with removal of entire fistulous tract with histopathologic evaluation because of presence of salivary tissue may lead to cyst formation.¹⁻⁸

Recognition of lip pits is important as multiple other anomalies associated can be detected. Proper evaluation and treatment of these associated abnormalities and genetic counseling is important. Popliteal pterygium syndrome is another rare autosomal dominant syndrome with a similar orofacial phenotype but it includes skin and genital anomalies. Clinical Manifestation of PPS are



Fig. 1. Photograph showing congenital lip pits of lower lip in patient (Son)



Fig. 3. Photograph showing patient's father with congenital lip pits of lower lip.

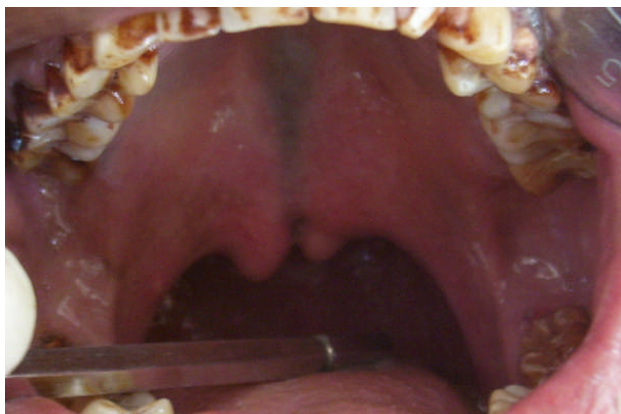


Fig. 2. Photograph showing submucous cleft palate in patient (son)



Fig. 4. Photograph showing both patients (son and father together) with congenital lip pits of lower lip

popliteal webbing, cleft palate with or without cleft lip and genital and nail anomalies, lower lip pits, syngnathia and syndactyly.^{3,6}

Conclusion and summary

Congenital lip pits associated with cleft lip/ palate is associated with many syndromes. One of them is Van der Woude syndrome. Early diagnosis helps a child in various necessary treatments like surgery, speech therapy and dental management

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