

Hemifacial Microsomia: Need Of A Dentist Can't Be Denied.

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Abstract: Hemifacial microsomia is a common craniofacial malformation resulting in oral, facial and ocular manifestations. The condition is relatively rare with important dental changes. The unclear aetiology with variable presentation makes it an interesting case study. This case report attempts to discuss the clinically relevant dental findings in hemifacial microsomia patient with management of the same.

Key words: Craniofacial, Goldenhar syndrome, Malformation, Microsomia.

I. Introduction

Hemifacial microsomia is the second most common craniofacial malformation following cleft lip and palate. The term Hemifacial microsomia was first described by Gorlin for conditions characterized by unilateral microtia, failure to form ramus and condyle of mandible and macrostomia¹. Owing to its diverse clinical presentation various names have been given to it such as Goldenhar gorlin syndrome², lateral facial dysplasia³, otomandibular dysostosis⁴ and oculoauriculovertebral spectrum⁵.

II. Case Report

A 17 year old girl reported to the OPD of Dr Ziauddin Ahmed dental college, AMU with chief complaint of asymmetric face. Patient also had a complaint of difficulty in chewing. Patient has once visited a plastic surgeon for treatment of defective ear. No treatment was rendered to her by plastic surgeon. Patient gave no history of any such disease in other family members. No antenatal drug history or history of trauma was found. History of a dental filling was given by patient. No other dental treatment history was found.

Upon extraoral examination asymmetric face with deviation towards right side was found (Figure 1). Flattening of face towards right side and underdeveloped ear pinna was also noted (Figure 2). Incompetent lips, inclination of lips and fullness of face towards left side was noted (Figure 3). Upon palpation underdeveloped ramus and condyle on the right side was noted. Upon intraoral examination posterior open bite on the right side and inclination of cant of occlusion was noted (Figure 4). Absence of maxillary first molar and mandibular second molar on the right side was also noted.

Oral hygiene of the patient was not adequate with deposition of plaque and calculus on the hard tissue surfaces. There was also narrowing of palate and decreased palatal width on right side. A Panoramic radiograph revealed underdeveloped ramus of mandible and condyle (Figure 5)



Figure 1



Figure 2



Figure 3



Figure 4



Figure 5

III. Discussion

Incidence of 1 per 3500 births has been suggested by Poswillo⁶. The ratio of male to female occurrence is 3:2 with 3:2 involvement of Right side vs left side⁷⁻⁹.

The exact etiology and pathogenic mechanism is yet not clear but it appears to be related to genetic and teratogenic components¹⁰⁻¹⁴. According to poswillo hematoma in the region of developing stepedial artery may cause necrosis in first and second arch branchial derivatives which may cause hemifacial microsomia¹⁵. Familial incidences in the patients of hemifacial microsomia has also been reported which suggests chromosomal involvement in the etiopathogenesis of the disease¹⁶. Autosomal dominant and autosomal recessive inheritance patterns have also been hypothesized to explain familial occurrence of the same¹⁷⁻¹⁸.

Although Hemifacial microsomia has extremely variable range of expressions, commonly it is recognized by facial asymmetry¹⁹ due to the underdeveloped mandible and malformed pinna. Maxillary, temporal and malar bones are flattened and reduced in size. Intraoral deformity includes hyperplastic or aplastic teeth and enamel with delayed and missing dentition on the affected side. Patients may have minimal to complete aplasia of mandibular condyle and/or ramus of the mandible²⁰.

Hypoplasia of facial muscles has also been reported with narrowing of palpebral fissures in about 10 percent cases²¹. Unilateral coloboma of the upper eyelid is a common finding.

Hemifacial microsomia is a progressive skeletal and soft tissue deformity which require a comprehensive health care. Hypoplastic mandible interferes with normal growth of maxilla and as the contralateral side grows, it produces secondary distortion of maxilla, nose and orbit. Principles of timings for surgical reconstructions of facial deformity are based on age, psychosocial aspects and severity of the condition. Surgical reconstruction of deformed facial bones, reconstructive ear surgery, orthodontic treatment and restoration of missing dentition are the most demanding challenges with the patients of hemifacial microsomia. Tissue engineering may proved to be of potential clinical application in the surgical reconstructive needs of patients of hemifacial microsomia in future.

IV. Conclusion

This case report is important because of the rarity of this condition and frequently undiagnosed or misdiagnosed cases. Many times patients visits a plastic surgeon or an otorhynolaryngologist where the diagnosis may be made correctly but dental component of the disease goes frequently unnoticed. Dental management of such patients is of paramount importance for functional and esthetic reasons. Proper counselling and stepwise approach in the treatment of such cases is required with a team of specialists from different fields. Reporting of such cases may propagate the word further which in future may result in better diagnosis and treatment of patients of hemifacial microsomia.

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