HEMIFACIAL HYPERTROPHY

ABSTRACT

Hemifacial hyperplasia is a rare developmental anomaly characterized by marked asymmetry of unilateral facial tissues. It involves orofacial soft tissues, bones of the face, and teeth. The cause remains ambiguous although several predisposing factors have been reported. A case report of a 11-year-old male patient with unilateral hemifacial enlargement is presented to highlight the clinical findings and to discuss the differential diagnosis and its management.

INTRODUCTION

Hemifacial hyperplasia (HFH) isa rare congenital developmental disorder characterized by unilateral overgrowth of facial structures ,involving bones,softtissues,cartilage and nerves.It is appropriate to refer it as hyperplasia rather than hypertrophy because it is characterized by tissue hyperplasia rather than hypertrophy. It may affect the face, or it may affect the whole side of the body.Meckel first described HFH in 1822, and Wagner first documented it in 1839.[1]HFH affects 1 in 86,000 live births.[2]
This syndrome causes facial asymmetry, which becomes more noticeable with age and fully develops during puberty.[5,6]Early diagnosis is essential as overgrowth typically accelerates during puberty and can results in functional and aesthetic complications like macrodontia,severe malocclusion,asymmetric growth of facial structures.

CASE REPORT

A 11year-old male came to the Department with the chief complaint ofasymmetry of the face. The parent stated that there were no prenatal or significant postnatal history and he was born by full term normal delivery. The parents noticed that his right face was gradually getting larger and width of his eyelids were different. The enlargement extended inferiorly to the lower border of the mandible and from the midline to the preauricular area. The affected right half of the skin appeared normal and had not changed in thickness. There was no family history of similar condition.

EXTRAORAL FEATURES

- Marked facial asymmetry, right side larger.
- Diffuse soft tissue swelling extending from right infraorbital region to the mandible
- Bony enlargement-Maxilla, Mandible, Zygoma
- Deviation of the nose
- Drooping of the right corner of the mouth
- Masseter muscle enlargement
- Clicking of TMJ present
- Deviation in mouth opening present

- Midline shift present
- Obliteration of nasolabial fold present
- Chin tilted

RADIOGRAPHIC FEATURES

- Enlarged mandibular canal
- Deviated Nasal Septum
- Soft tissue shadows overlap structures on right side
- Gonial angle steeper on right side
- Zygomatic process more prominent on right side
- Teeth width on enlarged side is wider
- Right Ramus width increased
- Tooth overlapping on right side in panoramic view
- Condylar size different on both sides
- Right palate enlarged

INTRAORAL FEATURES

- Occlusal slant present
- Macrodontia on affected side
- Complete right side buccal crossbite present
- Dentinalcaries-55,65
- Pre-shedding mobility 53,63 present

DIAGNOSIS

For an accurate diagnosis, abnormalities of the teeth , hard tissues and soft tissues in the jaw are crucial. Etiological factors of facial asymmetry include vascular malformations, fibro-osseous lesions such Paget's disease, fibrous dysplasia, dyschondroplasias, and malignant diseases (osteosarcomas, chondrosarcomas). The presence of foraminal enlargement in hemifacial hyperplasia and distinctive clinical-radiological features in the remaining entities allow for their differentiation.[27]In condylar hyperplasia growth,it is limited to condyle.Vascular or lymphatic malformations have fluid filled spaces in imaging with pulsations and skin dis-colourations present.

DENTAL MANAGEMENT

A multi-disciplinary approach involving, Pedodontists,Oral and maxillofacial surgeons, Orthodontists and Endodontists are needed for the management.Surgical treatment involves both hard and soft tissue surgeries; the hard tissues can be treated by a combination of condylar recontouring, osteotomies to achieve the necessary shape, followed by debulking of soft tissues. The treatment modalities range from subtle soft tissue contouring to extensive surgeries to correct the underlying bony defect and reshape the overlying soft tissues.Liposuction and other soft tissue contouring procedures have been popular for many years.[9]

Since early diagnosis and management prevent further worsening of facial asymmetry in this case extraction of 53, 63 done followed by lateral expansion using Jack screw was given for the correction of crossbite.This approach is typically used in the non-surgical phase of treatment to manage transverse maxillary deficiencies and in unilateral crossbites.

DISCUSSION

Rowe classified hemihyperplasia anatomically into three categories: HFH, which affects one side of the face, complex hemihyperplasia, which affects half of the body, and simple hemihyperplasia, which affects a single limb. Rowe further classified HFH into two categories: (1) true hemifacial hyperplasia (TFHF), which is characterized by a unilateral enlargement of the viscerocranium that extends superiorly from the frontal bone (excluding the eye) to the inferior border of the mandible and from the midline to the pinna of the ear with enlargement of all soft tissues, teeth, and bone in the area; and (2) partial hemifacial hypertrophy (PHFH) if the enlargement is restricted to a single structure.[7]
Three aspects of the dentition of affected side were noted by Rowe: the size of the crown, the form and size of the roots, and the rate of development. He pointed out that not every tooth was equally affected.[8] The most often impacted teeth were the cuspids in the permanent dentition and the second molar in the deciduous dentition, followed by the first molars and premolars. But compared to their counterpart, the enlargement was not more than 50%. It was also interesting to observe that while the second molars, which grow around the same time, are not as large as the premolars. On the afflicted side, early deciduous tooth shedding, delayed permanent tooth eruption, prematurely formed teeth with short roots, and congenitally absent teeth were frequently observed.[9]

According to Rowe, the alveolar bone of the affected side is thicker and larger with the highest bulk occurring distal to the largest tooth. Additionally, there was a tendency for an open bite because both posterior ridges produced exostoses that made contact with one another when the jaw closed.[10] The mandibular canal may also enlarge radiographically.[11]It has also been reported that the growth of the tongue is uniform and starts suddenly in the midline with excrescences that resemble large fungiform papillae.[12]

The etiology cannot be fully explained by a single explanation. Inherited chromosomal abnormalities are one of the reasons of this condition.[3,4]Hormonal imbalances, neurological disorders, vascular disorders (like hemangiomas and arteriovenous malformations), anomalies (like lymphangioma, incomplete twinning, abnormal intrauterine environment, somatic mutations, and central nervous system lesions), mechanical influences, and congenital syphilis are just a few of the numerous theories put forth to explain hemifacial hyperplasia.[13,10]

Noe and Berman proposed that damage to the mitochondria in half of the fertilized egg is the primary cause of the excessive number of cells.[14]Pollock et al. proposed an embryological theory that suggests the enlarged half of the neural tube has more neural crest cells.[15] According to Yoshimoto et al., the pathogenesis is believed to be caused by basic fibroblast growth factor and its receptor-stimulated osteoblastic differentiation on the afflicted side relative to the normal side of the face.[16] According to Pollock et al., the overgrowth process is histologically characterized by an increase in cell number rather than size.[17]

Men are more likely than women to be affected by HFH, and right-sided involvement is noticeably more common.[18] The skin on the affected side did not show any abnormalities extraorally, which is consistent with Gorlin,Meskinand Lawoyin*et al* findings.[19,20]

Nasal septum deviation was noted in this case which is in line with what Oktay et al. found.[21] The outcome was hypertrophied nasal conchae on the afflicted side and involvement of the face muscles surrounding the nasolabial fold.[22] The most notable characteristics of HFH in terms of the unilateral distribution of dental anomalies include rate of development, crown size, root size, and shape.[23]The size of the crowns on the right and left teeth differed significantly in the cervicoincisal, mesiodistal, and labiolingual dimensions. This result is in line with the Row study.[24]In addition, the right mandibular molars have larger roots than their contralateral counterparts. The differential diagnosis of the following partial hyperplasia should be taken into consideration because the majority of the characteristics of partial HFH typically appear in the orofacial region: CLOVES Syndrome, Klippel-Trenaunay syndrome, which is linked to capillary-lymphatic-venous deformation, fibrous dysplasia, and other overgrowth syndromes.[25]

IMPLICATIONS IN DENTAL PRACTICES

Asymmetry in primary dentition allows for better growth monitoring in HFH and in early intervention . Radiographic examination of affected side is necessary for assessing bone density and nerve pathways. Tooth on the affected side may erupt prematurely and expect macrodontia on the affected side and soft tissue enlargement can lead to obstructive sleep apnea and can risk airway of the patient so early management is crucial.

FURTHER RESEARCH

The future of managing hemi-fascial hyperplasia is shifting from reactive , extensive surgery towards precision based molecular-informed care.Reseachers are moving from treating the condition solely as physical deformity and are now targeting the genetic that drives the overgrowth.

CONCLUSION

Early diagnosis helps to differentiate from similar conditions and can decrease psychological distress making surgical and orthodontic treatment a primary goal. Regular follow up are essential to monitor skeletal and dental stability.

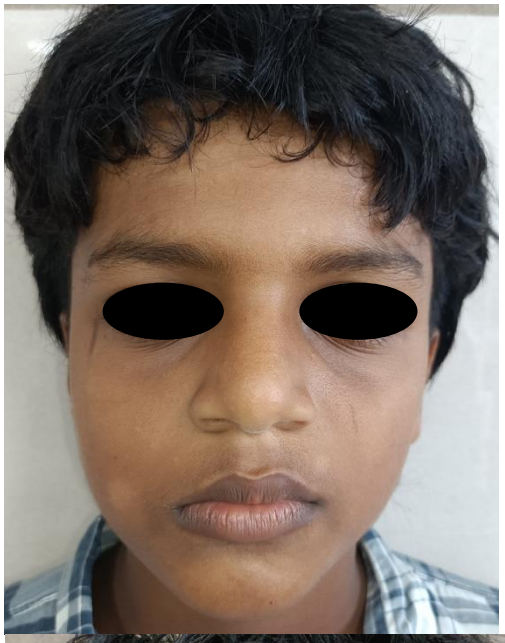

Figure 1:Demonstrated facial asymmetry

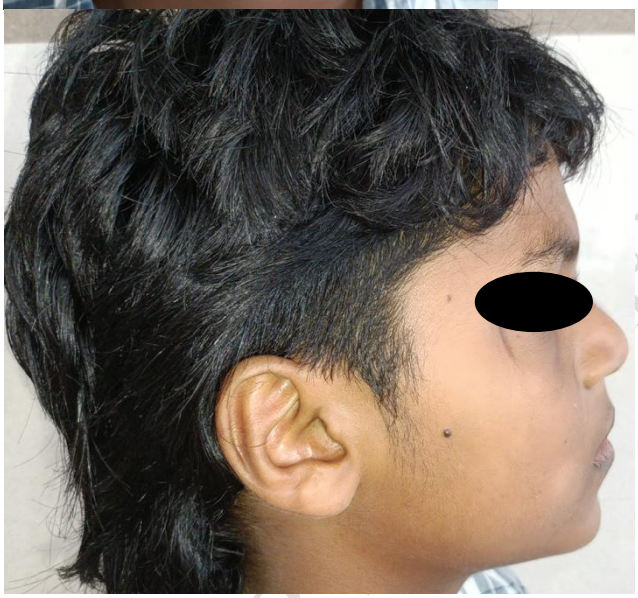

Figure 2:Right side of facial profile

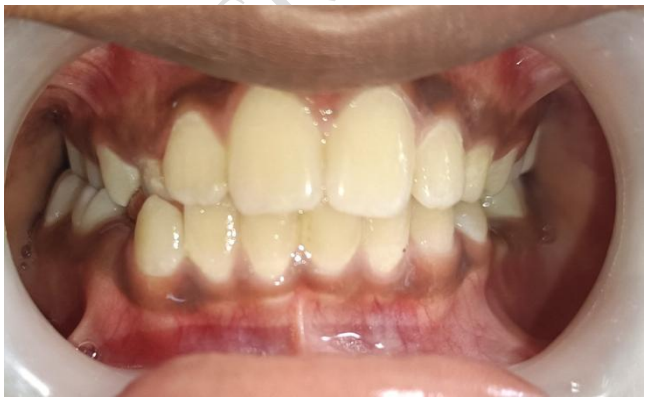

Figure 3:Intraoral view

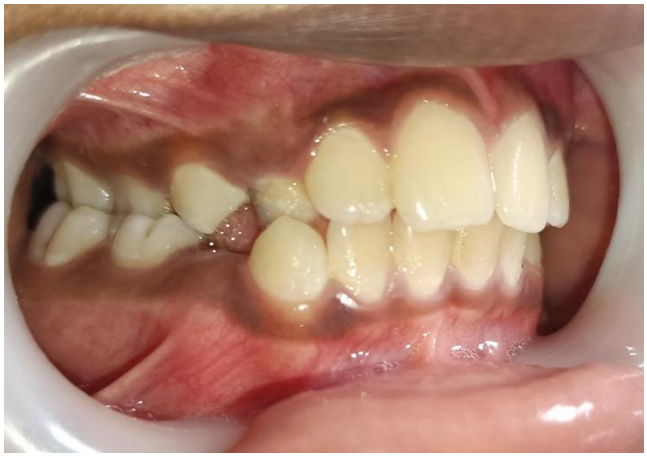

Figure 4:Intraoral view of buccal crossbite

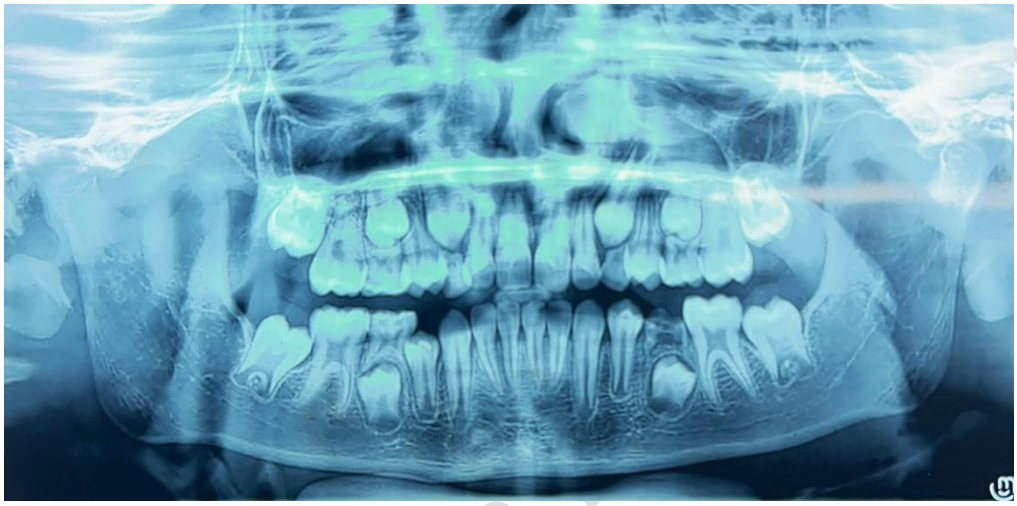

Figure 5:OPG showing enlarged mandible on right side.

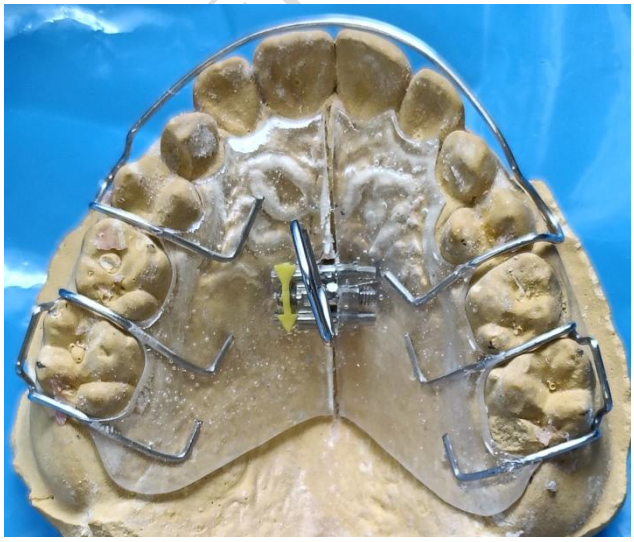

Figure 6 : Lateral expansion using Jack screw

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