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Case Report

Congenital Hemifacial Hypertrophy: A Case Report

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Abstract

It is a rare developmental anomaly characterized by progressive and marked unilateral facial tissue changes, which involves the soft tissues, facial bones, and the teeth. It is observed at birth and the growth ceases after puberty with pronounced facial asymmetry with an unknown etiology.

Keywords: Congenital hemifacial hypertrophy, developmental anomaly, facial asymmetry

Introduction

Symmetry is an important concept in biology, being related to health, mate selection strategies and ultimately the survival of the species. Although deviations from symmetry are critical perceptual units in detecting the appearance of health, the natural subtle asymmetry of the human face may be relatively unimportant for judging facial attractiveness.[1,2]

The affected side grows at a rate proportional to the unaffected side, such that the disproportion is maintained throughout life. Etiology for Congenital Hemifacial Hypertrophy (CHH) includes heredity, chromosomal abnormalities, atypical twinning and intrauterine environment.

Rowe (1962) described CHH as "an unusual condition that produces facial asymmetry by a marked unilateral localized overgrowth of all the tissues in the affected area, which is the facial soft tissues, bones, and teeth."[3] It is characterized by viscero-cranial enlargement bounded by the frontal bone superiorly (sparing the eye), inferior border of the mandible inferiorly, midline medially and ear including the pinna laterally.[3,4,5] In partial hemifacial hypertrophy not all structures are enlarged to the same degree or are limited to one structure. Rowe classified true CHH into the following three types:

Type I: Simple hemifacial hypertrophy—one or both the limbs
Type II: Complex hemifacial hypertrophy—the entire half of the body are affected
Type III: Hemifacial hypertrophy—the face, head, and associated structures are involved on one side

Hemifacial hypertrophy may be associated with a wide variety of abnormalities such as thickened skin and hair on the involved side, excessive secretion of sebaceous, sweat glands, vascular and pigmentary defects of the affected side.[5] Different textures and color variances of ipsilateral scalp hair have also been reported. In addition, skeletal abnormalities such as macrodactyly, polydactyly, syndactyly, ectrodactyly, scoliosis, tilting of pelvis, and clubfoot have also been described.[5]

Case Report

A 10-year-old boy reported to Geetha Multispeciality Dental Clinic, 5th main Road, near nagasandra circle, tata silk farm road, Basavanagudi Bangalore, with the chief complaint of pain in the lower right region of the mouth.

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Intraoral examination revealed dental caries involving the pulp of lower first right molar for which he was treated. The patient gave a history of abnormally enlarged right side of the face since birth, which became more prominent progressively. Taking into account of the patient's clinical features, he was diagnosed of Congenital Hemifacial Hypertrophy; however, no genetic studies were carried out to further reconfirm the diagnosis. The patient was born to a mother from non-consanguineous marriage by full-term normal delivery, he was normally built with an intelligence quotient ranging between 80 and 89 in the lower normal range.[6]

Extraorally the patient showed a diffuse soft and extended swelling, extending up to the lower border of the mandible on the right side with a significant development of the symphysis region [Figure 1]. Patient was in the mixed dentition phase with angles class III molar relation on right side , angles class I molar relation on left side ,class I canine relation on the right side, and unerupted upper lower canines on left side, anterior incisor cross bite and pulpally involved 46 [Figure 2]. The right involved side showed a rapid eruption of the teeth with broader basal jaw bones when compared to the left uninvolved side with delayed eruption of the canines and premolars. Molar crowns were larger on the right side than those on the left side.

Grummons analysis: It showed a marked facial asymmetry with the chin deviated to the unaffected left side.[7] Increase in the length and height of the body of the mandible was seen on the right side of the face. Upper facial ratio was decreased (cg-ANS/cg-me = 43%), lower facial height was increased (ANS-Me/cg-me = 60%), overall maxillary ratio was slightly increased (ANS-A1/ANS-me = 57%), and overall mandibular ratio showed a remarkable increase (me-B1/ANS-me = 80%)[6] [cg—crista galli, me—menton, ANS—anterior nasal spine, A1—upper central incisor edge, B1—lower central incisor edge] [Figure 3A and 3B].

Lateral cephalogram shows a negative angle of 4 degrees formed by joining point A, point B and Nasion angle decreased lower incisor mandibular plane angle of 82°, and mandibular plane angle of 142°, showing a vertical growth pattern [Figure 4A and B].

Discussion

CHH is a rare developmental anomaly, characterized by asymmetric overgrowth of one or more body parts. Malformation syndromes associated with hemifacial hypertrophy are Beckwith–Wiedemann syndrome, neurofibromatosis, Klippel–Trenaunay–Weber syndrome, Proteus syndrome, McCune–Albright syndrome, epidermal nevus syndrome, Langer–Giedion syndrome, multiple exostoses syndrome, Maffucci syndrome, and

Figure 1: Extraoral photograph of the patient

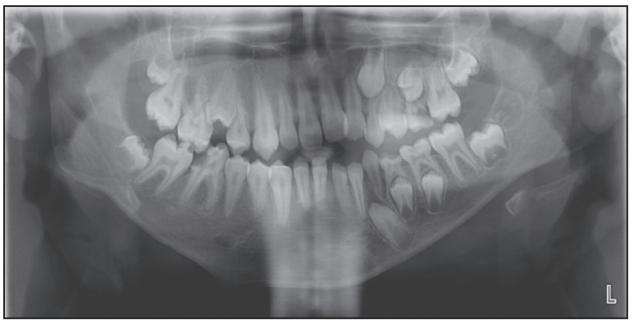

Figure 2: Orthopantomogram of the patient

Ollier syndrome.[5,8] Hoyme *et al*. anatomically classified hemifacial hypertrophy into complex hemihyperplasia, simple hemihyperplasia, and hemifacial hyperplasia.[8,9]

The etiology is unknown with vascular, lymphatic, central nervous system (CNS), or chromosomal abnormalities. Clinical features include enlargement, which is confined to one side of the body, unilateral macroglossia, premature development and eruption, as well as increased size of the dentition with permanent teeth on the affected side often show more than 50% enlargement. The maxillary and mandibular bone show enlargement with altered bony trabeculae.[8] The involvement of CNS in the form of cerebral enlargement, epilepsy, strabismus, and mental retardation in 15%–20% of patients has been reported in the literature.[5] Adrenal cortical carcinoma, nephroblastoma (Wilm's tumor), and hepatoblastoma can be occasionally associated with this disorder. Genitourinary system disorders (such as hypospadias, cryptorchidism, and medullary sponge kidney) were also noted occasionally.[5]

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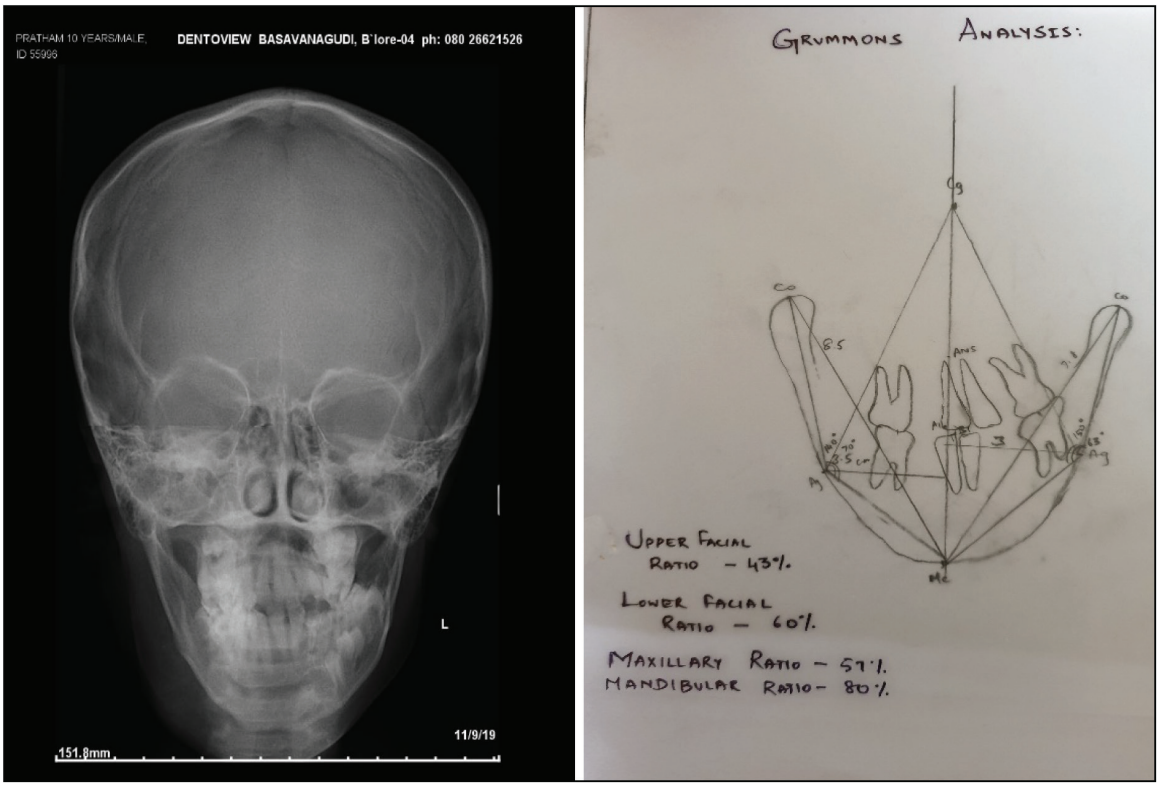

Figure 3: (A) Posteroanterior view of the patients skull (B) Grummons analysis for facial asymmetry

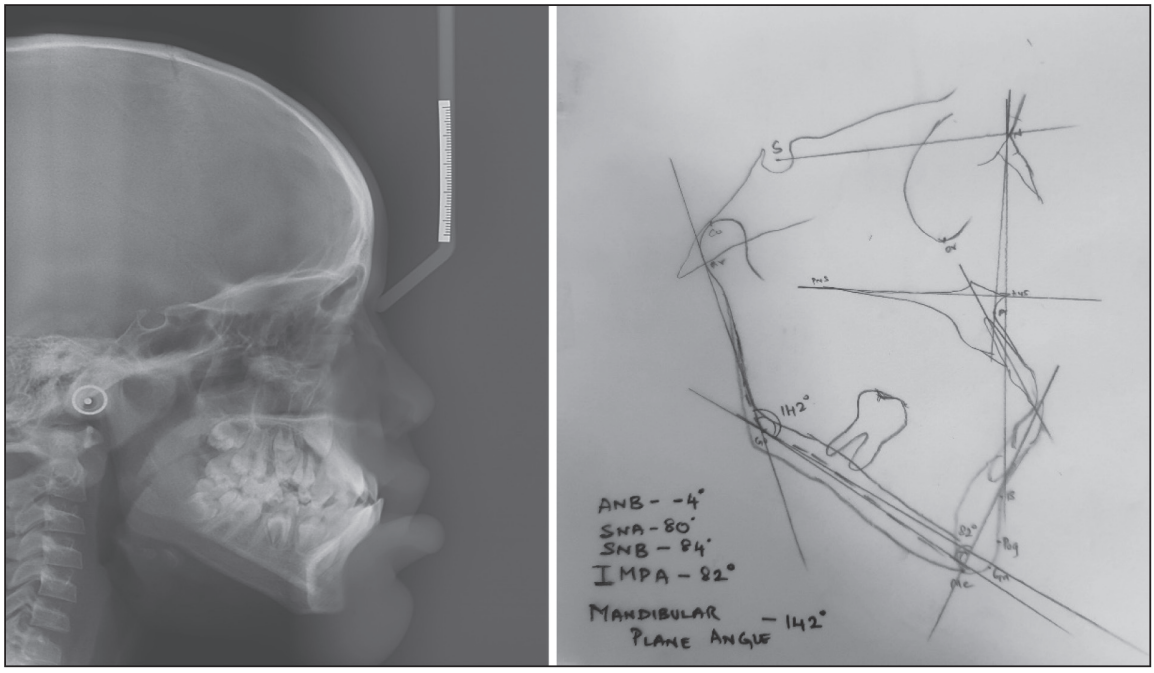

Figure 4: (A) Lateral cephalogram. (B) Linear and angular analysis on the cephalogram

Treatment involving cosmetic surgeries should be carried out after cessation of the growth; magnetic resonance imaging should be recommended to rule out tumors.[7,5]

Declaration of patient consent

A detailed informed consent has been obtained from the patient regarding the publication of his photos and radiographs obtained for various treatment purpose.

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The patient understands that his name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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