

Congenital Hemifacial Hyperplasia: Report of a Rare Case with Review of Literature

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Abstract

Hemifacial hypertrophy is a developmental disorder which is characterized by facial asymmetry unilaterally. It is a congenital malformation usually seen at birth and progressively increases with age. Although etiology is unknown, various theories such as chromosomal abnormalities, heredity, atypical twinning, endocrine dysfunction, and altered intrauterine environment have also been proposed. A few cases have been reported in the literature. We herein report a case of true hemifacial hyperplasia in 65 years old male.

Key words: Congenital hemifacial hyperplasia, Facial asymmetry, Hemifacial hypertrophy

INTRODUCTION

A minor asymmetry is acceptable characteristic of morphogenesis. However, asymmetry which is easily noticeable can affect esthetic, normal functioning and quality of life. A congenital developmental disturbances causing unilateral overdevelopment of tissues of face both hard and soft is seen in this rare disorder called hemifacial hypertrophy. In 1982, Hemihyperplasia was first described by Meckel.¹ Rowe in 1962 classified it as complex involving entire half of the body, simple involving one or both limbs, and hemifacial involving half of the face. The term hyperplasia is preferably used since an increase in number of cells is seen rather than increase in size of cells.² The aim of this report is to present a case of congenital hemifacial hyperplasia and to update the existing clinical knowledge.

CASE REPORT

A 65-year-old male complained of inability to open his mouth since many years. He gave a history of enlarged right

side of face since birth which has gradually increased to the existing size. His parents, all his siblings, and children did not suffer from his disorder.

On extra oral examination, a gross facial asymmetry was seen on the right side of the face. The soft tissue on maxillary and mandibular region along with ear pinna and ala of nose was enlarged as compared to the left side of the face. Chin was deviated toward the left side of the face. The mass was soft and nontender on palpation (Figures 1 and 2).

On intraoral examination, enlargement of the maxillary and mandibular alveolar ridge, tongue and gingiva were seen on the affected side. Reduced mouth opening with intermaxillary distance of 1.5 cm was present. The absence of teeth was noticed on the affected side which had exfoliated 2 years back (Figure 3).

Orthopantomogram, lateral view of the skull and posterior anterior view of skull revealed the absence of teeth on the right side and enlarged body of the mandible unilaterally. There was an increased height of the ramus and body of the mandible on the right side in comparison to left side. An oversized right condyle and deviation of chin toward the unaffected side could be appreciated (Figures 4 and 5). Based on these clinical and radiologic examinations, a diagnosis of congenital hemifacial hyperplasia was concluded.

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Figure 1: Profile of the patient showing gross asymmetry on right side of the face



Figure 2: Comparative profile of right and left side of the patient showing obvious asymmetry on right side



Figure 3: Intraoral examination showing absence of teeth and enlarged tongue on right side

DISCUSSION

Hemifacial hyperplasia is a rare congenital anomaly of hard and soft tissue where there is an enlargement of one half of the face. It is seen in 1 in 86000 live births and was first reported by Friedreich in 1863. Usually, it presents itself at birth with a male predominance. The right side of the face is more commonly affected.² This case was a male with right side of the face affected. The enlargement was present since birth and had then stabilized after 18 years of age.



Figure 4: Orthopantomogram absence of teeth on the right side and enlarged body of the mandible oversized right condyle unilaterally. Deviation of chin toward the unaffected side could be appreciated

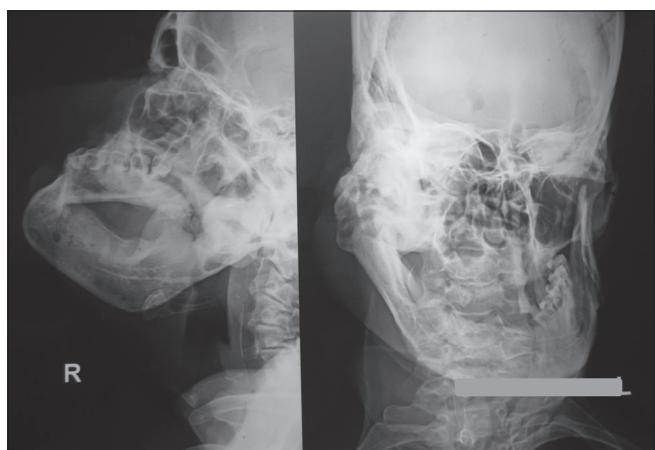


Figure 5: Lateral view of the skull and posterior anterior view of skull showed increased height of the ramus and body of the mandible on the right side in comparison to left side

Although etiology is unknown, few theories which have been proposed are hereditary, atypical twinning, anomalies in chromosome, altered environment in intrauterine life, endocrine dysfunctions, central nervous system disturbances and malformations of vascular or lymphatic systems. This etiological heterogeneity is a result of multisystem involvement.³

The integumentary, cardiovascular, neurological, genitourinary, musculoskeletal, respiratory, and endocrine systems are affected by this disorder. There is asymmetric growth and development of facial bones and dentition. Asymmetry is not a feature of deciduous dentition but early eruption of permanent dentitions seen.⁴ In our case, we could not record the size of the dentition on the affected side as there was an early shedding of the permanent teeth.

Hemiatrophy or Parry Romberg syndrome which manifests as unilateral underdevelopment along with weakness of muscles and deficit of neurological entities can pose as a differential diagnosis. However, it manifests later in life between 5 to 15 years of age. Bony tumors, fibro osseous lesions do not

involve the soft tissues and dentition. Cutaneous lesions and vascular malformations are usually bilateral. All these lesions with an exception of hemiatrophy can be diagnosed histopathologically. Proteus syndrome, hyperpituitarism, epidermal nevus syndrome, Maffucci syndrome, Ollier's syndrome, Klippel-Trenaunay-Weber syndrome, Langer-Giedion syndrome, Russell Silver syndrome, McCune-Albright syndrome, and multiple exostosis syndrome can mimic hemifacial hypertrophy. Unilateral allocation of dental abnormalities and coinciding unilateral tongue enlargement are the striking features of hemifacial hypertrophy.^{2,4}

CONCLUSION

Hemifacial hypertrophy creates a diagnostic dilemma and needs a thorough clinical and radiological evaluation. It

requires a multidisciplinary approach of management because of its multisystem involvement. Unless there is a necessity for cosmetic correction, treatment is not indicated.

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