True Hemifacial Hypertrophy – A Case Report and Review of Literature

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Abstract
Hemifacial hypertrophy is a rare congenital disease involving hypertrophy of both soft and hard tissues of the face, and is usually discovered soon after birth. This pathology may be classified as either true or partial hemifacial hypertrophy. Many conditions resemble hemifacial hypertrophy, potentially complicating diagnosis. We present a case of true hemifacial hypertrophy in a 6-year-old girl with radiological findings, differential diagnosis and a brief review of the literature. Myohyperplasia, reported previously as a separate entity, may actually represent a form of true or partial hemifacial hypertrophy. Management of true hemifacial hypertrophy requires meticulous planning, including multiple treatment modalities.

Introduction
The human body generally shows morphological symmetry, which changes with time and circumstances within a definitive range. However, in hemi-hypertrophy, the affected side grows faster and at a rate proportional to the unaffected side. This means the discrepancy between right and left sides remains constant until the process stops with the cessation of growth.

Hemifacial hypertrophy is an uncommon developmental disorder, initially described by Meckel in 1822 (1). Since then, over 250 cases of body and facial hemifacial hypertrophy have been reported in the literature. Classically, this condition presents as a unilateral, localized overgrowth of facial soft tissues, bones and teeth. Of the various classifications proposed, Rowe’s classification (2) is considered the simplest and most practical. He classified hemifacial hypertrophy into true and partial. True hemifacial hypertrophy must involve the contents between and including the boundaries of the viscerocranium: facial bones; facial midline; inferior borders of the mandible and ear, but not involving the eye. Partial facial hemifacial hypertrophy thus is limited to only one structure or does not involve all facial tissues.

Although, crossing the midline to involve parts of both the sides has been seen in some cases (3-5). It is important to mention that all these structures are neither necessarily involved nor enlarged to the same degree, as stated by Stafne and Lovestedt (6) and Hanley et al. (7).

Sometimes this condition may involve just the overlying musculature, in which case the condition is referred to as myohyperplasia (8). This study reviews the literature, discusses the etiology and classification and adds another case of hemifacial hypertrophy to the literature.

Etiopathogenesis
Multiple etiological factors have been implicated in the development of hemifacial hypertrophy, such as hereditary, anatomical, vascular and lymphatic abnormalities, endocrine dysfunction, altered environment, disturbances of the central nervous system and chromosomal abnormalities. However, most of these have not gained wide acceptance due to a lack of precise evidence (9, 10).

As a result, etiological heterogeneity may have been responsible for the varied clinical features, affecting single or multiple systems and the degree of tissue involvement.

Case report
An 8-year-old girl was referred by a local dentist for an opinion regarding enlargement of the left side of the face. The condition was noted at birth and asymptomatic. She was the youngest of 2 normal siblings from normal parents.
without any family history of the condition. The mother did not report any abnormalities during pregnancy and delivered the girl at full term.

Examination revealed unilateral enlargement of the left side of face, extending from the infraorbital margin to the base of the mandible, with a right-side shift of the chin. The left side of the lower lip was enlarged with drooping at the corner of the mouth (Fig.1). Movements of other facial muscles and bilateral vision were all normal. The overlying soft tissues showed significant hypertrophy. No temporomandibular joint dysfunction was detected, and no discrepancies in the range of mandibular motion were noticed (Fig. 2).

Intra-orally, the left side (involved side) of the dentition and tongue were significantly affected. The dentition was abnormal on the involved side (Fig.3) showing precocious development and eruption of the permanent teeth. All left lower permanent teeth except the second premolar and second molar were present in the oral cavity.

The changing dentition on the left side was equivalent to that of an 11- to 12-year-old girl, while the right side was normal for the patient’s age. No midline deviation or canting of the occlusal plane was evident. However, the patient showed a slight anterior open bite. Permanent teeth on the uninvolved (right) side remained unerupted, except the mandibular first molar and central incisor. Exact comparison of the teeth on both sides was thus not possible. However, teeth on the involved (left) side seemed to be larger than normal on measurement. The buccal mucosa was normal in appearance. The tongue showed unilateral enlargement with a sharp midline demarcation. The papillae were enlarged and resembled soft excrescences (Fig.4). Tongue movements and sense of taste appeared normal.

Radiographically, orthopantomography showed increased deposition of bone on the left side of facial structures, particularly the mandible and maxilla. The left side also showed a lower orbital level, with prominence of the zygoma. Significant bone growth was seen between root apices of the left mandibular teeth and the inferior alveolar canal (Fig.5). The left mandibular condyle and coronoid process appeared normal.

Considering the age of the patient and the expected growth and development, keeping the patient under observation was considered the best course, with reconstructive procedures to be planned and performed only after the completion of physiological growth. These surgical procedures would most likely involve osteotomies and/or orthognathic surgery, soft tissue debulking, particularly of masticatory, glandular and subcutaneous tissues, while attempting to preserve neuromuscular functions.
Differential diagnosis

Various entities showing similar manifestations should be excluded. These include: hemiatrophy, showing unilateral underdevelopment with muscle weakness and neurological deficit; fibro-osseous lesions and other bony tumors, which do not involve the soft tissues and dentition; and vascular and lymphatic malformations, cutaneous lesions, and neurofibromatosis, which are usually bilateral, involve different body parts and are diagnosed histologically. Other syndromes such as Beckwith-Wiedemann syndrome, Russell-Silver syndrome, Klippel-Trenaunay syndrome all have distinguishing features other than body asymmetry (has been deleted). Segmental odontomaxillary dysplasia shows unilateral enlargement of the maxilla and fibrous hyperplasia of the overlying gingival tissues.

All these conditions should be distinguished from hemifacial hypertrophy on the basis of specific clinical and radiographic findings.

Discussion

Hemifacial hypertrophy is a rare congenital disorder characterized by hyperplasia of tissues of one or more parts of the facial tissues, rather than hypertrophy. Some authors have reported heredity as an etiological factor in hemifacial hypertrophy (11), but no such contribution was established in the present case. Indeed, of the various theories suggested to explain the etiology of this condition, none was found to be relevant (9, 10).

Our patient was a girl with left-side involvement, concurring with the findings of Ringrose et al. (5). Whereas some studies state the opposite, males are more commonly affected than females and the right side is more commonly involved than the left (2, 12). Rowe (2) reports an equal distribution of the disease between sexes.

Clinical features of the patient were typical, as reported by various authors, particularly enlargement of the teeth, jaws and overlying soft tissues on the left side of the face. Other findings like an enlarged, thick tongue along with enlarged lips were also present on the involved side. However, Lawoyin et al. (13) have reported that neither of her cases of congenital hemifacial hypertrophy showed tongue enlargement. Larger roots, root resorption, open bite, trismus with or without uni-or bilateral ankylosis have been observed infrequently (9). Buccal mucosa was found to be normal. However, Miles (14) reported velvety, soft pendulous folds in the buccal mucosa. Extraorally, the skin did not show any kind of abnormality on the involved side, concurring with the findings of Gorlin and Meskin (1) and Lawoyin et al. (13). Conversely, Hayashi S et al. (15) reported skin abnormalities in a large number of reports (65.8% of Japanese reports of hemihypertrophy).

Ringrose et al. (5) analyzed 129 cases, finding the following as the most common features: mental deficiency; skin abnormalities; varicose veins; and compensatory scoliosis. Other clinical findings reported in the literature are thickened hair and skin, ipsilateral nevi and telangiectasis (9), macrodactyly, polydactyly, syndactyly, and club foot (12).

Pfister et al. (16) showed that abdominal tumors are usually present in hemifacial hypertrophy of the body.
However, in isolated hemifacial hypertrophy, not even a single case has been disclosed with abdominal tumors (17). As our patient was of poor socioeconomic status and neither she nor her parents ever complained of any abdominal problem, we did not perform ultrasonography, although we explained to her parents that she might require this examination for future management. Hemifacial myohyperplasia (8) has been added as a new clinical finding in this disease, where hyperplasia is only limited to the facial musculature. Generally, no treatment is required hemifacial hypertrophy unless cosmetic problems are involved. Long-term management of this patient should begin with psychological guidance and emotional support to adapt the patient to being confident in her environment. After growth ceases, corrective surgery consisting of soft-tissue debulking, facelift, and orthognathic surgery should be planned and performed. Every attempt should be made to preserve neuromuscular functions.

Radiography may not reveal the amount of soft tissue involvement, and should be carefully assessed. Nowadays, this can be easily done with the help of 2- and 3-dimensional computed tomography. Hall (18) has suggested the use of a device known as prosopometer, which precisely indicates the extent of deformity and necessary surgical corrections required in all the planes.

No formal report of malignancy has been published in the literature. Except for a few cases of fatty deposits recurring in the cheeks, no bony regrowth has been reported postoperatively (12).

Conclusion
A wide variety of conditions can resemble hemifacial hypertrophy. Importantly, cases where asymmetry between sides is limited are more difficult to classify as physiological or pathological. Clinical and radiographic information must always be kept in mind to achieve an accurate diagnosis. Finally, treatments involving reconstructive procedures should only be performed after the cessation of physiological growth.

References
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