Available online at http://journalijcmes.com

INTERNATIONAL JOURNAL OF CURRENT MEDICAL SCIENCES

CASE REPORT

ISSN: 2320- 8147

International Journal of Current Medical Sciences- Vol. 6, Issue, 8, pp. 142-145, August, 2016

HEMIFACIAL HYPERPLASIA –A RARE CASE REPORT

Raveendran Jayabalan., Adaikkappan M., Sivakkolunthu M.K., Sethurajan S and Rajadirajan, S

Department of Radiodiagnosis, Rajah Muthiah Medical College and Hospital, Annamalai University, Annamalai Nagar – 608 002

A R T I C L E I N F O

Article History:

Received 25th, May, 2016,
Received in revised form 6th,
June, 2016, Accepted 26th, July, 2016,
Published online 28th, August, 2016

Key words:

Facial asymmetry, Partial and Complete Hemi facial hyperplasia, macrodactyly, congenital anomaly.

A B S T R A C T

Asymmetric variations of contralateral structures of head and neck occur unusually in general population and are an accepted feature of morphogenesis. Gross asymmetry characterized by marked unilateral overdevelopment of hard and soft tissues of the face is a rare congenital malformation and has been termed as Hemifacial Hyperplasia (HFH). The etiology is unknown but several theories has been proposed including hormonal imbalances, diseases involving nerves, vascular conditions, lymphatic abnormalities and mechanical influences. Generally treatment is not indicated for Hemifacial hyperplasia unless cosmetic considerations are involved.

A 17 year old male referred for investigation to our department of radiology with the characteristic features of Hemifacial Hyperplasia (HFH). The case is presented to supplement existing clinical knowledge.

INTRODUCTION

Hemi facial hyperplasia (HFH) is a condition in which half of the face- maxilla alone, maxilla with the mandible, or half of the faces in concert with other parts of the body- grows to unusual proportions. Occasionally, a gross asymmetry easily perceptible to the eye may occur which progresses slowly but steadily and exhibits asymmetric development. One such entity characterized by marked unilateral overdevelopment of hard and soft tissues of the face is a rare congenital malformation. This has been termed variously in the literature as facial hemihypertrophy, hemimacrosomia, partial/unilateral gigantism, congenital hemifacial hyperplasia. Pollock AR (1985) emphasized the use of the term congenital hemifacial hyperplasia.

Hemihypertrophy was first documented by meckel in 1822 and was described as essentially a developmental anomaly. Asymmetric enlargement could be manifested in a unilateral / crossed configuration and may involve all the body tissues in the area or a single tissue.

HFH is classified as true and partial hemihypertrophy. True hemihypertrophy involves not only the soft tissues ofthe 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.

The purpose of report is to present the case history of an 17 years old male with hemifacial hyperplasia to supplement existing clinical knowledge.

Case Report

A 17-year-old male presented to Rajah Muthiah Dental College and Hospital, with facial asymmetry present since birth [Fig.1].

Figure 1 Photograph of patient of 17 yrs old demonstrating left facial enlargement

Corresponding author:* **Raveendran Jayabalan
Department of Radiodiagnosis, Rajah Muthiah Medical College and Hospital, Annamalai University, Annamalai Nagar – 608 002

He was referred to our department of Radiodiagnosis for relevant imaging. He was the last of two siblings without any previous family history of facial asymmetry.

Patient presented with diffuse left facial asymmetry. The left facial swelling is increasing in size with age causing gross facial asymmetry. The physical examination revealed a mentally healthy and active boy. The patient's face was asymmetrical with an enlargement of the left side, including the malar, maxillary, and mandibular region. The pinna of the left ear was slightly enlarged and there was deviation of the nose and the chin to the right side, which appeared to be the normal side [Fig1]. The skin and hairs appeared normal with the exception of small hyperpigmented nonhairy patches on the left cheek. The soft tissues of the left cheek and the lip were thick and fleshy. There was an obvious enlargement of tongue on the left side, which began abruptly at the midline [Fig2]. Speech was not impaired by this deformity of the tongue and the sense of taste appeared to be equally distributed. The general examination revealed normal symmetrical development of the upper extremities and the trunk. Both lower extremities show macrodactyly with flat foot [Fig3].

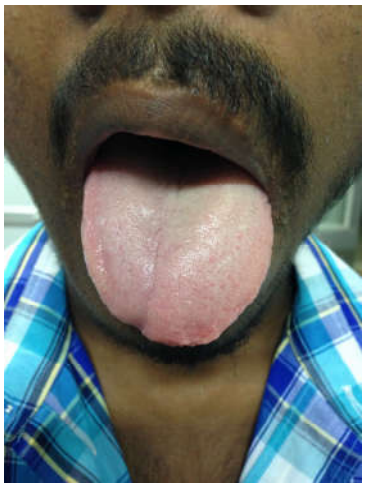


Fig 2 Enlargement of tongue on left side

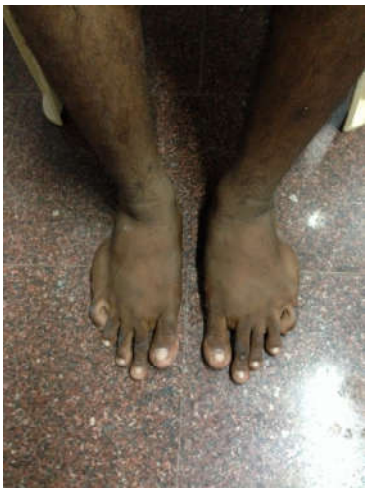


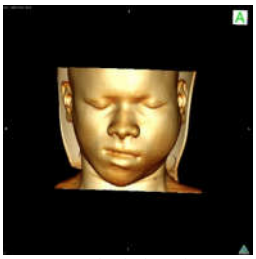
Fig3 Bilateral flat foot with Macrodactyly

Patient had early exfoliation of deciduous teeth in the left upper back tooth region and the erupting permanent teeth are very large in size causing disturbed occlusion. Considering history and all the clinical features the provisional diagnosis of congenital HFH was made and thedifferential 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.[6] Macrodontia noted in the left side teeth.There was accelerated root formation of all the left upper teeth. Slanted palatal drag suggestive of hyperplastic maxilla and hyperplastic zygomatic bone was observed on left side. 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.

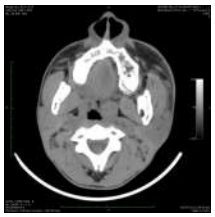
Imaging Findings

Patient was subjected to radiograph, USG,CT and MRI imaging modalities. PNS radiograph showed enlarged maxillary sinus of Left side. X-ray foot [Fig] showed bilateral Macrodactyly and flat foot. Magnetic resonance imaging (MRI) report was showing enlargement of the subcutaneous adipose tissue, masseter muscle and jaws on the left side. Facial asymmetry is mainly due to soft tissue and bony enlargement.

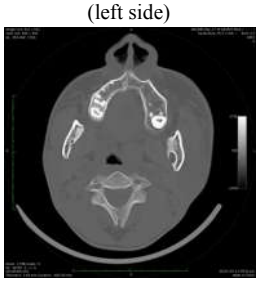
MDCT images and findings

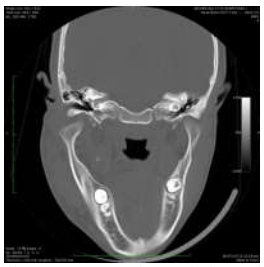


3D – Shaded surface display(SSD)technique - shows Hemifacial Hyperplasia/hypertrophy (left side)

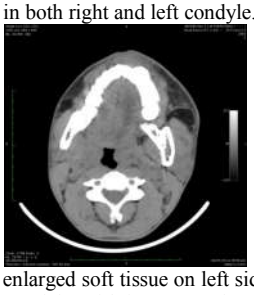


Hypertrophy with fatty infiltration noted in left side masseter and pterygoids muscles

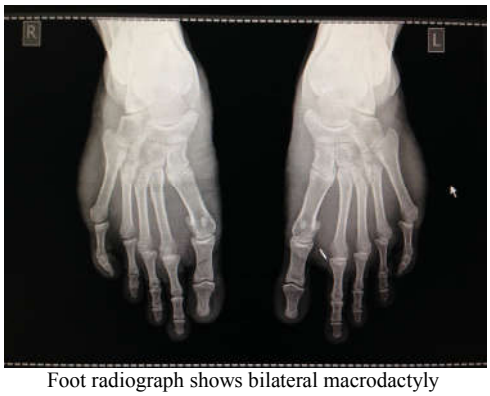




Left side Hemimandibular, Maxillary, Zygomatic and frontal bones all show bone enlargement/expansion. CT scan shows great discrepancy in both right and left condyle.



CT scan showing enlarged soft tissue on left side. Subcutaneous plane diffuse increased fat with intramuscular fatty replacement in left side of face



Foot radiograph shows bilateral macrodactyly

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

DISCUSSION

HFH is an unusual condition that produces the facial asymmetry by a marked unilateral localized overgrowth of all the tissues in the affected area, that is, facial soft tissues, bone, and teeth.[1,6] The disorder occurs more commonly in females with ratio 3:2 and the right side ofthe face is usually affected.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 the hypertrophic area.[6] 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.[1]

The multisystem involvement has resulted in etiological heterogeneity and no single theory explains the etiology adequately. Factors that can be implicated are heredity, chromosomal abnormalities, atypical forms of twinning, altered intrauterine environment, endocrine dysfunctions, anatomical and functional anomalies of vascular/lymphatic systems and disturbances of the central nervous system.[1,7] An interesting concept put forth by Gesell (1927) suggests an inequality of regulatory abilities in embryologic development leading to an aberrant twinning mechanism.[5,7]

Pollock and coworkers (1985) proffer a hypothesis that the neural tube and its precursors are unilaterally hyperplastic.[4] The enlarged half of the neural tube would give rise to proportionally more numerous neural crest cells on the involved side. The increased number of crest cells persists throughout much of the prenatal and formative postnatal growth periods of life and lead to unilateral overgrowth of the crest-derived bone, cartilage, teeth, muscles, and soft tissue. Some tube segments may be involved more often than others helping explain why cephalic areas are more often affected clinically than other areas.[4,8]

Involvement of oro-facial structures is related to asymmetric morphogenesis of teeth, bone, and soft tissues. Dentition abnormalities are with respect to crown size, root size, and shape and rate of development.[1,3] Tooth size enlargement is random with the frequency of involvement more in cuspid followed by premolars and first molars and least occurring in incisors, second molars, and third molars.[2,9] Usually enlargement does not exceed 50% of the normal size.[9] Root size and shape may be proportionally enlarged. Precocious eruption of permanent teeth by up to 4–5 years is usually seen.[1] Skeletal findings may be in the form of an asymmetric growth of the frontal bone, maxilla, palate, mandible, or condyles. Abnormal occlusal relationships such as midline deviation, unequal occlusal plane level, open bite, and widely spaced teeth on the involved side have been reported.[1,9] Soft tissue abnormalities include thickened and enlarged anatomical tissues on involved side with the sharply demarcated midline. The tongue, which is commonly involved, may show a bizarre picture of enlargement of fungiform papillae with unilateral enlargement and contra lateral displacement.[2,3,9] Other soft tissues such as lips, buccal mucosa, uvula, and tonsils are also affected.

Because most of the striking signs of congenital hemihypertrophy are usually manifested in the orofacial region, Gorlin and Meskin (1962) suggested them help in differentiation of this condition from other entities that may simulate hypertrophy[9]. Among the conditions that closely mimic HFH as suggested by various authors are fibrous dysplasia, dyschondroplasia, congenital lymphedema, arteriovenous aneurysm, hemangioma, lymphangioma, Klippel-Trenaunay syndrome, von Recklinghausen's neurofibromatosis, malignant conditions such as osteosarocma and chondrosarcoma.[2,7,9,10] All these conditions, however, exhibit sufficient clinical differences with HFH and should be distinguished on the basis of specific radiographs, clinical and laboratory findings.[3] To the unfamiliar clinician, hemi hypertrophy that is localized to oro-facial region can constitute a diagnostic problem. Deformities of the teeth and their related hard tissues in the jaw are key findings for correct diagnosis, particularly in hemihypertrophy limited to the face.[3,9,10].

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.[11] This disorder has been associated with adrenal cortical carcinoma, nephroblastoma (Wilms' tumor) and hepatoblastoma.[12] Occasionally genitourinary system disorders like hypospadias, cryptorchidism and medullary sponge kidney are also observed.[13] 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.

Management

Generally, treatment is not indicated for HFH unless cosmetic considerations are involved. Procedures usually are planned (multidisciplinary approach) when physiological growth ceases. These may include

reconstructive procedures such as osteotomies or orthognathic surgical procedure and soft tissue debulking by excision of excess masticatory and subcutaneous tissues, with preservation of neuromuscular functions.[2,3,7] HFH is generally associated with good prognosis with no reports of malignant degeneration.[14] 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.

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 patient to a single specialist.

References

1. Nayak R, Baliga MS. Crossed hemifacial hyperplasia: A diagnostic dilemma. J *Indian SocPedodPrev Dent.* 2007;25:39–42.[PubMed]
2. Islam MN, Bhattacharya I, Ojha J, Bober K, Cohen DM, Green JG. Comparison between true and partial hemifacialhypertrophy.OralSurg Oral Med Oral Pathol Oral RadiolEndod. 2007;104:501–9. [PubMed]
3. Khanna JN, Andrade NN. Hemifacial hypertrophy: Report of two cases. *Int J Oral Maxillofac Surg.* 1989;18:294–7. [PubMed]
4. Pollock RA, Newman MH, Burdi AR, Condit DP. Congenital hemifacial hyperplasia: An embryonic hypothesis and case report. *Cleft palate J.* 1985;22:173–84. [PubMed]
5. Gessel A. Hemihypertrophy and twinning. *Am J Med Sci*.1927;173:542.
6. Rowe NH. Hemifacial hypertrophy: Review of literature and addition of four cases. Oral Surg Oral Med Oral Pathol.1962;15:572–87. [PubMed]
7. Horswell BB, Holmes AD, Barnett JS, Hookey SR. Primary hemihypertrophy of the face: Review and report of two cases. *J Oral Maxillofac Surg.* 1987;45:217–22. [PubMed]
8. Tommasi AF, Jitomirski F. Crossed congenital hemifacial hyperplasia. Oral Surg Oral Med Oral Pathol. 1989;67:190–2.[PubMed]
9. Lawoyin JO, Daramola JO, Lawoyin DO. Congenital hemifacial hypertrophy: Report of two cases. Oral Surg Oral Med Oral Pathol.1989;68:27–30. [PubMed]
10. Ward J, Lerner HH. A review of the subject of congenitalhemihypertrophy and a complete case report. *J Pediatr* 1947;31(2):403-14.
11. Lee S, Sze R, Murakami C, Gruss J, Cunningham M. Hemifacial myohyperplasia: Description of a new syndrome. *Am J Med Genet* 2001;103(2):326-33.
12. Batsakis JG. Tumors of the head and neck: Clinical and pathological considerations (2nd ed). Baltimore: William and Wilkins 1982;301-02.
13. Eisenberg RL, Pfister RC. Medullary sponge kidney associated with congenital hemihypertrophy (asymmetry). A case report and survey of the literature. *Am J Roentgenol Radium Ther Nucl Med* 1972;116(2):773-77.
14. Hayashi S, Tomioka T, Aoki H, Nakakuki K, Mekaru K. Hemifacial hypertrophy.Report of two cases. Oral Surg Oral Med Oral Pathol 1973;35(2):750-61.

****