Case Report

An unusual presentation of partial hemifacial hyperplasia

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INTRODUCTION

Hemifacial hyperplasia is a rare congenital anomaly characterized by an increase in volume of all affected tissues of half of the face. Depending on extent of involvement it can be complete or partial. It is present at birth, subsequently grows proportionally, and stops growing before adulthood.

Unilateral condylar hyperactivity can manifest as hamimandibular hyperplasia or hemimnadibular elongation.¹ It causes asymmetric growth of the mandible and develops typically in early adulthood. Both disorders have an unknown aetiology.

Here we report a case of partial hemifacial hyperplasia patient, in whom a progressive growth pattern similar to hemimandibular hyperplasia subsequently occurred.

A high condylectomy along with malar arch recontouring was successfully performed to stop the progressive growth and improve facial asymmetry.

CASE REPORT

A 22-year old married female reported to out patients clinic with a complaint of gradually worsening right facial swelling over the past two years. She was aware of a somewhat asymmetric face since her school days. She did not seek treatment as it was not perceived to be severe by her parents. Subsequent body growth was normal, as was her cognitive development. Apart from esthetic concern, there was no functional problem whatsoever.

The patient was the only child of healthy, non-consanguineous parents. Her family history was negative for congenital anomalies or asymmetries. The mother’s pregnancy had been uneventful; birth was via normal vaginal delivery.

On clinical examination, there was obvious marked hypertrophy of right cheek. (Figure 1a)

On palpation, it was bony hard and appeared to follow the course of zygomatic arch continuous anteriorly with the body of zygoma.

ABSTRACT

Partial hemifacial hyperplasia is a rare congenital malformation characterized by prominent unilateral overdevelopment of some of the hard and soft tissues of the face. This uncommon diagnosis should be considered while examining facial asymmetry. We report a case of slowly enlarging right facial swelling which turned out to be enlarged zygomtic arch. Complete evaluation led us to believe continued condylar hyperactivity as the underlying reason for worsening deformity. We offer an explanation for the same. Minimal surgical intervention led to significant aesthetic improvement which was patients main concern.

Keywords: Condyle, Hyperplasia, Hemifacial
Intra oral examination revealed a normal Inter- incisal opening without deviation. Tongue was enlarged on right side with prominent fungiform papillae (Figure 1b). Tooth size was similar on two sides. There was edge to edge occlusion with bite just starting to open on the right side. Occlusal cant was absent.

Right corner of the mouth and mandibular lower border on the right appeared to be lower than the left side.

TM joint and Facial nerve examination did not reveal any abnormality. Right pinna was similar to the left. There was no cervical lymphadenopathy.

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Orthopantomogram (OPG) revealed large condyle with elongated neck on the right side. There was an increase in height of mandibular ramus and body with bowing down of inferior border on the right (Figure 2) which stopped at midline. Dental midline was not shifted. These features resemble that of hemimandibular hyperplasia. Most striking feature was increase in size of zygomatic arch resulting in pronounced outward bowing which was confirmed on 3D CT scan pictures (Figure 3).

Rest of the physical examination didn't reveal any obvious asymmetry. Systemic physiological findings were within normal limits.

As the patient complained of progression of asymmetry slowly over 2 years, bone scan was advised to assess continuation of bone growth.
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Based on clinical, radiological and nuclear medicine examination, diagnosis of case of partial hemifacial hyperplasia with continued condylar hyperactivity was made.

The patient was offered a comprehensive treatment plan consisting of high condylectomy followed by occlusal correction with orthognathic surgery involving presurgical orthodontics and sagittal split osteotomy of ramus of mandible. In addition, lower border osteotomy on right side to improve symmetry was also suggested. However patient’s only concern was cheek swelling with no subjective masticatory difficulty. The patient declined orthognathic treatment. Finally, it was decided to proceed with high condylectomy and zygomatic arch recontouring to prevent worsening of deformity and improvement in existing asymmetry.

Under general anesthesia, right condyle was accessed via a pre-auricular approach with temporal extension. High condylectomy excising 5 mm superior portion of condylar head was done preserving the articular disc. Lower portion of zygomatic arch was excised corresponding to overgrown ragged area and the remaining bone was smoothed (Figure 5a). Hospital stay was uneventful and wound healing was satisfactory.

The patient has been following up with us regularly and evaluation at two years shows considerable improvement in facial asymmetry. (Figure 6) She remained unconcerned with unchanged occlusion of edge to edge cuspal contact of right posterior teeth but was quite satisfied with facial appearance.

Histopathogical exam of excised condyle (Figure 5b) confirmed clinical diagnosis of condylar hyperplasia. (Figure 7a-d).

**DISCUSSION**

Hemifacial hyperplasia was first described in 1822 by Meckel.2 It was reported first by beck in 1836 as such.3 Almost all cases of hemi-hyperplasia occur sporadically. Hoyme et al found a 2:1 right-sided predominance and a 2:1 female predilection in 104 cases.4 It has estimated prevalence of 1 in 86,000 live births.5,6 Such is the rarity of this entity.

An anatomical classification initially described hemihyperplasia (half-body involvement) as simple hemihyperplasia, complex hemihyperplasia, or hemifacial hyperplasia. Hemifacial hyperplasia was further classified as ‘true’ or ‘partial’.

True hemifacial hyperplasia is characterized by unilateral visera-cranial enlargement of all components of the soft and hard tissues (excluding the globe) whereas in partial hemifacial hyperplasia there is less severe involvement of a few structures.
The term hemifacial hyperplasia is to be preferred over hemifacial hypertrophy although these are used interchangeably in literature. There is histological evidence of increase in the number of cells and not size.\(^5,6\)

Hemifacial hyperplasia (HFH) is said to be present at birth and then grows proportionally till end of puberty. The growth then ceases.\(^7\)

Tongue involvement with enlarged fungiform papillae is a feature which is described consistently in reported cases and our case is no exception.\(^8\)

Gingival hypertrophy and lower lip involvement has been reported in a few cases. These were absent in our case.\(^9\)

Meckel reported an increase in tooth size on the affected side.\(^2\)

In our case, tooth size was not measured objectively, as it was not observed clinically.

Functional problems include malocclusion and occlusal cant which was not present in our case. Pre-surgical orthodontics followed by mandibular sagittal split osteotomy was proposed to the patient to achieve correct centric occlusion. However, patient declined the same as existing occlusion posed no subjective masticatory difficulty.

Condylar involvement is rarely found in hemifacial hyperplasia and may be misdiagnosed as such. However when combined with additional features they should be correctly designated as partial hemifacial hyperplasia.\(^10\)

This condition is benign and affected individuals have normal life span.\(^9,11\)

A significant clinical feature observed in our case is progressive deformity of zygomatic arch which prompted patient to seek treatment. This was to a large extent responsible for hypertrophy of tissues of the right cheek. Following is our proposed hypothetical explanation for this deformity.

As mandibular ramus continues to lengthen under the influence of hyperactive condyle, masseter muscle also has to lengthen (hypertrophy) in order to maintain its insertion at the angle of mandible. In this process due to continued contraction over a period of time, it exerts outward force on the zygomatic arch. This resulted in ectopic bone formation and outward bowing of the arch.

Osteotomy followed by medial repositioning and miniplate fixation was an option in this case to address the arch deformity. But that would have resulted in stripping all periosteal attachments from the arch which would then be like free bone graft with potential for resorption. Hence recontouring was preferred.

Unilateral condylar hyperplasia results in facial asymmetry where the mandible is affected with secondary involvement of maxillary complex. Its two main types have been described by Obwegeser and Makek. These are hemimandibular hypertrophy, hemimandibular elongation and hybrid forms. Hemimandibular elongation is characterized by predominantly horizontal asymmetry, and hemimandibular hyperplasia, by mainly vertical distortion with increased volume.\(^1\)

In unilateral condylar hyperplasia, the asymmetry is not present at birth but develops later, typically in early adulthood (mean age 21 years). Progression can be confirmed with bone scintigraphy, and treatment consists of a partial condylectomy to stop the progressive growth.\(^12\)

Therefore, the present patient showed an overlap of clinical features of hemifacial hyperplasia and unilateral condylar hyperplasia.

Even though the present case showed histological features consistent with condylar hyperplasia, it is said in literature that histology does not confirm true hyperplasia in all unilateral condylar hyperplasia patients.\(^13,14,15\)

Etiology of hemifacial hyperplasia is far from clear. Proposed mechanisms include those involving the central nervous system, vascular and lymphatic abnormalities, endocrine dysfunction, altered intrauterine development, and chromosomal abnormalities, an embryological hypothesis due to neural crest abnormalities, first branchial arch overgrowth.\(^6,8,14,15\)

CONCLUSION

Partial facial hyperplasia although uncommon should be considered while examining facial asymmetry. Hemifacial hyperplasia per se does not always require treatment. When the patient does seek treatment, desire for cosmetic improvement overrides function in the majority of the cases. Due to involvement of facial tissues to a variable degree, treatment has to be individualized for each patient. We would like to stress that a stable and improved aesthetic result was achieved in present case with relatively straightforward operation of high condylectomy and zygomatic arch recontouring. However, this needs to be discussed with the patients as multiple procedures may be needed in some to achieve satisfactory result which was achieved in a single procedure here.

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