Case Report

# Hemifacial Microsomia in a Female Kid - A Clinico-Radiological report

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**ABSTRACT**

Otomandibular Auricular syndrome or Hemifacial Microsomia (HFM) is an asymmetric craniofacial malformation which results in hypoplasia of the components of the first and second branchial arches. HFM is the second most common congenital craniofacial anomaly after cleft lip/palate. The incidence of this disorder is 1:3000 26,000, which is usually seen at birth. Males are more commonly affected than females and right side is more affected than the left side. For the pre-surgical evaluation of this anomaly, diagnostic imaging and classification of the facial structures, based on OMENS classification, is of prime importance. The management of this developmental malformation is multidisciplinary. Here we report a case of Hemifacial Microsomia in a six-year-old female kid.

**Keywords:** Hemifacial Microsomia, Branchial arch syndrome, Condylar hypoplasia, Lateral facial dysplasia, Otomandibulardysostosis.

## INTRODUCTION

German physician Carl Ferdinand Von Arlt was the first person to describe Hemifacial Microsomia (HFM) in 1881. HFM comprises of unilateral microtia, malformation of the mandibular ramus, condyle with macrostomia.[1] Goldenhar syndrome is a variant of HFM, which includes vertebral anomalies and epibulbar dermoids. This is known as craniofacial microsomia when there is involvement of cranial deformities (Converse *et al*.). HFM is the second most common congenital facial anomaly following cleft lip/palate. The reported incidence cases of HFM are about 1 in 5600 live births.[2-4]

Other synonyms for HFM used in literature include First arch syndrome, Otomandibular-dysostosis (Francois and Haustrate, 1954), First and second branchial arch syndrome (Stark and Saunders, 1962; Grabb, 1965), Oculoauriculovertebral sequence (Gorlin et al., 1963), Goldenhar syndrome (Goldenhar, 1952; Gorlin et al., 1976), Lateral facial dysplasia (Ross, 1975), and Craniofacial microsomia (Converse et al., 1979). [5]

Here we report a case of Hemifacial Microsomia in a six year old female kid.

## CASE DESCRIPTION

A six year old female kid reported with a chief complaint of deposits and decay in her tooth since one year. Patient's mother gave a medical history of malnourishment and she did not breast feed the patient. Patient's mother observed the asymmetry of patient's face and ear, and there was difficulty in hearing since birth. Patient's mother gave no history of forceps delivery. Patient had undergone 2-D Echocardiogram and Doppler study at the age of 6 months which was normal. Audiogram done at the age of 4 years revealed right side moderate to severe conductive hearing loss for which the patient was advised an ENT opinion. Patient had already consulted a dentist, near their locality 2 days back for the same complaint as mentioned above and the doctor had advised for an orthopantamogram (OPG). Family history revealed first-generation consangious marriage for her parents.

General examination was normal. On extraoral examination, facial asymmetry was evident due to mandibular hypoplasia on the right side with frontal bossing along with deviation of angle of mouth towards the right side. [Figure 1a, 1b, 1d]

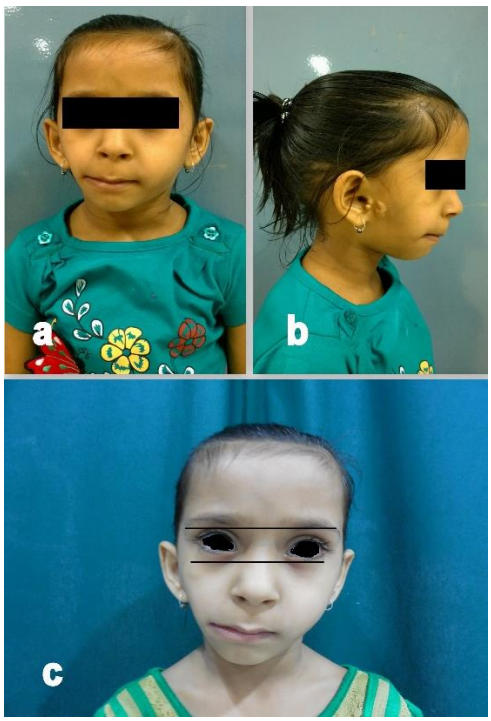


**Figure 1:**

1a: Evidence of facial asymmetry due to mandibular hypoplasia on the right side with frontal bossing.

1b: An accessory or a supplemental pinna or a pre auricular tag noted anteriorly to right tragus of the ear.

1c: Raised level of orbital floor on right side

Mandible appeared to be deficient on the right side and fullness of face on the left side. [Figure 1c] An accessory or a supplemental pinna or a pre-auricular tag was noted anteriorly to right tragus of the ear. [Figure1b] The accessory ear tag at right side was painless and firm. Mouth opening was normal and jaw movements were within normal limits with deviation of mandible towards right side during opening and closing of mouth. [Figure 2a]

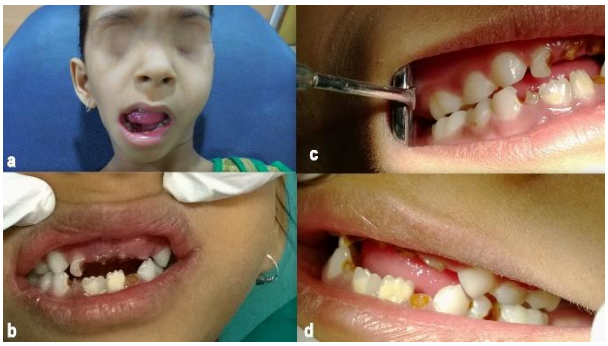


**Figure 2:**

2a: Deviation of mandible towards right side during

2b,2c,2d: Evidence of multiple grossly decayed and root stumps

No tenderness was elicited and TMJ movements were not palpable on the right side. No crepitus /clicking sounds. Masseter, temporal and pterygoid muscles were found to be hypoplastic on the affected side. Intraoral examination revealed multiple grossly decayed tooth and root stumps as shown in Figure 2b, 2c, 2d. On correlating all the positive findings, a provisional diagnosis of Hemifacial Microsomia of the right side, was made. Differential diagnosis included Condylar hypoplasia-unilateral, Coronoid hypoplasia-unilateral, Mandibulofacialdysostosis, and Treacher-Collin's syndrome.

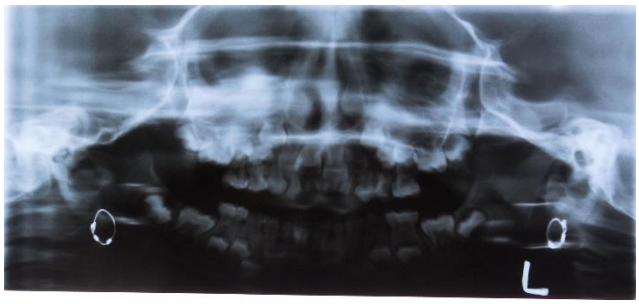

**Figure 3:** OPG showing dysmorphic ramus, condyle and coronoid process on the right side.

The OPG was studied and the patient was advised to take a Computed Tomography (CT). OPG revealed dysmorphic ramus, condyle and coronoid process on the right side as shown in Figure 3. CT revealed hypoplasia of right posterior condyle of the mandible with flattening of the rudimentary condyle, widening and flattening of the mandibular notch, complete opacifications of both maxillary sinuses, mucosal thickening at right ethmoidal and left sphenoidal sinuses, hypoplastic frontal sinus, bilaterally sclerosed mastoid air cells, bony lesion or an osteoma at right external auditory canal and hypoplastic middle ear ossicles at the right side as shown in Figure 4. The left side appears to be normal. On correlating the clinical and the radiographical findings, the final diagnosis was made as Hemifacial microsomia or OtomandibularAuricular syndrome.

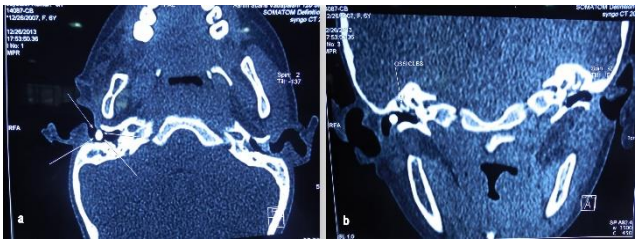


**Figure 4:**

4a: Computed tomographic images showing bony lesion or an osteoma at right external auditory canal

4b: CT showing hypoplastic middle ear ossicles at right side

## DISCUSSION

HFM is a complex malformation syndrome with genetic and teratogenic associations with abnormalities in facial skeleton and other organs. [5,6] The incidence of HFM is 1:3000-26,000 and females appear to be less frequently affected while compared to males (2:3) and the right side is affected more than the left side. [7] It is usually unilateral (70%). Ours is a case of a female kid. Although the term "hemifacial" refers to one half of the face, this condition may occur bilaterally in 31% of cases, in which one side is more severely affected than the other. However, this condition may be a part of some other syndromes like Goldenhar syndrome in 48% of cases. [7] The two most frequently used classifications for HFM are the skeletal–auricular–soft tissue (SAT) and the orbital asymmetry–mandibular hypoplasia–ear malformation–nerve dysfunction–soft tissue (OMENS) deficiency classifications. Among these, OMENS classification is the more comprehensive and, is one of the most commonly used systems.[7,8] In our patient, facial nerve dysfunction was absent. The diagnostic features were observed in clinical finding and in the CT of our patient. In this context, this case is an important addition to the existing literature.

The differential diagnosis of HFM includes Pierre Robin syndrome, Treacher Collins syndrome, and Parry–Romberg syndrome. However, Pierre Robin syndrome always consists of micrognathia, glossoptosis, and cleft palate, which were not seen in the present case. Features of Treacher Collins syndrome like "fishlike or birdlike" appearance of face were not seen in our case. Parry–Romberg syndrome (facial hemiatrophy) is characterized by progressive wasting of subcutaneous fat, sometimes accompanied by atrophy of skin, cartilage, bone, muscle, whereas in HFM, there is underdevelopment of one side of

the face primarily affecting the ear (aural), mouth (oral), and jaw (mandible).

Our patient was referred to department of Pediatric dentistry for management of her poor oral hygiene and multiple decayed tooth. The available surgical options were discussed with a multidisciplinary team. Surgery aims to correct facial asymmetry and restore function. The management should begin early in life and should involve a team of specialists which include geneticists, audiologists, speech pathologists, physicians, plastic surgeons, maxillofacial surgeons, pedodontists, prosthodontists, and orthodontists. At the age of 2–4 years, mildly affected children require no treatment. In cases where the mandible is severely underdeveloped, rib bone grafting or distraction osteogenesis is advised to lengthen the mandible, followed by orthodontic treatment to correct alignment. At the age of 6–8 years, external ear reconstruction can be done. At the age of 8–10 years, esthetic correction of the face is done like genioplasty and microvascualr free flaps, which is very crucial. During the teenage years, orthognathic surgery should be performed on those patients whose mild condition did not require treatment in early childhood. Mandibular growth that occurs in adolescence in severely affected patients may require further surgery. Therefore, these patients require both pre-.and post-surgical orthodontic treatment due to the surgical movement of the jaws. [9,10]

## CONCLUSION

Being a rare developmental malformation which is characterized by deficiency of skeletal and soft tissue components of the maxillofacial region on one side of the face, early diagnosis and early intervention improves proper functioning and esthetics which will aids in better results, better prognosis, fewer complications, smaller number of surgical re-interventions and an improved quality of life of patients.

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**Conflict of Interest:** None declared.