A 7 Year Follow up of a Case of Lobster-Claw Syndrome : A Heredo-Familial Disorder

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Abstract

Lobster claw syndrome is an autosomal dominant, hand-foot malformation with oro-dental features. This is a rare condition and the incidence is reported to be 1 to 4 in 100000 live births. Most reports have focused on the hand-foot deformity of this syndrome. This case report is a 7-yr follow up of a 17-yr old female that highlights the typical oro-dental features of this heredo-familial disorder with reference to the functional and esthetic management of the dentition.

KeyWords: Lobster-claw, Ectrodactyly, EEC syndrome, Intrafamilial variations.

Introduction

Lobster-claw syndrome is a developmental defect of the hand and feet. Cleft hand deformity is secondary to wedge-shaped defect in primary hand plate. Severe cases are associated with syndactyly of thumb and index finger and of ring and little finger with absence of third metacarpal ray. The remaining sets of digits appose each other giving a Lobster-Claw appearance.

Autosomal dominant inheritance is most commonly known. It usually appears bilaterally and is associated with cleft feet. Associated anomalies include cleft feet, congenital heart disease, cataracts,defect of tibia, radioulnar synostosis and fusion of kidneys. [1]

In 1804, Franzheinrich Mortens first identified Lobster-Claw in a patient named Johann Gottlob.[2] The effect of Heredo-familial cleft deformity and its effects on normal function were studied by Meyerding HW and Upshaw in1947.[3] Subrato Sarkar in 1999 was the first to report such a case with intra-oral manifestations like hypodontia, retained deciduous tooth with Lobster- claw deformity.[4] A familial Lobster-Claw syndrome was reported by Sejal Thakkar and Yogesh Marfatia in the year 2000 highlighting the intrafamilial variations indicating variable penetrance.[5] This is a rare case report that presents the intra oral features of such a heredo-familial cleft defect with reference to the functional and esthetic management of the dentition.

Case Report

This is a 7 yr old follow up of a girl who is currently seventeen years who reported to the Department of Pedodontics and Preventive Dentistry, Rajah Muthiah Dental College and Hospital with a complaint of missing lower front teeth.She first reported to the department when she was around 10 yrs old. At that time general examination was done which revealed abnormalities present in both upper and lower extremities in the form of missing 2-nd and 3-rd digits and clefting of the hand and feet since birth. The family history revealed that a similar defect was present in the father as well, but the missing digit varied i.e., only 2-nd digit. History of consanguinity of parents, drug intake, and exposure to radiation during pregnancy were ruled out. No other child in the family i.e., elder sister and younger brother were affected. On extra- oral examination mid-facial hypoplasia, hypertelorism, saddle nose and thick everted lips were evident. On intra-oral examination many permanent teeth were found to be missing.

Dental History

Patient gave a history of fixed orthodontic treatment before one year. Glass ionomer and amalgam restorations were present in posterior teeth. She was wearing a mandibular removable partial denture in relation to 31,41,32,42 since 1 year which got fractured and needed a replacement.

Intra- Oral Examination

The size and shape of the individual teeth were studied by the help of the study cast. The intra oral examination along with panoramic radiograph analysis revealed the absence of following permanent tooth germs: 12, 13, 22, 23, 25, 28, 31, 32, 33, 35, 38, 41, 42, 43, 45, 48

Number of teeth clinically present were:

Maxilla	:	Mandible
18 (erupting)	:	37 (microdont)
17 (microdont)	:	36
16	:	34 (malformed)
15	:	73 (malformed)
14 (malformed)	:	83 (microdontia)
52 (esthetically build up with composite to simulate 12)	:	44 (malformed)
11 (retroclined)	:	46
21	:	47 (microdont)
62 (esthetically built up with composite to simulate 22)	:	
24	:	
65	:	
26	:	
27(microdont)	:	

The characteristic appearance of the palms and feet and the intraoral appearance of hypodontia paved the way in diagnosing the present case as Lobster-Claw Syndrome. The patient's father also presented with congenital absence of 2nd digit of both hands and feet and several permenant teeth were missing and malformed both in maxillary and mandibular arch indicating a heredofamilial defect with variable penetrance.

As the chief complaint of the patient was missing lower anterior teeth decision was made to fabricate a fixed partial denture in relation to 41,42,31 and 32. Crown preparation was done in relation to 73 and 83 and a metal ceramic bridge was delivered in relation to 73, 32, 31, 41, 42 and 83.

Orthodontic correction of retroclined 11 was done. Oral hygiene instructions were given to the patient to maintain a better hygiene and to improve the prognosis of the dentition. Patient was advised to have regular recall visits.

Discussion

Split hand and split foot deformity may occur as the only malformation or may occur in association with consistent pattern of skeletal or other system disease (syndromatic). The typical split hand anomaly can be inherited in any one of the three Mendelian ways: autosomal dominant, autosomal recessive or X linked recessive. However, if they are associated with other abnormalities or a family history, the risk to future pregnancies may be as high as 50%.[6]

Brasky described two types of split hand/split foot defect. One is typical and the other is atypical . In typical type, all four limbs are involved, with perhaps an even greater propensity for only the feet to be affected. The cleft is typically deep and are "V" shaped. It is familial and usually inherited as an autosomal dominant mode, consistent with the observation that multiple limb and symmetric defects are often genetic. Other associated abnormalities include cleft lip and cleft palate, reduction in the number and size of phalanges, syndactyly, polydactyly, triphalangeal limb, scalp defect, genitourinary anomalies and atresia of nasolacrimal duct. The atypical type involves a single upper arm and always spare the feet. The cleft is a wider, U-shaped palm, with only thumb and small finger remaining and those digits may be attenuated soft tissue nubbins, remnants of embryonic fingers, may be found in the cleft. This upper extremity malformation involving one arm occurs as a sporadic event and has no genetic basis.[7]

Syndromes presenting with limb deficiency

Aetiology	Syndrome	Limb deficiency
Autosomal dominant	EEC (Ectrodactyly, Ectodermal dysplasia, Cleft lip/palate)	Split hand and foot(ectrodactyly)
	Adams Oliver	Transverse defect
Autosomal recessive	Roberts	Longitudinal affecting whole limb
	Grebe	Hypoplastic radii,ulnae and tibiae
X-linked recessive	Split hand/foot	Split hand/foot
Chromosomal	Trisomy 18	Longitudinal all degrees
	Trisomy 13	Radial Absence
Drugs	Thalidomide	Longitudinal all degrees
	Aminopterin	Hypoplasia(forearm), Hypodactyly
Infection	Varicella	Hypoplasia, hypodactyly
Sporadic syndromes	Poland/Moebius	Hypoplasia forearm, oligodactyly
	Oro-facial digital syndrome	Oligodactyly
	Apert's syndrome	Hypoplastic forearm,oligodactyly

Trans vaginal ultrasonography performed at 14 weeks gestation demonstrated a bilateral cleft lip, Lobster-claw deformities of hands and feet, in keeping with the diagnosis of this syndrome.[8] Early diagnosis will allow parents to avail counselling and in particular obtain reassurance regarding the low risk of mental handicap.The management of Lobster-claw syndrome requires a multidisciplinary team including a plastic surgeon, dental surgeon, ophthalmologist, dermatologist, nephrologist and speech therapist. [9]

From the dental standpoint following should be done

- Preserve the primary dentition as long as possible.
- Saliva substitutes in case xerostomia is severe.
- Restoration of carious primary as well as permanent teeth
- Prosthesis using support of any or all the remaining teeth.
- Complete dentures in case no teeth are present.

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Legends

Fig 1. Photograph showing Lobster Claw deformity
Fig 2. Extra oral Photograph showing saddle nose, hypertelorism and everted lips
Fig 3. Photograph showing patient and her father with the deformity
Fig 4. Intra oral photograph showing microdontia and malformed teeth
Fig 5. Orthopantamogram showing retained deciduous teeth and hypodontia
Fig 6. Intra oral photograph showing retroclined 11 and edentulous area in relation to 31,32,41,42
Fig 7. Intra oral photograph showing metal ceramic bridge cemented in relation to 31,32,41,42
Fig 8. Intraoral photograph showing orthodontic correction of retroclined 11

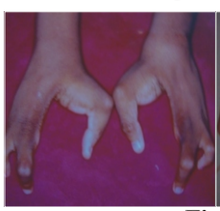


Fig. 1

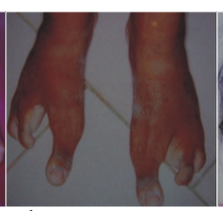
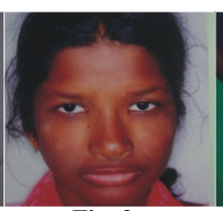


Fig. 2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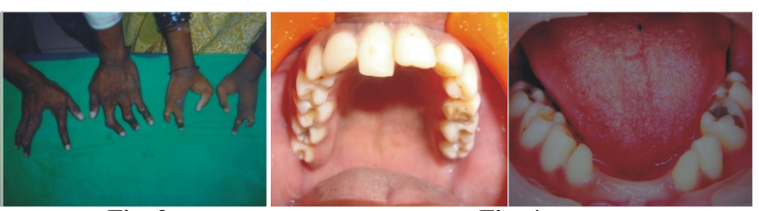
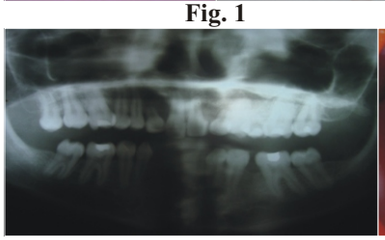


Fig. 4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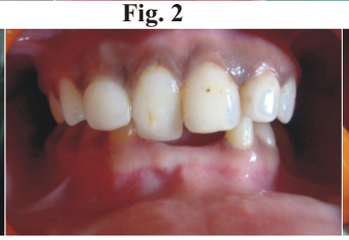
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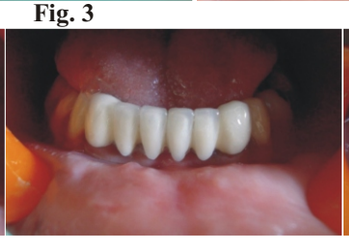
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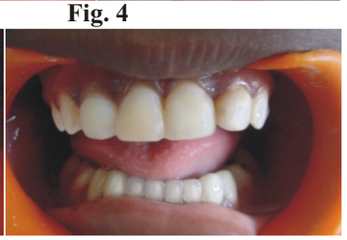
Fig. 8