HEMIFACIAL MICROSOMIA : CASE REPORT

Dr. Amit Shaw*, Dr. Bides Bhaumik, Dr. Ali Asger Nakib***

Abstract

Hemifacial microsomia is a congenital anomaly of face caused due to interference with migration of neural crest cells and charecterized by lack of tissue on the affected side of face. The affected child suffers from gross disfiguration of face, functional disturbances, malocclusion and poor psychosocial acceptance. An early detection and management is therefore necessary which require a team approach by orthodontist, radiologist, oral surgeon, plastic surgeon and prosthodontist. In this article etiology, clinical features and treatment outline of Hemifacial microsomia is discussed along with two case reports.

Key Words Hemifacial microsomia , Hypoplasia

INTRODUCTION

Hemifacial microsomia is a birth defect in which the lower half of one side of face does not grow normally resulting in a gross asymmetric face. It is the second most common facial birth defect after clefts with an incidence range of 1 in 5600 live births.[3,5,6,8] Right side is affected more than left.[10] In some cases it is a part of a larger syndrome such as Goldenhar syndrome (oculo-auriculo-vertebral dysplasia) which includes vertebral anomalies and epibulbar dermoids.[2] This is known as craniofacial microsomia when there is involvement of cranial deformities.

EMBRYOLOGY

Hemifacial microsomia results from the abnormal development of the first and second branchial arches and the first branchial membrane These arches are the mounds of tissue that contribute to the development of facial structures such as maxilla, mandible, zygomatic bone and ear.[9]

During the 4th week of human embryonic life the neural crest cells migrate extensively and forms most of the mesenchymal tissue in the facial region which later differentiate into the skeletal and connective tissues, including the jaw bones and the teeth. In Hemifacial Microsomia neural crest cells with the longest migration path, those taking a circuitous route to the lateral and lower areas of face, are affected.[7] The structures most commonly affected are ascending ramus of themandible, temporo-mandibular joint, zygomatic arch, external and middle ear(incus, the malleus, and the tympanic bone), muscles of mastication and great vessels(aorta, Pulmonary artery).

PATHOGENESIS

1. Due to hemorrhage from the stapedial artery at 6th intrauterine period when the maxillary artery takes over the blood supply to the affected area.

ABOUT THE AUTHORS

*Clinical Tutor
**Professor
Department of Orthodontics, Dr. R. Ahmed Dental College & Hospital

2. Recent study suggests that although hemorrhage at a critical time may be involved Hemifacial microsomia arises primarily from early loss of neural crest cells.[7]

ETIOLOGY

Teratogens like thalidomide, ante acne drug isotretinoin may be responsible for such pathologic effects.

CASE REPORT 1

A 12yr old female patient reported to the Orthodontics OPD of Dr R Ahmed Dental College with the chief complain of gross asymmetry of face

CLINICAL EXAMINATION

On general examination, the patient was moderately built, well nourished, cooperative and well oriented to time and place. The gait and posture were normal.

Extraoral examination

revealed, on inspection, facial asymmetry on the right side of the face that appeared short and flattened as compared to the left side. The body and ramus of mandible are short on the right side as compared to the left side of the face. The chin and midline were deviated to the right side. The ala of the nose and corner of the mouth were placed higher on the right side. The corner of the mouth was deviated to the right side. The lips were competent. Right ear is aberrantly placed, underdeveloped (microtia) with presence of preauricular skin tags. Lower third of face on profile examination appears longer compared to upper and middle thirds. The temporomandibular joint revealed deviation on opening to the right side following normal mouth opening (40 mm). On palpation, the right masseter and temporalis muscle was deficient. [fig.1, fig.2]

Intraoral examination

revealed mixed dentition with retained 53,63,65,75,85. The teeth were of normal size. 16 and 15 are in crossbite and there is posterior open bite on left side. This may be because ramus and body of the mandible is longer on left side compared to affected right side and resulting deviation of mandible to the right. Also maxilla is narrow on the affected right side.[fig.3, fig.4, fig.5, fig.6]

CBCT imaging

1. Hypoplasia of the right side of face

2. Absence of right condyle and coronoid process

3. Absence of right glenoid fossa.

4. Hypoplastic right ramus of mandible.

5. Hypoplastic right zygomatic bone .

On the basis of clinical and CBCT scan findings, the diagnosis of Hemifacial microsomia was made. [fig 7, fig 8, fig 9, fig 10]

CBCT with its dual advantage of low radiation dose and high image resolution is becoming an integral part of dentistry. Direct volume rendering with CBCT generates excellent 3D images which are very helpful for orthognathic surgeries. In the present case, the 3D images generated with CBCT, revealed aplasia of right mandibular condyle and coronoid process, hypoplasia of left mandibular ramus, hypoplastic right zygoma.

Differential diagnosis

HFM can be easily confused with Treacher Collins syndrome. The differentiating features are: HFM is unilateral, it is not hereditary and colobomas of the lower lids do not occur in HFM.[4]

CASE 2

A 8yr old female patient reported to the Orthodontics OPD of Dr R Ahmed Dental College with the chief complain of gross asymmetry of face, deformed right ear , hearing impairment and irregular teeth.

CLINICAL EXAMINATION

Extraoral examination reveals body and ramus of mandible are hypoplastic on the right side as compared to the left side of the face. The chin and midline were deviated to the right side. Right external ear is almost absent (anotia).

There are presence of depressed forehead, maxillary and malar bone hypoplasia more on right side than on left side giving a cocave facial profile.

The corner of the mouth were placed higher on the right side giving rise to an oblique lip line.[fig 11, fig 12]

Intraoral examination reveals over retained right maxillary lateral incisor. Maxillary arch is constricted giving rise to bilateral posterior crossbite. Lower arch midline is deviated to the left. There is anterior open bite on left side. [fig 13, fig 14, fig 15]

Radiograph was not available. On the basis of clinical examination the case was provisionally diagnosed as Hemifacial microsomia.

Fig 1: extraoral photograph-frontal

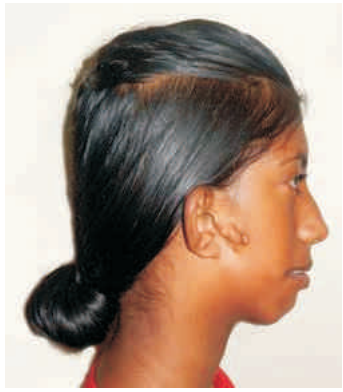

Fig 2: extraoral photograph-profile

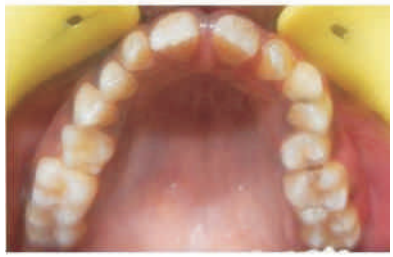

Fig 3: intraoral view-upper occlusal

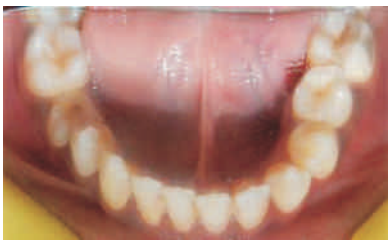

Fig 4: intraoral view-lower occlusal

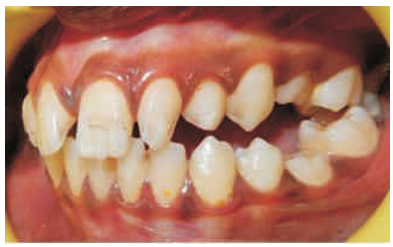

Fig 5: intraoral view-left lateral

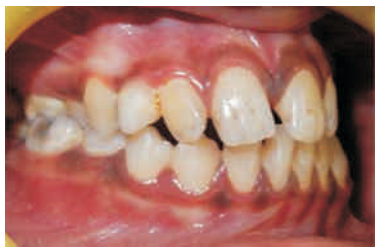

Fig 6: intraoral view-right lateral

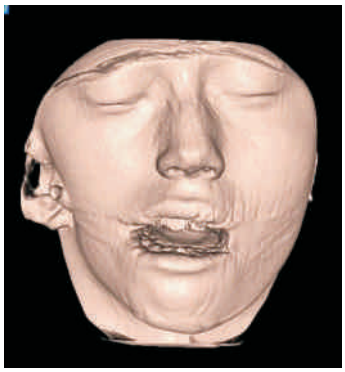

Fig. 7: CBCT image of facial soft tissue showing hypoplasia on the affected side

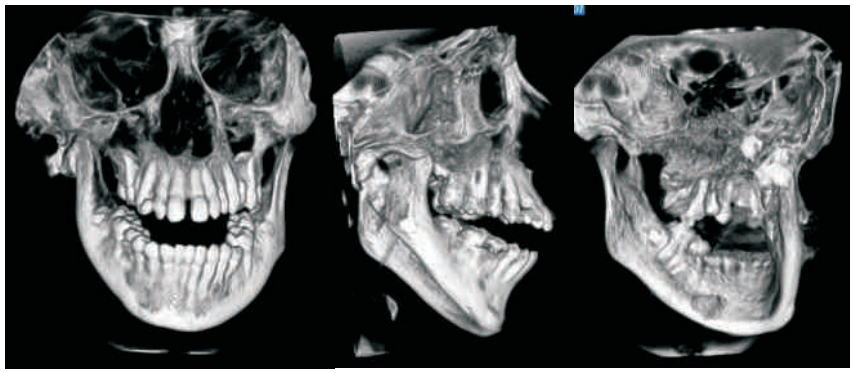

Fig 8,9,10 : CBCT image of cranium and facial skeleton showing hypoplastic ramus and condyle of mandible on right side

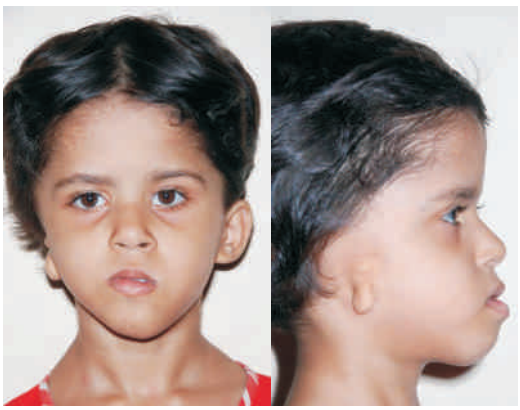

Fig 11, 12: extraoral photographs (frontal, profile)

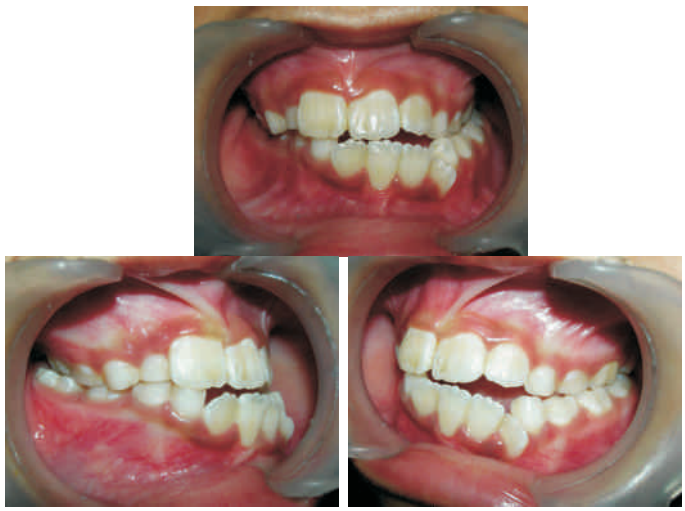

Fig 13, 14, 15: intraoral photographs

MANAGEMENT

A newborn patient with Hemifacial Microsomia is assessed for vital functions – breathing, eating and sleeping. Severe cases may require tracheostomy to maintain the airway.

Early orthopedic intervention:

Functional appliance: according to a study by Prof Bärbel Kahl-Nieke and Dr Roman Fischbach1 successful orthopedic intervention can be done at an early age using functional appliance. By means of the construction bite, the mandible is kept in a slightly forward and overcompensated centered position in order to establish a change in muscle activity that could lead to enhanced bone apposition and optimal growth direction of the condyle, or in case of a missing condyle, in the condylar region. In addition, the adjusting of the activator on the affected side stimulates passive eruption of the upper buccal teeth. Their study showed with excellent patient cooperation there is improvement of function and occlusion and reduction of facial asymmetry. However neither size of the deformed hypoplastic condyle nor muscle volume and density have improved.

Conventional surgical method:

The deficient ramus of the mandible is partly replaced by an autologous costo-chondral bone graft. A costo-chondral graft provides length to the ramus, as well as a joint; it also acts as a growth centre. The chin should be re-positioned in the centre of the face during this procedure. For most children, a single operation is sufficient to correct the asymmetry. The problem with some grafts, however, is that they show overgrowth.[2]

Distraction osteogenesis: after surgical insertion of the distractor parents are instructed to rotate the screw at home as guided by the operator. This gradually lengthens the mandible on the affected side in proper direction and corrects the asymmetry but creates a gap between maxillary and mandibular teeth. The orthodontist then insert an interocclusal block to allow the upper teeth to erupt gradually into the gap and contact the lower teeth. In this way, the mouth and teeth are leveled.

External ear reconstruction: according to the American Society of Plastic Surgeons external ear reconstruction should be done at the age of 7-10 yrs.

CONCLUSION

The treatment objective in Hemifacial Microsomia is "to improve facial symmetry and mandibular function and to avoid maxillary growth disturbance". Success of the treatment of asymmetric facial growth seems to depend on the severity of the abnormality. Qualified and quantified hard and soft tissue CT evaluation is one way to improve diagnosis, treatment, and research concepts for asymmetric facial growth.[1] HM should be treated in craniofacial teams with enough clinical experience in treating these dentofacial malformations. This definitely will lead to more predictable and better results, fewer complications and a smaller number of surgical re-interventions.

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