CASE REPORT

Hemifacial Microsomia : Clinicoradiological Insight and Report of a Case

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OPEN ACCESS

Citation: Nidhi Chhabra, Anuj Chhabra. Hemifacial Microsomia : Clinicoradiological Insight and Report of a Case. Ethiop J Health Sci 2017;27(1):91-94. doi: http://dx.doi.org/10.4314/ejhs.v27i1.12.
Received: August 03, 2016
Accepted: October 21, 2016
Published: January 1, 2017

Funding: None
Competing Interests: The authors declare that this manuscript was approved by all authors in its form and that no competing interest exists.
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ABSTRACT

BACKGROUND: Hemifacial microsomia is a congenital malformation characterized by deficiency in the amount of hard and soft tissue on one side of the face. It is primarily a syndrome of the first branchial arch, involving underdevelopment of the temporomandibular joint, masticatory muscles, mandibular ramus, ear and, occasionally, defects in facial nerve and muscles.
CASE DETAILS: The clinical and radiological manifestations of a 14-year-old male patient having hemifacial microsomia is highlighted in this article to enhance our knowledge and diagnostic skill of this rare entity.
CONCLUSION: This case illustrates that early diagnosis and intervention in a patient with hemifacial microsomia is quintessential for proper functioning and esthetics of the orofacial structures, which will lead to a better prognosis.
KEYWORDS: Hemifacial microsomia, congenital malformation, hypoplasia

INTRODUCTION

Hemifacial microsomia (HFM) was first described by the German physician Carl Ferdinand Von Arlt in 1881 (1). It is also known as first and second branchial arch syndrome, oculo-auriculo-vertebral dysplasia, otomandibular-facial dysmorphogenesis and lateral facial dysplasia (2). HFM is comprised of unilateral microtia, macrostomia and malformation of mandibular ramus and condyle (1). Goldenhar syndrome is a variant of HFM, which includes vertebral anomalies and epibulbar dermoids.

During the development of the jaw, the neural crest cells migrate to the first pharyngeal arch from the posterior mesencephalic fold and from the first rhombomeres, which gives rise to the skeletal maxillo-mandibular component. Damage or disruption of these neural crest cells results in HFM and some related syndromes (3).

The HFM is characterized by asymmetrical defects of the first pharyngeal arch derivatives. The most commonly affected structures include ascending ramus of the mandible, temporo-mandibular joint, zygomatic arch and external and middle ear, which includes the incus, the malleus, and the tympanic bone. There are chances of deviation of the mandible accompanied by malocclusion and hearing defect (2). This case report describes the clinical and radiological features of hemifacial microsomia from the literature and adds one new case to enhance our knowledge and diagnostic skill of this unusual entity.

CASE REPORT

A 14-year-old patient presented to our department complaining of facial asymmetry since birth. On general examination, the patient was moderately built, cooperative and alert and had no history of hearing deficits or any other physical deformities. The gait and posture were normal. The medical, family and habit histories were non-contributory. No other family member had similar cases.

Extraoral examination revealed facial asymmetry with deviation of the lower jaw towards the right side, shifting the midline of the mandible. The right side of the face appeared short compared to the left side one. The ala of the nose and the corner of the mouth were placed higher on the right side. The corner of the mouth was deviated to the right side (Figure 1). The temporomandibular joint revealed no abnormality except for the deviation on the opening to the right side following normal mouth opening (40 mm). On palpation, the right masseter and the temporalis muscles were deficient.

Intraoral examination revealed a high arched palate, a significant dental crowding in right maxillary teeth. Maxillary left central and lateral incisors were carious (Figure 2). The incisors, premolars and molars of the right mandibular arch were missing (Figure 3).

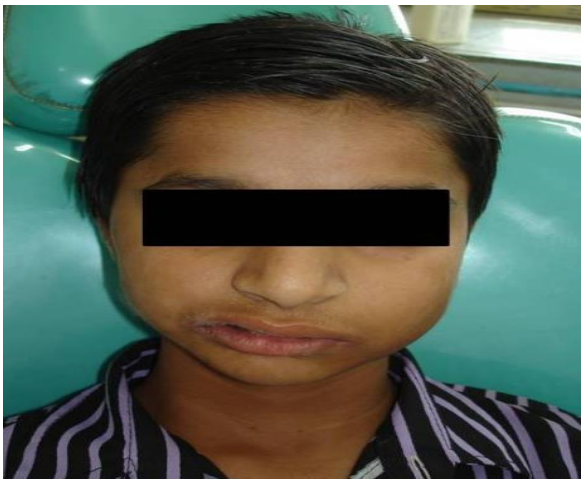

***Figure 1**: Frontal view of the patient revealed facial asymmetry with deviation of the lower jaw towards right side, shifting the midline of the mandible. The ala of the nose and corner of the mouth were placed higher on the right side*

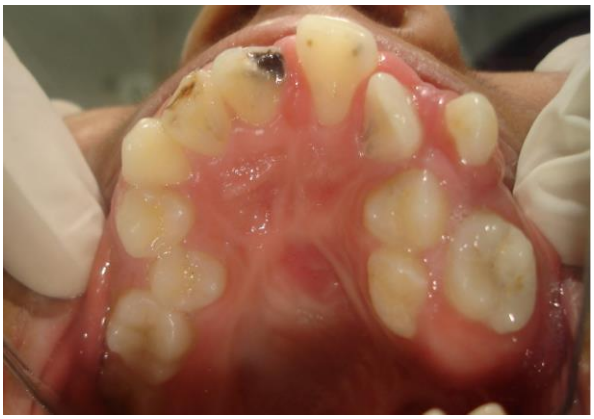

***Figure 2**: Intraoral examination of maxillary arch revealed a high arched palate, significant dental crowding in right maxillary teeth and carious left central and lateral incisors*

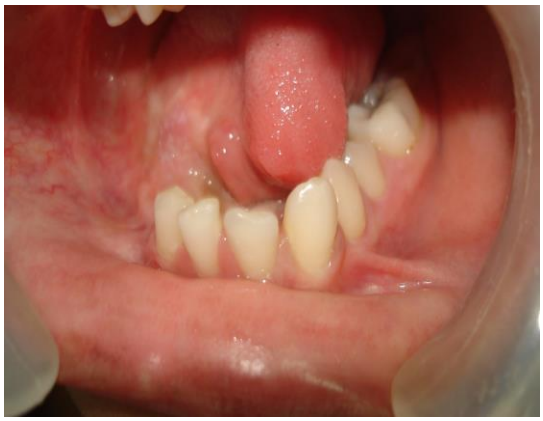

***Figure 3**: Mandibular arch examination showing missing incisors, premolars and molars in right mandibular arch*

The patient was diagnosed with HFM or facial hemiatrophy on the right side of the face. The differential diagnosis included Pierre Robin syndrome, Treacher Collin Syndrome, Parry-Romberg syndrome, Down's syndrome, hypoplasia of the condyle on the right side and congenital unilateral ankylosis on the right side.

Orthopantomograph examination revealed missing right mandibular incisors, premolars, molars, right hypoplastic mandibular ramus, small condylar head, reduced height of the body of mandible and high level of the external oblique ridge (Figure 4). Radiographic examination showed that there were no skeletal anomalies affecting the vertebrae. Intravenous pyelogram showed no renal abnormalities. Cardiac function, eyes and sight were excellent, and no mental retardation was found. Aural examination and

DOI: http://dx.doi.org/10.4314/ejhs.v27i1.12

audiological testing showed that both auricular structures and hearing were normal.

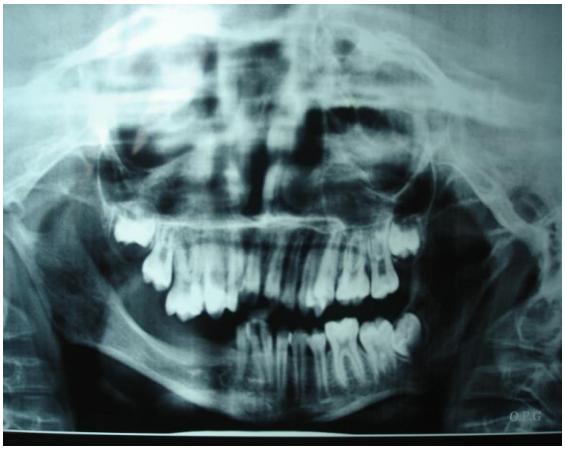

Figure 4*: Panoramic radiograph showing missing right mandibular premolars and molars; right hypoplastic mandibular ramus and small condylar head, reduced height of body of mandible, high level of external oblique ridge*

The treatment proposed for the patient was distraction osteogenesis to bring about the growth of the mandible to the desired size, restoration of the carious teeth and alignment of the teeth. The condition and proposed treatment were explained to the patient and to his family, and written informed consent was taken.

DISCUSSION

HFM is a complex malformation syndrome with a large host of genetic and teratogenic associations and a wide spectrum of clinical features, which involves the facial skeleton and other organ systems (4). Deformities may include auricular defects, preauricular tags and fistulae, microtia-atresia, mandibular, maxillary and orbital hypoplasia, microphthalmia, epibulbar dermoid, strabismus, conductive or sensoneural hearing loss and hypoplastic facial muscles (5). HFM which is associated with vertebral, cardiac and renal defects is called Goldenhar syndrome. The term, 'oculoauriculovertebral dysplasia' (OAVD) was suggested by Gorlin for this disorder (6).

HFM manifests as little asymmetry in the face, to severe underdevelopment of one facial half, with orbital implications, a partially formed ear or even a total absence of the ear. The chin and the facial midline are off-centred, and they get deviated to the affected side. Often, one corner of the mouth is situated at a higher level than the other, giving rise to an oblique lip line. The most common symptoms are the unilateral hypoplastic maxillary and temporal bones with or without unilateral shorter zygomatic arch (2).

In the present case, there was gross facial asymmetry; the right side of the face was underdeveloped with prominent and short zygomatic arch. The chin and the midline were deviated to the right side with the corner of the mouth situated slightly higher on the right side compared with the left side, which was consistent with the literature given.

Intraoral manifestations include aplastic teeth and enamel, significant delay of tooth development on the affected side and increased frequency of missing teeth on the affected side, all of which were seen in the reported case. The subject in the present case presented with hypodontia in the mandibular incisors, premolars and molars on the affected side, which is consistent with the literature. The incidence of delayed tooth development with hemifacial microsomia is proportional to the extent of mandibular deformity (7,8).

Chalky opacifications of the enamel are occasionally found on the maxillary central and lateral incisors of the underdeveloped side as a markers of development for hemifacial microsomia, which were present in this case. In HFM, hypoplasia of facial muscles such as the masseter, temporalis, pterygoids and those of facial expressions on the affected side has been observed (9). In the present case, the masseter and temporalis muscle were hypoplastic on palpation.

HFM patients may have minimal underdevelopment of the condyle to unilateral aplasia of the mandibular ramus and/or condyle with the absence of the glenoid fossa. There is narrowing of the maxilla on the affected side accompanied with decreased palatal width, which was seen in this case. Associated cleft lip and/or palate are found in 7% of hemifacial microsomia patients (11). In this case, there were an underdeveloped condyle and hypoplasia of the mandibular ramus. However, additional characteristics like absence of antegonial notch, reduced depth of the sigmoid notch and superiorly placed alveolar canal were absent. This is

DOI: http://dx.doi.org/10.4314/ejhs.v27i1.12

inconsistent with the literature. The severity of the clinical presentation depends upon the extent of loss of neural crest cells, which in turn reflects in the degree of severity(10).

In the most severe cases of hemifacial microsomia, there is underdevelopment of the external and middle ear, the side of the skull, the thickness of the cheek tissue, the upper and lower jaws and the teeth and some facial nerves (11). The absence of certain cheek muscles or nerves, which supply those muscles, can result in an asymmetrical smile, which was noticed in the reported case. Cheek tissue (fat and muscle) is often underdeveloped, which makes one side of the face fuller than the other.

Decision about the need and type of surgical correction depends upon age and growth status of the patient, severity of the skeletal deformity and soft-tissue deficiency (2). The objective of the treatment is to enhance the maxillofacial symmetry and stomatognathic function. The management of HFM involves multidisciplinary approach to craniofacial anomalies, which includes surgery done during or after growth phase; limited autogenous bone grafting of deficient portions of the craniofacial skeleton, bilateral mandibular jaw advancement in patients with mild to moderate mandibular micrognathia with a combined Le Fort I osteotomy. The genioplasty and microvascular free flaps can be used for augmenting the soft tissue of the affected side of the face and costo-chondral grafts, which can be used to provide a new growth center for treating this anomaly (2). The costo-chondral graft, however, has the no-growth spurt like the condyle; it grows at another rhythm (slower and irregular) independently from the healthy condyle. Overgrowth is often seen on the grafted side (12). Moreover, like in every surgical procedure where tissue has to be transplanted, there is always a risk of no acceptance of the graft.

Distraction osteogenesis is increasingly advocated in treating patients with HFM as it is considered a good alternative to the classical surgical interventions (like osteotomies and bone grafts) and its presumed positive effect on the soft tissue as claimed by the advocators (13). Therefore, distraction osteogenesis was considered the treatment of choice in the present case.

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DOI: http://dx.doi.org/10.4314/ejhs.v27i1.12