



True Hemifacial Hyperplasia-Clinical Presentations and Treatment Options

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Abstract

Congenital hemifacial hyperplasia (CHFH) is a rare congenital malformation characterized by marked unilateral overdevelopment of the hard and soft tissues of the head and face region. The exact etiology is not clear; it is usually evident at birth and increases with age, mostly till puberty. In some cases multiple systems are involved. Treatment is usually indicated in cases where esthetics is a concern.

A 6 year old female reported to the Department of Oral Medicine and Radiology with the features of congenital hemifacial hyperplasia, we are making an attempt to present a rare case of CHFH in order to highlight the clinical manifestations and treatment options.

Keywords: Asymmetry; Hemifacial hyperplasia; Hypertrophy; Congenital.

Introduction

Hemifacial hyperplasia or hypertrophy is a condition in which one half of the face either maxilla or mandible alone or soft tissues of one half of the face grows to unusual proportion as compared to the opposite side. It is a rare congenital malformation. This condition has been termed as facial hemihyperplasia, partial / unilateral gigantism, hemimacrosomia, or hemifacial hypertrophy [1].

CHFH was first noted by Meckel JF in 1822, as a developmental anomaly and the first case was reported by Wagner in 1839[2]. Hyperplasia can be seen in association with several syndromes like Beckwith Weidemann syndrome, Proteus syndrome, Russell silver syndrome and Sotos syndrome. Gessel in 1927 described CHFH as essentially a developmental anomaly antedating birth. Rowe[3] in 1962 proposed a classification of hemifacial hyperplasia based on its anatomical location as simple (one or both limbs), complex (entire half of the body), and facial hemihyperplasia(face, head and associated structures). Based on soft tissue involvement HFH can be classified as true (increased growth of not only the soft tissues of the face but the hard tissues as well) and partial (increased growth limited to one structure only) [4,5].

We present an interesting case of a 6 year old female with a true congenital hemifacial hyperplasia.

Case Report

A 6 year old female patient reported to the Department of Oral Medicine and Radiology with the chief complaint of a large upper left front tooth since six months. Patient's mother was concerned about the esthetics. They had noticed an enlargement of the left side of the face since birth, which has been progressing in size. Patient had an early exfoliation of the upper left deciduous tooth which was followed by an early eruption large sized permanent tooth. The child was the second of two siblings born of a non-consanguineous marriage. There was no significant prenatal or relevant medical history. Milestones were normal and she has normal intelligence. Patient was moderately built and nourished with normal gait. The vitals were within normal limits.

Extraoral examination revealed an obvious swelling of the left upper half of the face, leading to asymmetry extending superiorly from the infra orbital rim to the left upper lip inferiorly. Medially extending from the bridge of the nose, obliterating the nasolabial fold and laterally 2 cm anterior to the pretragal area on the left side. The left half of the upper lip was hypertrophied and the angle of the mouth on the left side was tilted downwards lip was incompetent. The overlying skin appeared normal. On palpation the swelling was soft in consistency, non-tender, with no rise in temperature (Figure 1).



Figure 1: Extraoral Photograph.

Intraoral examination revealed premature eruption of 23, 24, 27 with macrodontia of 23. Grossly decayed 54, 65, 74, 85 and 26. Dentinal caries in 53, 62, 75, deep carious lesion in 84, rest of the dentition was normal with age. Thickened and enlarged alveolar ridge extending from 23 till 27. Mandible was normal. No midline shift was noticed. Buccal mucosa and tongue appeared normal (Figures 2,3).



Figure 2: Macrodontia of 13.



Figure 3: Enlarged left alveolar ridge.

Considering the history and clinical features a provisional diagnosis of a congenital true hemifacial hyperplasia of the left side was made, and a differential diagnosis of segmental odontomaxillary dysplasia was given.

An orthopantomogram showed early exfoliation of 63, 64 followed by early eruption of 23, 24 and 27. Enlarged pulp chamber of 23, 24 and 27. Incomplete root formation in 23, 24 and 27. 65 also shows resorbed roots and half the root formation of 25 as compared to the opposite side where only the crown formation is complete. Slanted palatal drag suggestive of hyperplastic maxilla and hyperplastic zygomatic bone on the affected side. The left maxillary sinus appears to be enlarged as compared to the right side. There was no obvious enlargement of the left mandible including coronoid and the condylar process (Figure 4).



Figure 4: Panoramic radiograph.

Both the clinical and radiographic features suggested congenital true hemi-hyperplasia on the left side.

Patient was then referred to the department of pedodontics for detailed examination and management. They advised full mouth oral prophylaxis, extraction of all the teeth with grossly destructed crown structure which includes extraction of 54, 74, 84, 85 followed by placement of space maintainer. Since the prognosis of 26 was poor they advised extraction of 26 also. Pit and fissure sealants and periodic follow up was also advised. Since radiograph shows unerupted 21 and 22 due to lack of space patient was advised orthodontic treatment followed by esthetic recontouring of 23. Once the growth spurts are over patient was advised non-surgical management for the facial asymmetry.

Discussion

CHF is a rare developmental anomaly characterized by an increased growth of both hard and soft tissues of the face present since birth. The prevalence of HFH is approximately 1 in 86000 births [2]. In some cases it is associated with the syndromes of head and neck region. It usually increases with age till puberty. It is usually seen more commonly among females as compared to males mostly on the right side of the face [6]. Many theories have been proposed as a cause of HFH including hormonal imbalances, diseases involving the neural system, other conditions like hemangiomas, lymphangiomas, incomplete twinning, abnormal intrauterine development, somatic mutations, mechanical influences and congenital syphilis [7]. Numerous clinical findings may be seen like gross asymmetry of the facial structures and presence of hypertrophied areas. The soft tissues usually have the same consistency as that of the normal tissues. In some cases unilateral macroglossia and prominent fungiform papillae are also seen.

Permanent canine, bicuspid, and first molars are most commonly involved. Premature exfoliation and early eruption of the teeth of the affected side is noticed. In most cases teeth have large crown size (macrodontia) and large root with root resorption. Midline shift and malocclusion with deviated occlusal plane is also noted. Involvement of the upper lip causes displacement of the philtrum [8,9]. Palate can also show an arch shaped deformity of the affected side. Differential diagnosis includes fibrous dysplasia, dyschondroplasia, hemangioma,



lymphangioma, A-V aneurysms, congenital lymph edema and odontomaxillary dysplasia[10]. HFH may be associated with skeletal deformities like polydactyly, syndactyly, scoliosis and macrodactyly. CNS defects like epilepsy, mental retardation affects 20% of the affected patients. HFH may also be associated with adrenal cortical carcinoma, Wilm's tumor and hepatoblastoma [11].

Treatment Options

A multidisciplinary approach is needed in most of the cases, includes subtle soft tissue contouring to extensive surgeries to correct the soft and hard tissue defects. Like condylar recontouring, osteotomies followed by debulking of the soft tissues [12]. Khanna and Andrade did hard and soft tissue debulking to achieve acceptable results[13]. Pollock also did osteotomies and treatment of soft tissue deformities like cheiloplasty [14]. Occlusal irregularities were corrected by orthodontic therapy. Due to the difficulties encountered in the treatment like aggravating growth, suboptimal post-operative esthetics has dissuaded many surgeons from approaching the condition in an aggressive manner. Liposuction the tumescent technique has been widely used which was introduced by Klein [15]. The other technique includes ultrasound assisted liposuction, powered liposuction. Phosphatidylcholine preparations have been widely used for localized reduction of subcutaneous fat. Once injected into the site the reaction will remain for 8 to 10 weeks [16]. Despite temporary discomfort Phosphatidylcholine injections can be successfully used in the treatment of localized fatty areas of the face as it is safer than liposuction.

Dental abnormalities like macrodontia, premature exfoliation, premature eruption and malocclusion can be managed according to the abnormalities. Macrodontia with open apex can be treated once the root completion is over either by coronoplasty or crown placement. Premature exfoliation leading to closure of space can be managed with space maintainers. Malocclusion can be orthodontically corrected. Carious formation due to the premature eruption can be treated prophylactically by pit and fissure sealants and fluoride applications.

Conclusion

To conclude this article highlights the importance of a thorough case history, clinical examination and diagnostic evaluation in oral medicine for the proper diagnosis and treatment planning of such congenital developmental disturbances. Modern technology has revolutionized treatment options, in rare conditions like this the possibility of achieving facial symmetry is not an easy task, and it has to be done with a multidisciplinary approach. Non-surgical management is considered a better treatment option as compared to cosmetic surgeries.

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