

## Three-dimensional Imaging of Congenital Hemifacial Hypertrophy: A Case Report

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### ABSTRACT

*Hemihypertrophy or hemihyperplasia is a condition in which there is asymmetrical overgrowth of the cranium, face, trunk, and limbs on one side of the body. It is characterized by hyperplasia of tissues rather than a hypertrophy. It is more commonly seen in females than in males and usually has the right side of the face involved. It may be an isolated finding or may be associated with a variety of syndromes. We report here a case of isolated non-syndromic right facial hemihypertrophy associated with diffuse warty pigmentation on the right side of the face, which is rarely reported. The clinical features, various radiological and imaging findings, and differential diagnoses to be considered are discussed in this case report.*

**Keywords:** Hemifacial, pigmentation, 3D computed tomography, unilateral.

### INTRODUCTION

Hemihypertrophy or hemihyperplasia is defined as asymmetry between the right and left sides of the body due to the overgrowth of one or more body parts (1). It may be an isolated finding or may present with certain syndromes (2). It can be classified as complex hemihypertrophy, where one half of the body is involved; simple hemihypertrophy where a single limb is involved; and hemifacial hypertrophy where one side of face is involved (3). The condition can occasionally be crossed, involving different areas on both sides of the body. The prevalence for hemihyperplasia was reported approximately as one in 86 000 births (4), although the exact prevalence of non-syndromic facial hemihypertrophy could not be obtained from the available literature. A case of non-syndromic facial hemihyperplasia has been discussed here.

### CASE REPORT

A 16-year-old male patient presented to the Department of Oral Medicine and Radiology with a complaint of swelling in the right side of the face since birth (Fig. 1). The swelling had increased in size since 2–3 years. The

patient also complained of hearing impairment on the right side and watering of eye on the right side during reading. There was no history of pain in that area. The past medical and family history was non-contributory. The patient was well oriented to time and place and was moderately built and nourished. The vital signs were all under normal limits. Extraoral examination revealed a facial enlargement on the right side of the face. The enlargement measured roughly 12 cm in size and caused decreased eye opening on the right side. There was diffuse warty and papular pigmentation noted on the same side extending onto the neck but not crossing the midline (Fig. 1).

There was deviation of chin towards the left side, frontal bossing, malar prominence, and hyperplasia of lips on the right side when compared to the left side. The nasal bridge was flattened with deviation of the nasal tip. Intraoral examination revealed diffuse enlargement of buccal mucosa and macroglossia on the right side (Fig. 2). On palpation, the palatal bone and mandible appeared to be enlarged on the right side when compared to the left side. Gingival hyperplasia was seen near the right posterior teeth. The patient had high arched palate

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Fig. 1: Facial asymmetry due to the enlargement on the right side of the face.

and cross bite on the left side (Fig. 2). The teeth appeared to be larger on the right side when compared to the left side (Fig. 2).

On palpation, the swelling was smooth, non-tender and soft in consistency. The enlargement was confined only to the facial region. No other body parts were involved. Based on the clinical features, a provisional diagnosis of progressive right facial hemihypertrophy was made. The differential diagnoses considered were fibrous dysplasia and neurofibroma. The haematological investigations undertaken revealed that all values were within the normal range. On radiographic investigations, orthopantomograph revealed increased width and height of the mandible on the right side. The maxillary and mandibular teeth on the right side appeared to be more bulbous along with altered trabeculae in the bone. The condyle appeared to be enlarged on the same side (Fig. 3).

A computed tomography (CT) with 3D reconstruction was made which revealed enlarged maxillary sinus, condyle, mastoid air cells, and subtle soft-tissue thickening seen on the right side (Fig. 4).



Fig. 2: Intraoral picture showing unilateral macroglossia, enlarged right side of the jaws and cross bite.



Fig. 3: Orthopantomograph showing enlarged right side of the jaws.



Fig. 4: Plain axial computed tomography scan of head with bone window showing enlarged maxillary sinus, condyle, mastoid air cells, and subtle soft-tissue thickening seen on the right side.

3D reconstructed CT scan revealed prominent supraorbital ridge and infraorbital bone, enlarged ramus of mandible on the right side and deviation of mandible towards the left enlarged ramus of mandible and condyle at the base (Fig. 5).

Audiologist consultation was obtained and hearing deficit was elicited on the right side. An ophthalmological examination was also conducted, which revealed diminished eyesight on the right side when compared to the left side. The patient was also referred to a skin specialist for the pigmentation who suggested laser treatment. A detailed full body work-up was done in our case to rule out any other abnormalities. The treatment suggested to the patient was reconstructive orthognathic surgery followed by correction of malocclusion and then laser surgery for the pigmentation on the face. As the

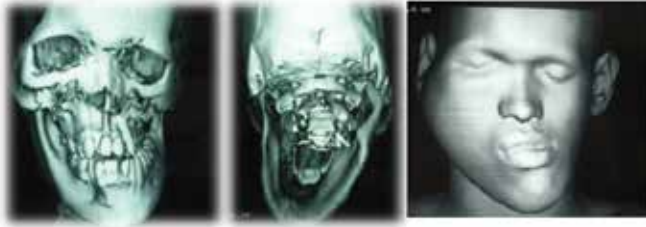


Fig. 5: 3D reconstructed computed tomography scan showing prominent supraorbital ridge and infraorbital bone, enlarged ramus and condyle of mandible on the right-side and deviation of mandible towards the left.

patient was only 16 years of age, the treatment is postponed till the physiological growth of the patient ceases.

## DISCUSSION

Hemifacial hyperplasia (HH) presents a developmental anomaly characterized by unilateral enlargement of the face involving single or multiple tissues including the underlining bone on the affected side (5). Congenital HH was first described by Meckel in 1882 (6). Ward and Lerner (1947) stated that ‘asymmetric enlargement could be manifested in a unilateral/crossed configuration and may involve all the body tissue in the area (*ie*, total) or a single systems such as muscular, vascular, skeletal or nervous (*ie*, limited)’ (6). Congenital HH has been classified by Rowe in 1962 as involvement of (7):

- the entire half of the body: complex hemihypertrophy,
- one or both limbs: simple hemihypertrophy,
- the face, head and associated structures: hemifacial hypertrophy.

The case reported here falls under the third sub-heading. The aetiology of this condition is unknown. The aetiology of HH remains unknown and many theories have been proposed for that, including abnormalities in vascular or lymphatic flow, lesions of nervous system, hormonal imbalance, chromosomal abnormalities, neural tube defects, and altered intrauterine environment or the deviation from normal twinning process (8). The unilateral enlargement of the face is bounded superiorly by the frontal bone not including the eye, inferiorly to the border of mandible, medially by the midline of face and laterally by ear, pinna being included within the hypertrophic area (7). The disproportionate growth is almost always evident at birth and the enlarged side generally grows at the rate proportional to but slightly faster than the normal side. The disproportionate growth rate is maintained until the time of skeletal maturation and results in an asymmetry existing throughout life (6). The condition is usually accentuated with age,

especially around puberty. The similar history was given by the patient, as was seen in the case presented here. Clinically, the condition represents as unilateral overgrowth of the orofacial soft tissues, bones and teeth, as seen in our case. The right side of the face is affected most often than the left side as seen in our case, and HH is more common in women than in men with the ratio of 3:2 (9). Some reports in the literature mention the condition is more prevalent in males (7). Whites are more commonly affected than blacks (10). One case report has mentioned the presence of epidermal nevus on the face (11). The case reported here also had diffuse warty papular pigmentation like nevus on the face which makes it unique from the cases reported earlier.

The dentition abnormalities are with respect to crown size, root size and shape and rate of development (6). The skeletal findings are in the form of an asymmetric growth of the frontal bone, maxilla, palate, mandible, or condyles (6), as seen in our case. Abnormal occlusal relationships such as midline deviation, unequal occlusal plane level, open bite, and widely spaced teeth on the involved side have been reported (6). Our case had cross bite on the unaffected side. The soft-tissue abnormalities include enlarged anatomical tissues on the involved side. The tongue shows unilateral macroglossia as seen in our case. The other soft tissues such as lips, buccal mucosa, uvula, and tonsils are also affected. In our case, the lips and buccal mucosa were involved. The imaging features noted for hemifacial hypertrophy are an increase in the size of condyle, ramus and mandibular canal, along with hyperplasia of the maxillary and mandibular bone (7), as seen in our patient. The other uncommon features include early eruption and idiopathic root resorption, which were not present in our case. Isolated HH should be a diagnosis of exclusion.

The differential diagnosis that can be considered are Proteus syndrome, neurofibromatosis type 1 and vascular malformations such as Klippel–Trenaunay syndrome and lymphaticovenous malformations, fibrous dysplasia, dyschondroplasia, congenital lymphedema, arteriovenous aneurysm, haemangioma, lymphangioma, and malignant conditions such as osteosarcoma and chondrosarcoma (7). A full neurological examination and brain imaging are prudent to rule out these entities. A detailed full body work-up was done in our case to rule out the above-mentioned entities. Treatment is usually not indicated for isolated cases of facial hemihyperplasia unless cosmetic considerations are involved. The treatment is usually planned when physiological growth ceases, which includes reconstructive procedures such

as orthognathic surgical procedure combined with orthodontic therapy and soft-tissue debulking by excision of excess masticatory and subcutaneous tissues, with preservation of neuromuscular functions (7). Congenital hemifacial hypertrophy has usually good prognosis.

## REFERENCES

1. Ballock RT, Wiesner GL, Myers MT, Thompson GH. Current concepts review—hemihypertrophy. Concepts and controversies J Bone Joint Surg Am 1997; **79**: 1731–8.
2. Mutafoğlu K, Cecen E, Cakmakci H. Isolated hemihyperplasia in an infant: an overlooked sign for wilms tumor development. Iran J Pediatr 2010; **20**: 113–7.
3. Rowe NH. Hemifacial hypertrophy: review of the literature and addition of four cases. Oral Surg 1962; **15**: 572–6.
4. Ringrose RE, Jabbour JT, Keele DK. Hemihypertrophy. Pediatrics 1965; **36**: 434–48.
5. Sugiyama M, Tanaka E, Ogawa I, Ishibashi R, Naito K, Ishikawa T. Magnetic resonance imaging in hemifacial hyperplasia. Dentomaxillofac Radiol 2001; **30**: 235–8.
6. Nayak R, Baliga MS. Crossed hemifacial hyperplasia: a diagnostic dilemma. J Indian Pediatr 2007; **25**: 39–42.
7. Deshingkar SA, Barpande SR, Bhavthankar JD. Congenital hemifacial hyperplasia. Contemp Clin Dent 2011; **2**: 261–4.
8. Yoshimoto H, Yano H, Kobayashi K, Hirano A, Motomura K, Ohtsuru A et al. Increased proliferative activity of osteoblasts in congenital hemifacial hypertrophy. Plast Reconstr Surg 1998; **102**: 1605–10.
9. Rudolph CE, Norvold RW. Congenital partial hemihypertrophy involving marked malocclusion. Dent Res 1944; **23**: 133.
10. Panat SR, Chakarvarty A, Chinnavar SN, Aggarwal A, Jha PC. Hemifacial hyperplasia: a case report. Oman Med J 2013; **28**: e059.
11. Saritha M, Chandrashekar L, Thappa DM, Ramesh A, Basu D. Late onset epidermal nevus with hypertrichosis and facial hemihypertrophy. Indian J Dermatol 2014; **59**: 210.

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