ABSTRACT
Asymmetric variations of the contralateral structures of the head and face occur commonly in the general population and are an accepted feature of morphogenesis. A gross asymmetry characterized by marked unilateral overdevelopment of hard and soft tissues of head and face is a rare congenital malformation and has been termed as hemifacial hypertrophy (HFH).

The etiology is unknown, but several theories have been proposed including hormonal imbalances, diseases involving the neural system, vascular conditions, lymphatic abnormalities, mechanical influences and congenital syphilis. Generally, treatment is not indicated for HFH unless cosmetic considerations are involved.

An 8 years old boy reported to the Department of Oral Medicine and Radiology with the characteristic features of HFH. The case is presented to supplement existing clinical knowledge.

Keywords: Facial hemihypertrophy, Hemifacial hyperplasia, Asymmetry, Congenital anomaly.

INTRODUCTION
Hemifacial hypertrophy (HFH) is a condition in which half of the face—maxilla alone, the maxilla with the mandible, or half of the faces in concert with other parts of the body—grows to unusual proportions. Asymmetric variations of the contralateral structures of the head and face occur commonly in the general population and are an accepted feature of morphogenesis. Occasionally, a gross asymmetry easily perceptible to the eye may occur either due to physiologic reasons or due to a growth abnormality. One such entity, characterized by marked unilateral overdevelopment of hard and soft tissues of head and face is a rare congenital malformation. This has been termed variously in literature as HFH, partial/unilateral gigantism, hemifacial hyperplasia.1

Hemihypertrophy was first documented by Meckel in 18222 and was described as essentially a developmental anomaly antedating birth and arising in some way as a partial deflection of the normal process of birth.3 Asymmetric enlargement could be manifested in a unilateral/crossed configuration and may involve all the body tissues in the area (i.e. total) or a single tissue.4

HFH is classified as true and partial hemihypertrophy. True hemihypertrophy involves not only the soft tissues of the body but the hard tissues as well. It may be classified as being total or partial. Total, if an entire side of the body is involved and partial, if only a portion. Some cases are associated with genetic diseases such as Beckwith-Wiedemann syndrome.5

The purpose of report is to present the case history of an 8 years old boy child with hemifacial hyperplasia to supplement existing clinical knowledge.

CASE REPORT
An 8 years old boy was referred to department of oral medicine and radiology with the chief complaint of painless swelling of the right side of the face since birth and large size of teeth in right upper back teeth region.

The swelling is increasing in size with age causing gross facial asymmetry. Patient had early exfoliation of deciduous teeth in the right upper back tooth region and the erupting permanent teeth are very large in size causing disturbed occlusion.

There was no history for extraction of any tooth. The child was the second of the two siblings born of a non-consanguineous marriage, at full term by normal delivery. No significant postnatal illness was reported. The developmental milestones were normal. Child was of normal intelligence, doing well at school. There was no history of similar complaints in the family. General examination showed, patient was having normal gait, average built, moderately nourished. All the vital signs were in normal limits.

Extraoral examination of the patient revealed an obvious asymmetry of the right side of the face over the cheek region. The right half of the upper lip was hypertrophied and angle of mouth is tilted downwards (Fig. 1). On palpation, zygomatic bone of right side was found hyperplaslic.

Fig. 1: Extraoral photograph
A diffuse swelling was located on right side of face over cheek region. Swelling extends superiorly from infraorbital margin of right side and inferiorly 2 mm below the inferior border of mandible. Medially 2 mm behind the philtrum of right side of upper lip and laterally 3 cm in front of pretragal area. On palpation swelling was soft in consistency, non tender and with no local rise in temperature over it. No pulsations were appreciated over the swelling.

Intraoral examination showed the presence of precocious permanent dentition with macrodontia in right upper quadrant—13, 14, 15 and 16 (Fig. 2). Pit and fissure caries was found in relation to 16 and deep dental caries with respect to 84. Rest of dentition was normal. Thickened and enlarged palate along with hyperplastic maxilla on right side was appreciated. Mandible was normal. A midline shift was evident along with malocclusion on right side. Buccal mucosa and tongue appeared normal.

Considering history and all the clinical features the provisional diagnosis of congenital HFH was made and the differential diagnosis of segmental odontomaxillary dysplasia, lipoma in the right cheek region, arteriovenous aneurysms, congenital lymphedema, Proteus syndrome, Beckwith-Wiedemann syndrome, Silver’s syndrome was considered.

Intraoral periapical radiograph of 13 and 14, 15 and 16 regions showed macrodont 13, 14, 15 and 16. Enlarged pulp chambers of right upper canine and right upper first molar were noticed.

An orthopantomogram showed advanced dentition development on the right side of upper jaw with erupted 13, 14, 15 and 16 teeth. There was accelerated root formation of all the right upper teeth. Slanted palatal drag suggestive of hyperplastic maxilla and hyperplastic zygomatic bone was observed on right side (Fig. 3).

PNS radiograph showed enlarged maxillary sinus of right side (Fig. 4). PA view of the skull showed normal study except an obvious enlargement of the maxilla on the right side and midline deviation (Fig. 5). Magnetic resonance imaging (MRI) report was suggestive of well defined benign soft tissue mass lesion in right cheek suggestive of lipoma (Fig. 6).

Hematological and serological values were within normal limits. Urinary gonadotropin hormone levels were normal. Ultrasound examination of abdomen does not reveal any abnormality in the kidney, liver, intestine, genitourinary tract, etc.

Based on the findings of the investigations, intraoral incisional biopsy was done and histopathological examination revealed fibrolipomatous lesion.

After considering both clinical and radiographic features the final diagnosis of congenital HFH was made.

DISCUSSION

HFH is a rare developmental anomaly exhibiting asymmetric growth of one or more body parts and is usually referred to as hemihypertrophy. In some instances, the hyperplasia may be isolated or may be associated with a variety of multidimensional and variably facetted complicated syndromes. The condition is usually accentuated with age, especially around puberty. The disorder occurs more commonly in females with ratio 3:2 and the right side of the face is usually affected.

HFH may be associated with other conditions, such as acromegaly and pituitary gigantism, or with hypertrophy of other parts of the body. Many theories and postulates have been proposed as a cause for HFH, including hormonal imbalances, diseases involving the neural system, vascular conditions such as hemangiomas, lymphatic abnormalities such as lymphangioma, incomplete twinning, abnormal intrauterine environment, somatic mutations, mechanical influences, and congenital syphilis.

True HFH is an unusual condition which produces facial asymmetry by a marked unilateral localized overgrowth of all the tissues in the affected area, i.e. facial soft tissues, bone and teeth. The unilateral enlargement of the viscerocranium is bounded by frontal bone superiorly (not including eye), inferiorly by the border of mandible medially.
by midline of the face and laterally by ear, pinna being included within hypertrophic area.9

The asymmetry is almost always evident at birth and the enlarged side generally grows at a rate slightly faster than normal side until the skeletal maturation. Numerous clinical findings may be seen, of which the more common include gross asymmetry of the facial structures with occasional resemblance to hydrocephalus and the presence of edematous hypertrophied areas. Soft tissues that are involved may seem to have normal consistency with few exceptions, together with thickened hair, thickened skin, ipsilateral nevi and telangiectasias, asymmetric tongue, macroGLOSSIA featuring enlarged and prominent fungiform papillae and there also may be enlargement of other oral soft tissues.10

Radiographically, the mandibular canal may be increased in size. Some of the cases exhibit larger crowns of the permanent cuspids, premolars, and first molar teeth on the affected side. In addition, precocious eruption and premature development of these involved teeth is not unusual. Teeth exhibit larger root sizes and in some cases root resorption is seen. Occasional malocclusion with open bite and trismus with or without unilateral or bilateral ankylosis.11 HFH may sometimes be associated with deformities of the skeletal system like macrodactyly, polydactyly, syndactyly, ectrodactyly, scoliosis, tilting of the pelvis and clubfoot. Central nervous system defects may include cerebral enlargement, epilepsy, strabismus and mental retardation in 15 to 20% of the affected persons, and in some cases dilatation of the pupils on the affected side.12 This disorder has been associated with adrenal cortical carcinoma, nephroblastoma (Wilms’ tumor) and hepatoblastoma.13 Occasionally genitourinary system disorders like hypospadias, cryptorchidism and medullary sponge kidney are also observed.14

Generally, treatment is not indicated for HFH unless cosmetic considerations are involved. Therapy includes soft tissue debulking by excision of excess masticatory and subcutaneous tissues, with preservation of neuromuscular functions. Reconstructive procedures like osteotomies or orthognathic surgeries are planned when physiological growth ceases. Other options include facelifts and orthognathic surgical procedure in conjunction with orthodontic therapy.15

HFH is generally associated with good prognosis with no reports of malignant degeneration.16 There may be severe psychological sequelae due to the facial deformity, but little or no associated morbidity is featured. Considerable patient cooperation is warranted because, as reported in the literature, surgical correction usually involves extensive
maneuvers and stepwise, arduous, slow, long-standing therapy is anticipated.

MANAGEMENT

A multidisciplinary approach was planned regarding the surgical removal of buccal pad of fat on right side that was causing the facial asymmetry and unesthetic appearance. Patient’s parents refused to get any invasive procedure to be performed. Pit and fissure caries with respect to 16 was treated with preventive resin restoration. Stainless steel crown was given, after doing pulpotomy of 84. Patient’s parents were advised to give follow-up visit of child every 2 to 3 months, to check the status of growth of mandibular condyles. The orthodontic intervention was planned to treat the occlusal asymmetry, on follow-up visits.

CONCLUSION

The importance of a thorough diagnostic evaluation in oral medicine and radiology cannot be overstated. However, the timing and sequence of treatment in such disorders is effective with a multidisciplinary team approach rather than restricting the child patient to a single specialist.

REFERENCES


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