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Case Report

CROSSED HEMIFACIAL HYPERPLASIA: REVIEW OF THE LITERATURE AND REPORT OF A CASE

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ABSTRACT:

Crossed hemifacial hyperplasia (CHH) is rare developmental condition that manifested as facial asymmetry along with co-existent enlargement of the opposite extremities. We report a case of CHH in a sixteen years old Libyan female who exhibited obvious enlargement of the lower half of the left side of the face, cheek fullness, swollen lips, displaced philtrum and fissured tongue. The teeth on left side of the jaw are larger than their counterparts on the right side, with deviation of the midline and unequal occlusal plane levels. There is an early eruption of the third molars on the left side of the mouth and the ipsilateral maxillary and mandibular alveolar ridges are enlarged. Extra-oral findings included cutaneous pigmentation on the left side of the neck, elongated and enlarged middle finger of the right hand and hypertrophied toe of the right foot without any mental or neurological defects

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تضخم شق الوجه العابر: مراجعة الاوراق المنشورة وتقرير عن حالة جديدة

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تضخم شق الوجه العابر (CHH) هو حالة نادرة بها خلل بالنمو, تتجلى في عدم تماثل قياسات الوجه مصحوبة بوجود تضخم بالأطراف في الجهة المعاكسة من الجسم. نحن هنا بصدد التبليغ عن حالة CHH لدى فتاة ليبية ذات ستة عشر سنة تعاني من تضخم واضح في النصف السفلي من الجانب الأيسر من الوجه والخد مع تورم بالشفاه و تشقق باللسان . كما أن الأسنان على الجانب الأيسر من الفك كانت أكبر من نظرائها على الجانب الأيمن، مع انحراف خط الوسط و عدم تكافؤ مستويات الإطباق. لوحظ كذلك بزوغ مبكر للأضراس الثالثة على الجانب الأيسر مع توسع بالتلال السنخية بالفك العلوي و الفك السفلي. اشتملت المظاهر خارج الفم عن وجود تصبغ جلدي على الجانب الأيسر من الرقبة، و تضخم بالإصبع الأوسط لليد اليمنى و كذلك تضخم بالقدم اليمنى و لم يلاحظ على المريضة أية عيوب عقلية أو عصبية.

INTRODUCTION:

Human body may show slight asymmetric variations which affect the head, face or even half of the body; occasionally gross asymmetric body enlargement without any local lesion or condition can be noticed. This has been most often termed in literature as hemihypertrophy [1-3], even though the use of the term hyperplasia is said to be more histologically precise as there is abnormal proliferation in the number of cells [4-8]. Hemifacial hyperplasia (HH) is produced by marked unilateral overgrowth of all tissues including soft tissues, bone and teeth in the orofacial area [2,9]. HH was first described by Merckelin 1822 and a case was presented clinically by Wagner in 1839 [2,10]. The asymmetric enlargement of the body can be limited to a single system such as muscular, vascular, nervous system or can involve all body tissues[11] whereas in CHH there is facial asymmetry along with co-existent enlargement of the opposite extremities[5]. The etiology of HH remains obscure and many theories have been proposed for that, including abnormalities in vascular or lymphatic flow[5], lesions of nervous system [9], hormonal imbalance [22], chromosomal abnormalities [12], neural tube defects [6] and altered intrauterine environment or the deviation from normal twining process [2]. One interesting theory proposed that fibroblast growth factor and its receptors are selectively involved in the affected fibroblasts leading to hypertrophy [14]; however no single theory was able to explain adequately the exact etiology HH[3]. Many cases of HH have been reported in association with deformities of skeletal system such as macrodactyly, polydactyly, scoliosis, thinning of the pelvis and clubfoot [7]. Central nervous system defects include cerebral enlargement, epilepsy and rarely mental retardation [10]. Furthermore, HH has been reported to associate with Wilm's tumor, adrenal cortical carcinoma and hepatoblastoma[12]. The present article describes a rare case of CHH involving the left side of the face along with changes of the extremities in the right side of the body in association with cutaneous pigmentation, but without mental defects or CNS malformations or internal organs changes. The orofacial and dental findings are described in detail.

CASE REPORT:

Sixteen years old Libyan girl presented to the department of Oral Pathology, Oral Medicine, Diagnosis and Radiology in Dental faculty of Benghazi University as a routine case complaining of toothache associated with decayed tooth. Her mother had noticed abnormal alterations and disproportionate differences in the size of various parts of the body when the girl was just one year old. These differences remained as child grew up. O/E, the lower half of left side of the face was obviously enlarged with slight local hypertrichosis, figure 1.

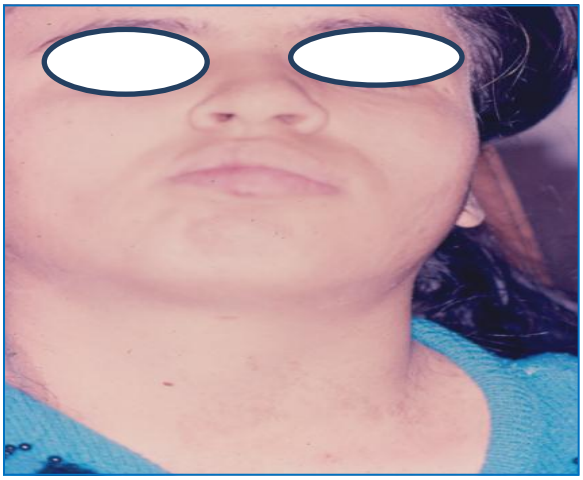

Fig: 1. Clear enlargement of the middle part of the face.

There was fullness of the cheek, enlargement of the lips and displaced philtrum. Cutaneous pigmentation was observed on the left side of the neck, figure 2.

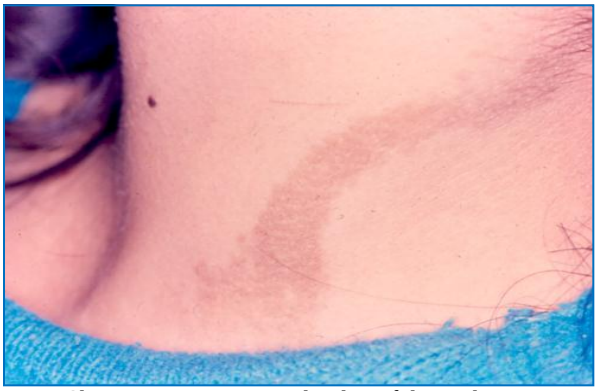

Fig: 2.Skin pigmentation over the skin of the neck

The middle finger of the right hand was abnormally enlarged and longer than the other fingers, figure 3. The right foot appeared hypertrophied with gross deformity of the toes, figure 4.a &b.

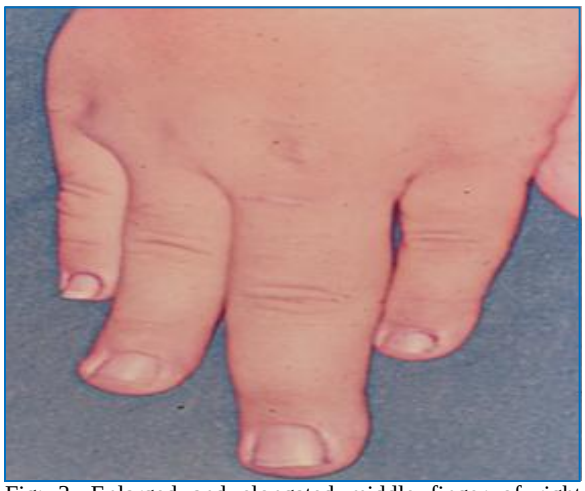

Fig: 3. Enlarged and elongated middle finger of right hands

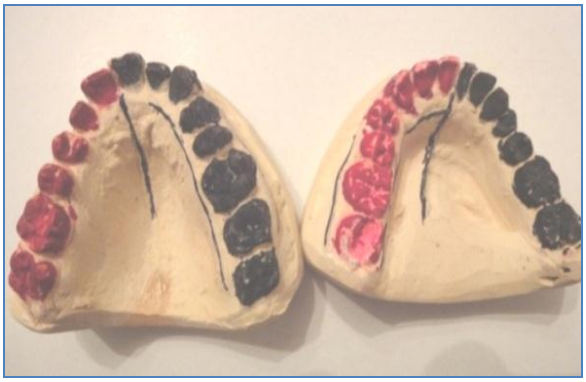

Fig: 6. Upper and lower dental casts with enlarged teeth and alveolar bone and clear deviation of the midline

The middle finger of the right hand was abnormally enlarged and longer than the other fingers, figure 3. The right foot appeared hypertrophied with gross deformity of the toes, figure 4.a &b. Intraoral examination revealed thick and fleshy soft tissue of the left cheek and lips. The tongue is enlarged, fissured and its midline is shifted towards the right side, figure 5. All the teeth (incisors, premolars and molars) on the left side are larger than their counterparts on the right side with an early eruption of third molars at that side. The ipsilateral maxillary and Mandibular alveolar ridges are enlarged. The midline deviation and unequal occlusal planes level on either side of the jaw were clearly visible, figure 6. Further radiological examination revealed accelerated jaw development at the left side with an early eruption of third molars, increased teeth size, and abnormalities of the occlusal plane. On further interviewing of the patient no mental retardation or CNS symptoms were noticed. No abdominal screening was requested by authors and we do not have any further information about any other

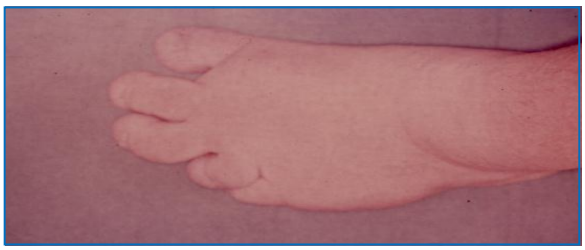

Fig: 4 a. Deformity of the toes of the right foot

Fig: 4 b. Radiograph of the right foot showing the toes deformity

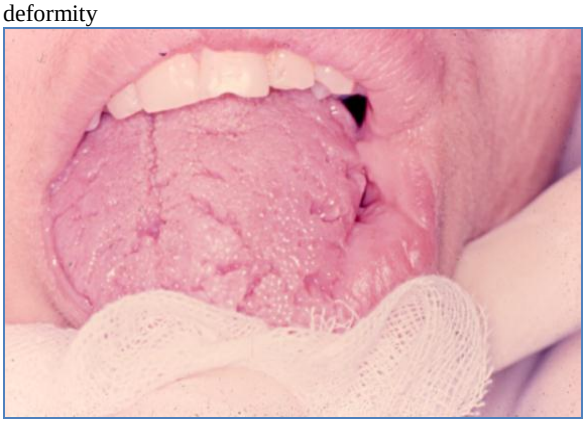

Fig: 5. Grossly enlarged & fissured tongue with well-demarcated midline

internal organs defects. The patient was apparently leading ordinary life with normal intellectual capability.

DISCUSSION:

HH is an unusual condition that produces facial asymmetry due to marked unilateral localized overgrowth of all the tissues in the affected area (facial soft tissues, bone and teeth). These asymmetries are often noticed at birth or shortly after that and accentuated during puberty[7], [13]. The growth is faster but proportionate in the affected side than that in the normal side, and its rate is maintained until the time of skeletal maturation and results in asymmetry which continue to exist throughout life [2]. Classification system for hemihyperplasia based on anatomic site of involvement has been proposed by Rowe [7]. According to this classification, a complex hemihyperplasia means the involvement of half of the body, while in simple hemihyperplasia there is an involvement of one or both limbs

whereas in HH there is an involvement of one side of the face. The asymmetric enlargement of the body could be manifested in a unilateral or crossed configuration. He further classified HH into true and partial varieties. According to this classification the present case is considered as simple true HH associated with crossed enlargement of both limbs. Pollok reviewed 192 cases of HH and found five cases of CHH involved the face and an opposite lower extremity [6]. However, by and large, only few cases of CHH have been reported worldwide so far [5]. Children with hemihyperplasia have an increased risk for developing embryonal tumors, particularly Wilm's tumor and hepato-blastoma [8,12,15], so an early diagnosis of hemihyperplasia coupled with abdominal screening is the best option for the detection of those malignancies at their earliest possible stage [4,16]. Involvement of central nervous system such as mental retardation, strabismus or epilepsy were reported in many previous cases [10,12,13], in our case there were no CNS changes or mental problems. HH appears to affect males more commonly than females [16] nonetheless our case were female patient. The right side of the face is affected than the left side, despite the fact that our case involved the left side of the face and the limbs on the right side of the body[17]. There have been diverse dental abnormalities reported to associate with HH, such as abnormal dental occlusion relations, open bite, spaces between teeth, midline deviation and unequal occlusal plane levels [17-19].Furthermore, asymmetric skeletal and soft tissues changes of excessive growth of the frontal, maxilla, palate, mandible, or condyles have also been reported [20]. Dental changes detected in our case included midline deviation and unequal occlusal plane levels and thickness of the alveolar ridges and moderate maxillary and mandibular enlargement with hypertrophied buccal mucosa, swollen lips on the left side and sharply demarcated midline of the tongue. Cutaneous pigmentation, telangiectasia and hypertrichosis are common findings in many previous studies [21, 22] and the present case showed pigmentation and slight hypertrichosis. Involvement of orofacial structures is related to asymmetric morpho-genesis of soft tissues, teeth and bone [18]. Teeth abnormalities with respect to crown and root sizes shape and root development can be observed. Tooth size enlargement is random with the frequency of involvement more in cusped followed by premolars and first molars and at least occurring in incisors, second molars, and third molars, associated with precocious eruption of permanent teeth [15,18,19]. In the present case there is macrodontia of all the teeth on the affected site. The diagnosis of HH can easily be established when the teeth and related tissues are involved, which are considered as clinical findings for correct diagnosis [24].Several conditions may clinically simulate HH and should be considered in the differential diagnosis such as congenital lymphedema, hemangioma, lymphangioma, Albright syndrome, neuro-fibromatosis, Klipple-Trenaunay syndrome, osteosarcoma and condrosarcoma [25]. These conditions can be distinguished from HH on the basis of their specific clinical, radiological, and laboratory findings. There are many reports of association between emberyonal tumors and hemihypertrophy, so an early and frequent ultrasound screening is recommended, in order to detect these tumors at the earliest possible stage [3,4,6,8]. HH is generally associated with good prognosis and surgical treatment is indicated only for cosmetic consideration, usually planned when physiological growth ceases. Reconstructive procedures such as osteotomies or orthognathic surgery and excision of soft tissues can be considered[24-27].

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