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Mohr-Claussen Syndrome: A Rare Case

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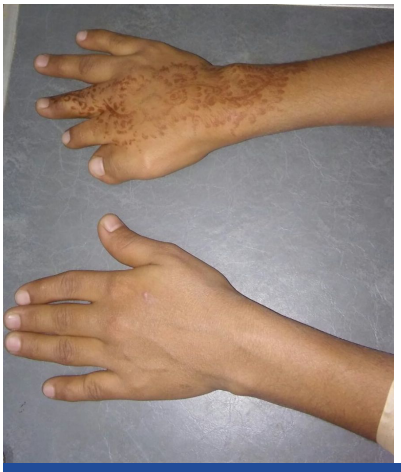
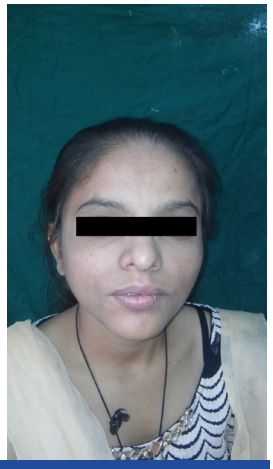
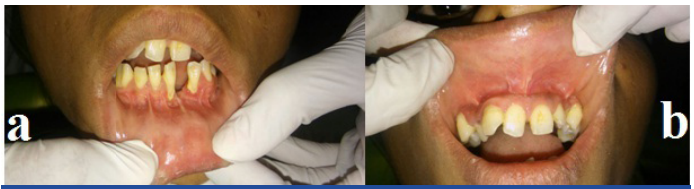
A 22-year-old, female patient reported with complaint of poor esthetics. Her past medical, dental, personal as well as family histories were non-contributory. After performing thorough general and extra oral clinical examination under aseptic precautions, various orofacial abnormalities were noted down which included brachydactyly of a thumb and an index finger of upper extremities on right side [Table/Fig-1] and hypertelorism with frontal bossing and depressed nasal bridge [Table/Fig-2]. Intra oral examination revealed hyperplastic frenula [Table/Fig-3], high lingual frenal attachment with lobed tongue [Table/Fig-4], multiple supernumerary teeth and fused teeth [Table/Fig-5]. Moreover, radiographic investigations were done which showed supernumerary teeth, fusion of two teeth and alveolar clefts in OPG and IOPA [Table/Fig-6,7]. A hand and wrist radiograph showed defect in phalanges [Table/Fig-8]. Review of literature shows that the diagnosis is based on clinical findings [Table/Fig-9] [1]. There are total 13 types of Oro-Facial Digital Syndrome (OFDS) out of which clinical features of Type 1 and Type 2 must be ruled out properly.

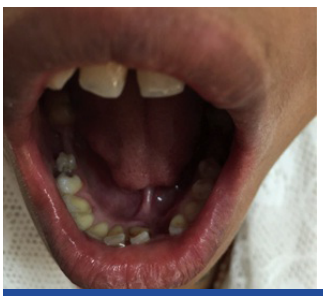


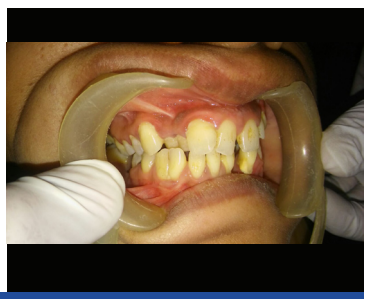


[Table/Fig-1]: Showing brachydactyly of a thumb and an index finger of upper extremities. **[Table/Fig-2]:** Showing frontal bossing and depressed nasal bridge.

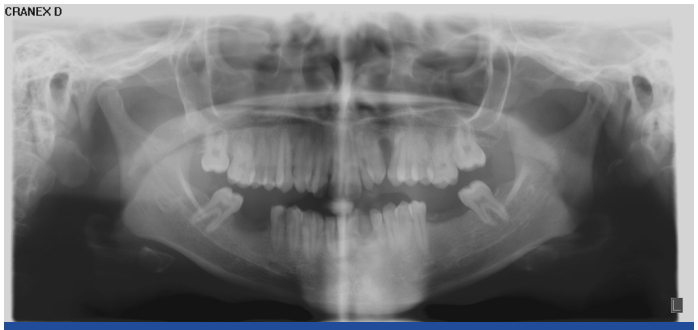


[Table/Fig-3]: Showing hyperplastic frenula a) maxilla b) mandible.

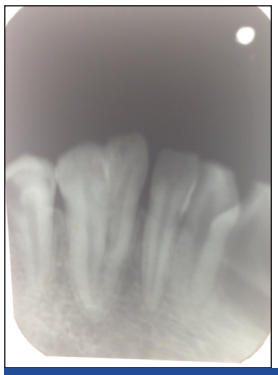


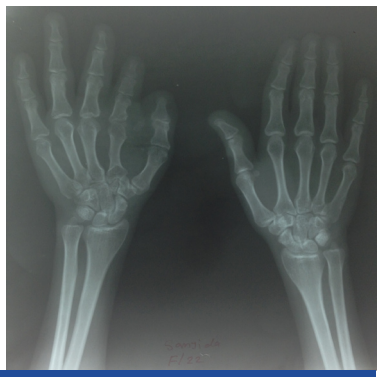


[Table/Fig-4]: Showing high frenal attachment with lobed tongue.
[Table/Fig-5]: Showing multiple supernumerary teeth and fused teeth.



[Table/Fig-6]: Showing radiographic evidence of alveolar clefts in upper and lower front tooth region.





[Table/Fig-7]: Fusion of canine and a supernumerary tooth in lower right back tooth region. **[Table/Fig-8]:** Showing hand-wrist radiograph with defect in phalanges.

Differential diagnoses which can be included are Type 1 OFDS and Acro–fronto–facio–nasal syndrome. Type 1 can be differentiated on clinical grounds whereas mental retardation and broad notched nasal tip are prime features of Acro–fronto–facio–nasal syndrome apart from oral, facial and digital abnormalities.

After primary assessment and detailed case-history, patient was diagnosed as case of OFDS Type 2, as majority of the features were in favour of the same. Patient was then advised full mouth rehabilitation with multi-disciplinary approach.

DISCUSSION

According to Gorlin and Psaume, the first observation about OFDS was back in 1883, which is characterized by developmental defects of the oral cavity, facial structures and digits of hands and feet [2]. There are about 13 subtypes of OFDS reported in literature [3] out of which Type 2 OFDS is the most common variant that is seen clinically, which is named as Mohr-Claussen Syndrome (MCS) [4].

MCS has a wide range of clinical features. Developmental defects of the oral cavity, abnormal facial features and digits are main manifestation of this syndrome. Abnormalities that occur in various

Type of OFDS	Clinical features
OFDS-I (Papillon-Léage-Psaume) X-linked	Oral Cleft lip and palate lobulated tongue tongue nodules (hamartomas) Facial Hypertelorism Digital Brachydactyly, syndactyly, clinodactyly reduplicated hallux Others Cerebral abnormalities renal dysplasias mental retardation
OFDS-II (Mohr-Claussen)	Oral Cleft lip and palate lobulated tongue micrognathia Facial Broad nasal bridge frontal bossing Digital Brachydactyly, syndactyly, clinodactyly, polydactyly reduplicated hallux Others Conductive hearing loss congenital heart disease normal intelligence

[Table/Fig-9]: Differences between orofacial features in Type 1 and Type 2 OFDS.

types of oral-facial-digital syndrome include a cleft in the tongue, unusually lobed tongue and benign tumors or nodules on the tongue [5]. Affected individuals may also have extra, missing, or defective teeth. Another common feature is cleft palate. Some people with OFDS have hyperplastic frenula that abnormally attach the lip to the gums.

Distinctive facial features often associated with OFDS include cleft lip; a depressed nasal bridge; and hypertelorism (increased inter papillary distance) [5].

Abnormalities of the digits can affect both the fingers and the toes in people with OFDS. These abnormalities include syndactyly, brachydactyly, clinodactyly and polydactyly [6].

Presence of clinical features may vary depending upon type of OFDS. Although, many of the oral, facial and digital abnormalities should be noted to give clinical diagnosis of OFDS irrespective of their intensity.

Treatment of malformations of the oral cavity involves teamwork and multispecialty approach in different areas of dentistry. Hence, we as a dental professional must have sound knowledge so that such rare conditions are not missed out. One should be able to correlate these dental abnormalities with the general physical abnormalities and not just merely concentrate on oral conditions.

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