

Hemifacial microsomia: a clinicoradiographic report of a case

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Hemifacial microsomia is a rare congenital malformation of craniofacial structures. Its characteristic features are unilateral underdevelopment of the face and ear malformations. This study describes clinical and radiographical features of a rare case of a 4-year-old hemifacial microsomia patient with underdevelopment of the left side of the face and preauricular skin tags on the affected side. Management of these patients requires a multidisciplinary approach to provide the most effective and appropriate treatment.

Key words:

•**Hemifacial microsomia** •**Facial asymmetry** •**Congenital abnormalities**

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Case Report

A 4-year-old girl was referred to the Department of Pediatric Dentistry of Hamadan University of Medical Sciences complaining of toothache. Extraoral examination of the patient revealed underdevelopment of the left side of the face and unilateral preauricular skin tags. The frontal aspect of the face showed marked asymmetry with mandibular deviation to the left and the height of the left ramus was reduced compared to the right side. Mild mandibular hypoplasia was detected from the lateral facial aspect. On mouth opening, left mandibular deviation with no limitation in the degree of opening was recorded. On temporomandibular examination, a TMJ clicking sound was heard but there was no tenderness of the joint on palpation. Intraoral examination showed a midline deviation to the left side and severe rampant caries with pulpal involvement. Mesiodistal dimensions of the left posterior teeth were less as compared to the right side. Panoramic radiograph showed hypoplasia of the ramus of the mandible and a missing condyle and coronoid process on the left side. No missing teeth were found on radiography.

The family and medical history of the patient was unremarkable. The patient was then referred to an otolaryngology and surgeon ophthalmologist for further investigation. Hearing tests were normal. The skin tags were removed by the otolaryngology surgeon. Pathological study of the specimens showed the presence of cartilage in the skin tags. The above skeletal, mandibular, dental and dermatological features were suggestive of hemifacial microsomia.

Dental treatment included pulp therapy and restoration of the primary canines and molars, and extraction of the incisors was performed. A Nance esthetic appliance was placed in the maxillary arch. The patient was then referred to the Department of Orthodontics for functional therapy. However, due to a lack of cooperation because of her age, functional therapy was postponed until her cooperation could be achieved.

Discussion

Hemifacial microsomia (HFM) is a rare congenital and multisystemic defect characterized by

underdevelopment of one side of the face, including the maxillary and mandibular bone and soft tissue, and usually the ear is also involved.⁽¹⁾ Because these parts of the face embryologically derive from the branchial arches, this suggests that this anomaly is due to abnormal morphogenesis and development of the first and second branchial arches.⁽²⁾ Another term, Goldenhar syndrome, is also used for this anomaly but according to some authors, Goldenhar syndrome is characterized by some features in addition to the typical features of hemifacial macrosomia, such as vertebral anomalies and epibulbar dermoids.⁽³⁾ Most cases of HFM are unilateral, such as in our case, but bilateral microsomia is found in 10–33% of cases. The ratio of the right to the left side being affected is approximately 3:2.⁽⁴⁾ The etiology of this anomaly is still unexplained in many cases. It seems that HFM is more related to mechanical and environmental factors than genetic factors. Most studies suggest that this syndrome is due to abnormal vascularization of the first and second branchial arches during the 4th week of pregnancy. The grade of malformation from small to more complex anomalies seems to be associated with the extent of vascular damage.⁽⁵⁾ A positive family history is mentioned in some studies⁽²⁾ but no strong genetic risk factors have been found as a cause of this anomaly and the majority of HFM cases are sporadic.⁽⁶⁾ In our case, the family history was negative, which indicates that environmental factors may be responsible for the anomaly in this patient. Hypoplasia of one side of face and external ear malformations are the typical features of patients with HFM. Our patient had facial hypoplasia and preauricular tags on the left side of her face.⁽⁷⁾ The condyle of patients with HFM may have minimal underdevelopment to unilateral aplasia of the ramus and condyle, and agenesis of the ramus on the affected side occurs in 50% to 70% of HFM patients, such as in our patient.⁽⁸⁾ Some studies have shown that the mesiodistal dimensions of the primary and permanent molars of patients with HFM on the affected side are smaller than on the other side, but no differences were found between the incisors and canines. This suggests that the dental lamina in HFM is affected.⁽⁹⁾ In our patient, the crowns of the first primary molars were destroyed by car-

ies but the mesiodistal dimensions of the maxillary and mandibular second primary molars on the left side were smaller than the right side, which is consistent with the findings of previous studies.⁽⁹⁾

Treatment of patients with HFM needs a multidisciplinary approach and treatment varies according to the severity of the abnormality as well as the patient's age. The timing of surgical correction of HFM depends on the severity of the defects and the patient's treatment plan and expectations, and therefore the timing of surgery is still controversial. Some studies support early reconstruction by surgery, because some clinicians believe that by providing early surgery they will prevent secondary growth abnormalities. Otherwise, the mandibular asymmetry will worsen over time and also early treatment helps to improve the patient's quality of life.⁽¹⁰⁾ Some other studies suggest waiting for the completion

of growth, because some clinicians believe that after early surgery, asymmetric growth will continue and additional surgery will be required.⁽¹¹⁾ Surgical correction of defects in patients with feeding and swallowing problems should be done as soon as possible.⁽²⁾ In patients with jawbone hypoplasia, reconstruction can be performed using bone grafts and distraction osteogenesis. Orthodontic treatment begins with functional (hybrid) orthodontic appliances in children. It is then continued with fixed orthodontic appliances.⁽¹²⁾ In our case, a non-surgical treatment plan was chosen because the parents preferred it and the extent of deformity was not severe. Reconstructive surgery is needed to correct anomalies of the soft tissues and ears and to improve aesthetics and the patient's quality of life. The skin tags on our patient were removed by an otolaryngology surgeon for aesthetic reasons.

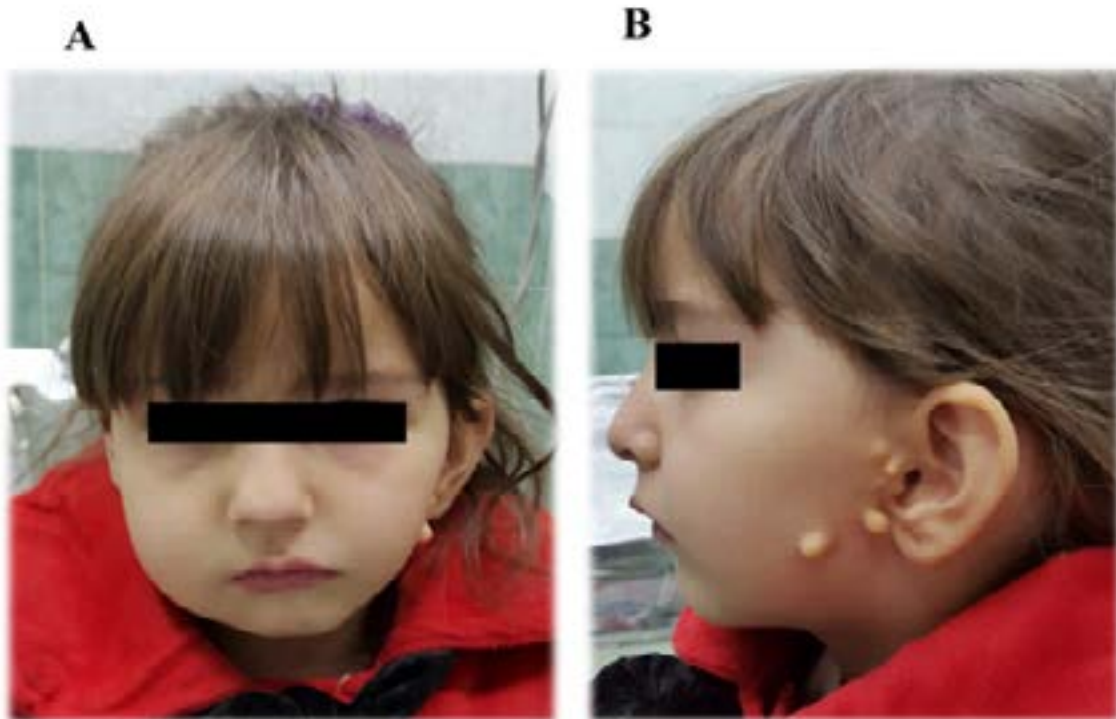


Figure 1. Frontal and lateral views of the patient. (A) Frontal view of the face showing marked facial asymmetry, (B) Lateral view of the face showing preauricular skin tags.



Figure 2. Panoramic radiograph of the patient showing severe mandibular hypoplasia on the left side associated with a hypoplastic temporomandibular joint.



Figure 3. Intraoral photographs revealing rampant caries before dental treatment.



Figure 4. Intraoral photographs after dental treatment. A Nance esthetic appliance was placed in the maxilla.

Conclusion

Management of HFM requires a multidisciplinary approach to provide the most effective and appropriate treatment and the prognosis depends on the severity of the malformations. Dental care with consultation with orthodontists and maxillofacial surgeons is necessary. Structural anomalies of the ears can be corrected by reconstructive surgery. The treatment usually begins at early ages and long-term follow up is required.

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