

Non-Syndromal Hemifacial Microsomia in Two Iraqi Children

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1. Abstract

Hemifacial microsomia is a rare congenital, heterogeneous malformation disorder affecting predominantly unilateral face and involving head malformations mainly in the region of the first and second branchial arch and varying associated malformations. The disorder is associated with unusual strange distortion of the face with facial asymmetry and may cause psychological problems in the affected child and even the parents.

Hemifacial microsomia is best managed by a multidisciplinary team including the otolaryngologist, audiologist, plastic surgeon and temporal bone radiologist. However, physicians working in many geographic areas of the world lacking active effective multidisciplinary teams and major craniofacial centers, will face a serious challenge and a tough time in making the appropriate referrals which ensure satisfactory management. The complexity of the defects in this condition, the potentially non-static nature of the condition and the lack of extensive management experiences with such rare condition make management challenging even when the appropriate timely referrals can be ensured.

The aim of this paper is to report the occurrence of non-syndromal hemifacial microsomia in two unrelated Iraqi children for the first time.

2. Keywords: Non-syndromal; Hemifacial

microsomia; Iraqi children

3. Introduction

Hemifacial microsomia is a congenital and heterogeneous complex lesion disorder affecting predominantly unilateral face and involving head malformations mainly in the region of the first and second branchial arch and varying associated malformations. The condition affects the development of the lower half of the face, most commonly the ears, the mouth and the mandible [1].

The condition has also been called Oral-mandibular-auricular syndrome (Stark and Saunders, 1962), first and second branchial arch syndrome [2-4] (Edgerton and Marsh, 1977), Otomandibular dysostosis and dysostosis otomandibularis [3], Lateral facial dysplasia [4], Asymmetrical otocraniofacial syndrome and hemifacial microsomia (Pérez Alvarez F et al, 1984), Oto-mandibular syndrome (Fasano et al, 1985). Craniofacial microsomia (Munro, 1987) [1-5].

The condition usually occurs on one side of the face, but both sides are sometimes affected. Severe cases can be associated with difficulties in breathing because of obstruction of the trachea and may need

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sometimes tracheotomy. The disorder shares some similarities with Teacher Collins syndrome. The condition arises at about four weeks of gestational age as a result of vascular problem such as blood clotting causing insufficient blood supply to the face restricting its development. The initiating factor could be physical trauma, but hereditary factors have also been liked [1].

Hemifacial microsomia is associated with variable clinical manifestation depending on the severity. The severity of the condition depends on the size of the area affected by intra-uterine reduction of blood supply and the gestational age of the fetus. Mild cases may be associated only with a small and underdeveloped external ear. Severe cases can involve multiple parts of the face. Goldenhar syndrome is probably a severe form of hemifacial microsomia associated whit extra-cranial anomalies [1,6].

The aim of this paper is to report the occurrence of non-syndromic hemifacial microsomia in two unrelated Iraqi children for the first time.

4. Case Presentation

The first Iraqi patient with hemifacial microsomia was a girl who was first seen at about the age of five with parents main complain being abnormal shape of the face (Figure 1). The girl didn't have other abnormalities.



The second Iraqi patient with hemifacial microsomia was a boy (Figure 2) who was first seen at about the age of eighteen months during the middle of April, 2018. The parents main complain being poor hearing.



The boy had cleft palate which was operated before four months and delayed speech. The boy also has large mouth (Macrosomia) and low set ears. The boy had mild developmental delay as he was unable to walk alone, but was able to walk holding furniture with some difficulty.

5. Discussion

Hemifacial microsomia is a rare congenital, heterogeneous malformation disorder affecting predominantly unilateral face and involving head malformations mainly in the region of the first and second branchial arch and varying associated malformations. Hemifacial microsomia is associated with unusual strange distortion of the face with facial asymmetry and may cause psychological problems in the affected child and even the parents. Hemifacial microsomia is best managed by a multidisciplinary team including the otolaryngologist, audiologist, plastic surgeon and temporal bone radiologist. However, physicians working in many geographic areas of the world lacking active effective multidisciplinary teams and major craniofacial centers, will face a serious challenge and a tough time in making the appropriate referrals which ensure satisfactory management. The complexity of the defects in this condition, the potentially non-static nature of the condition and the lack of extensive

management experiences with such rare condition make management challenging even when the appropriate timely referrals can be ensured [1].

6. Conclusion

The occurrence of non-syndromal hemifacial microsomia in two unrelated Iraqi children for the first time.

7. Acknowledgement

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