Corrective Surgery of the Hemifacial Hypertrophy in Klipple_Trenaunay Syndrome

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Introduction: Klipple-Trenaunay syndrome (KTS) is characterized by three symptoms including hemangioma, varicose veins and hypertrophy of bony and soft tissues. Hypertrophy usually appears unilaterally, and mostly involves lower limb, however, trunk, upper limb, and face may also be affected. The occurrence of orofacial manifestations in KTS is rare. **Case Report:** In this study a case with hemifacial hypertrophy as a rare manifestation of KTS who underwent surgery based on patient demand of aesthetic has been presented. Further, presentations of KTS in head and neck region and considerations for corrective surgery of these patients have been discussed. **Result:** During the six month post-surgical follow ups, bone and soft tissue were healed with no complications, like severe bleeding or extensive ecchymosis. The amount of asymmetry was improved after six months and it became psychologically acceptable for the patient. **Conclusion:** Surgical intervention of a case of KTS with hemifecial hypertrophy,vascular malformations were not entirely excised and mostly fibrous and adipose in soft tissue were removed. However, the outcome was satisfactory for the patient.

Keywords: Capillary Malformation; Corrective Surgery Haemangioma; Hemifacial Hypertrophy; Klipple-Trenaunay Syndrome,

Introduction

Klipple-Trenaunay syndrome (KTS) was first described in 1900 by two French physicians (1). This congenital syndrome is characterized by the triad of hemangioma, varicose veins and bony and soft tissue hypertrophy (1).

Sixty three percent of patients diagnosed with KTS have all of the three symptoms, however, by presence of two out of three of them, KTS would be diagnosed (2). Port wine appearance (capillary hemangioma) as birth mark is the most common symptom (3) which is diffuse telangiectasias of the superficial skin vessels showing a pink to purplish color. Cavernous hemangioma and lymphangioma can also be found (4). Varicose veins can appear in deep venous system (mostly popliteal vein(3)) as well as superficial veins (5) and even if it could not be detected clinically, it may appear in sonograph (6).

Hypertrophy usually appears unilaterally and its appearance depends on the nature of vascular malformation. Venous malformations may result in varicosities, venous stasis changes, ulceration, phlebitis, thrombosis, and pain (7) while lymphatic malformations can cause soft tissue swelling, cellulitis, and hypertrophy (8). In these patients soft tissue enlargement is evident in all tissue layers. Cutaneous hemangiomas of varying extent and irregular contour appear in the hypertrophic regions (4).

Hypertrophy mostly affects lower limbs however trunk, upper limbs and face may also be affected (4, 9). The effect of gravity on venous drainage from the head and neck region may be the reason for the scarcity of varicose veins in the head and neck region in comparison to lower extremities (10).

Due to variety of KTS presentations, treatment options for KTS vary extensively and each patient should be analyzed individualy (4, 9). In the current study a rare case of KTS with hemifacial enlargement who underwent surgical treatment has been presented.

Case Report

A nineteen- year old man was referred to the department of oral and maxillofacial surgery, Taleghani hospital, Shahid Beheshti University of Medical Sciences. The patient had severe facial asymmetry stillborn. The lesion had not been shrunk after puberty

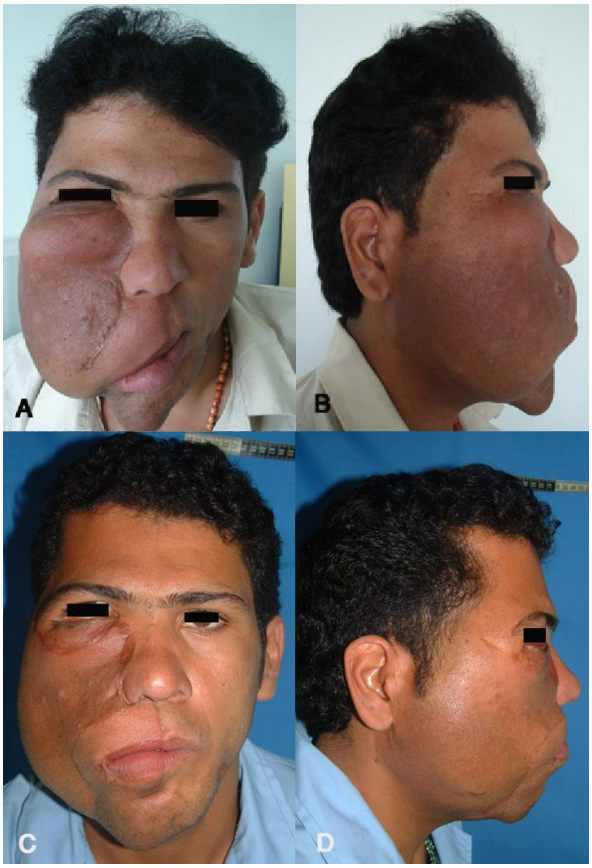

Figure 1. Hemifacial enlargement of the face with oblique lip line. A:Frontal view, B: Profile view. C, D:six months follow up

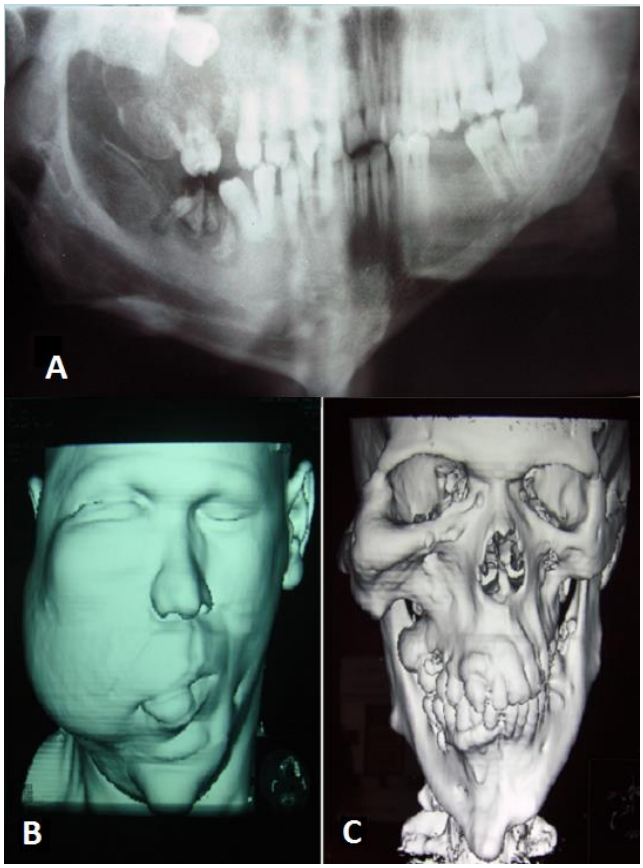

Figure.2. Radiographic evaluation of hemifacial hyperplasia. *A*) Panoramic view. *B,C*) Computed tumographic evaluation of the facial bones

(Figure 1). The patient had no pain and the chief complaint was psychosocial problems due to facial appearance.

A prominent swelling in the face extended from right temporal region to zygoma and lateral aspect of nose was clearly noticeable. The surface of skin of right side was flat and slightly darker than the other side. Vascular lesion sign and symptom was not detected clinically. Presence of the lesion during the growth caused hemifacial hypertrophy, which affected the size and the shape of the facial bone. The intraoral examination showed abnormally enlarged teeth in the affected side, swelling in soft and hard palate, and deviated uvula to the left. Right tuberosity was also involved. The tongue was deviated to left. Facial nerve in the right side of the face had no pronounced activity. Body examination revealed no other similar lesion in upper or lower limbs.

Result

Radiologic examination was performed to assess the extent of the lesions. The results revealed extensive unilateral expansion of both jaws with buccal, lingual and vertical expansion. In the mandible, this was basically in the body region with the rami relatively spared. There was no evidence of encapsulation or cyst formation in the lesions; rather the lesions melded imperceptibly with the surrounding bone. These manifestations were seen unilaterally in mandible, maxilla, and buccal bone complex (Figure 2). The bone consistency was granular and could be described as having a ground/frozen glass appearance with a mottling within. The ground glass appearance had a vaguely "multilocular" appearance. There was marked displacement and delayed eruption of the anterior teeth in the mandible and dental spacing in the maxilla. The mentioned radiographic appearance was seen in ipsilateral zygomaticomaxillary complex.

Primarily, a complete evaluation of coagulation was performed to avoid bleeding complication. Incisional biopsy from three areas of maxilla, mandible, and surrounding soft tissues was done under general anesthesia. Histopathologically the incised specimens were similar to cavernous lymphangioma (Figure. 3).

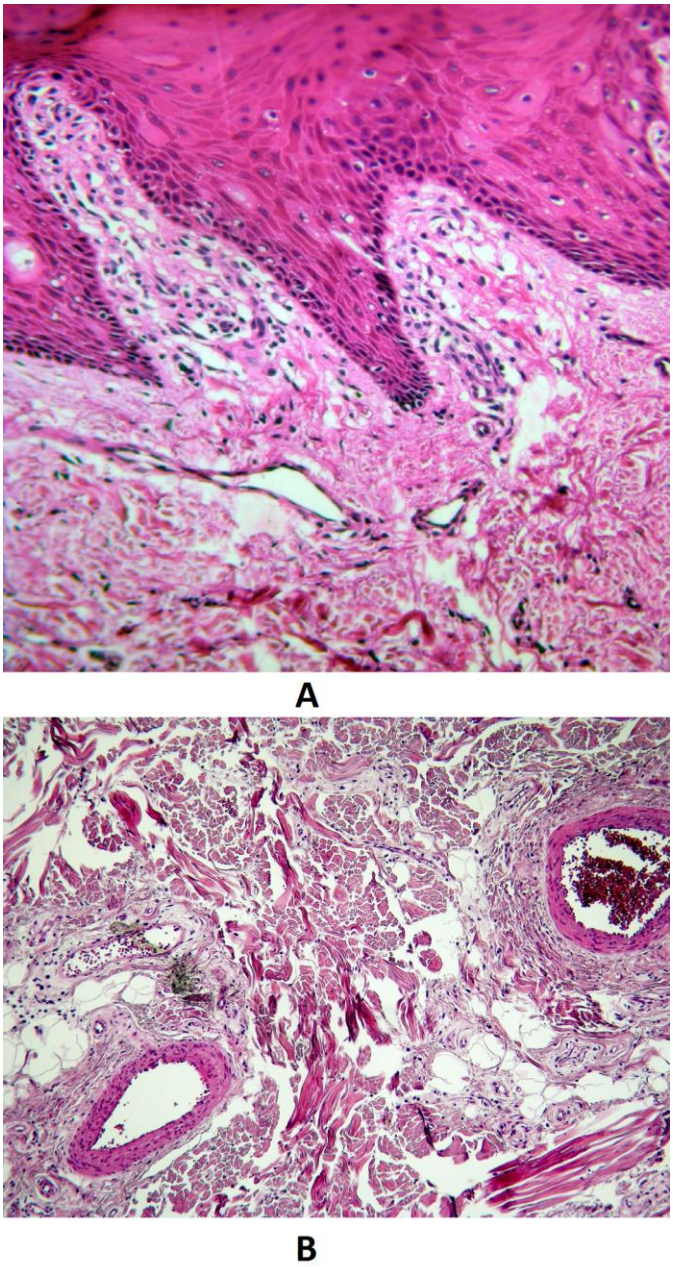

Figure 3. Histopathologic evaluation similar to cavernous lymphangioma

Considering the unilateral clinical appearance, radiographic and histopathologic findings, and the history of congenital disorder, the most probable diagnosis of KTS was determined. In order to improve patient's esthetic and reduce psychological problems, corrective surgery of the lesion was determined. Three surgical procedures were performed to remove the pathologic lesion and correct bone malformations.

During the first surgery, prior to application of general and local anesthesia, subscilliary and lateral nasal skin incision (Weber-Fergusson access), as well as vestibular mucosal incision were used to expose the left side of zygomaticomaxillary complex entirely. The vestibular incision was also made at mandibular symphysis area and extended posteriorly to molar region. After reaching the bone and subperiosteal extension of the flap, left maxillary bone, cheek, and infraorbital rim were exposed. Mandibular bone was also exposed by subperiosteal flap, while spotting the trunk of the mental nerve. In maxillary area the bone was spongy and bloody. Zygomatic and infraorbital bone were shaved by large round surgical bur. Soft tissue, including fibrous and fat tissue, was dissected and debulked with Metz scissor. Samples obtained were sent to laboratories for further histopathological investigations. Samples showed skin tissue with fibrofatty tissue underneath. No evidence of malignancy was found.

Seven month after the debulking of soft and hard tissues, the second stage of surgical procedures; orthognathic surgery, genioplasty, and cheek osteotomy was performed in order to restore the maximal esthetics. Orthognatic surgery was done with the classic maxillary Le Fort 1 access, sagittal split mandible incision (BSSO), and intraoral genioplasty incision (from mental to mental). Approximately 10 mm superior repositioning of the right side of maxilla was done with the removal of bony wedge and it was secured rigidly in place by locking plates and screws. Mobilized mandible was also fixed with bicortical screws. After splinting of ramus and intermaxillary fixation, the unilateral bony wedge in the right chin area was reduced and dislocated to the right. The chin region was secured with titanium plate.

During the six month post-surgical follow ups, bone and soft tissue were healed with no complications, like severe bleeding or extensive ecchymosis. The amount of asymmetry was improved after six months and it became psychologically acceptable for the patient (Figure 1).

Discussion

Diagnosis of KTS is usually made based on presence of any 2 of the following: capillary malformations, varicose veins or venous malformations, soft and/or hard tissue hypertrophy (2). Because of variability of the vascular deformities, a wide range of symptoms may be detected clinically. The presented

case was diagnosed with KTS based on presence of hypertrophic bone and soft tissues and detection of vascular malformations in histopathologic analysis.

KTS enters differential diagnosis with some other rare conditions such as Proteus syndrome, Hemihypertrophy, Sturge-Weber Syndrome, Maffuci syndrome, and Neurofibromatosis (11). The nearest diagnosis in this case was Sturge-Weber Syndrome which was ruled out by absence of intra-cranial calcifications and history of seizers (12).

The occurrence of orofacial manifestations in KTS is very rare. In study of Gloviczki *et al.* (4) only two out of 40 patients with KTS had involvement in head and neck region. Craniofacial findings may include cavernous hemangiomas of facial skin (10, 11, 13, 14), hypertrophy of soft tissue such as enlarged lip, cheek, and asymmetric growth of jaw bones (11, 13). When oral cavity is involved, capillary hemangiomas of gingiva (10, 11, 15), and tongue(15) as well as dental malforamation (13, 14) and gingival hyperplasia (10) may be present. The overall craniofacial and oral involvement can result in asymmetry, malocclusion (10, 14, 16) and anterior open bite (14, 17).

Histopathologic assessment of the lesion in the current case showed that the nature of soft tissue lesions was lymphangioma. Previous authors also reported facial lymphangiomas in patients with KTS (13, 14). When the nature of the underlying lesion is lymphatic, the presentation would be as cellulitis, soft tissue swelling, and hypertrophy (8). The most prominent clinical sign of KTS in the presented case was hemifacial hypertrophy of hard and soft tissue. A review of English literature showed that hemifacial hypertrophy has been reported in a limited number of cases with KTS (10, 13, 14, 18). Bathi *et al* (10) reported 3 patients that had hypertrophy of the jawbone and concomitant malocclusion with no clinically obvious varicose veins. In addition, 2 out of these 3 cases showed exophytic pedunculated growth arising from the gingiva. Our patient is one of the rare cases of KTS that has hemifacial enlargement. Intra oral examination showed enlargement of the right maxillary buccal and palatal gingiva.

In order to achieve better aesthetic for the patient, corrective surgeries were performed. To the extent of our knowledge this is the first report of surgical management of KTS with orofacial manifestations. Previously vascular and orthopedic surgical interventions of lower limb for patients with KTS have been reported (4). Gloviczki *et al.*, (4) reported surgical operations performed in 13 KTS patients including femoral and tibial osteotomy and epiphysiodesis as well as repeated excision of hemangioma and varicectomy.

Clinicians should consider the high risk of bleeding during and prior to the surgery for patients with KTS. These patients may suffer thrombocytopenia (19), platelete dysfunction, and clotting and fibrinolytic abnormalities (16). Gaiser *et al.,* (20) reported that coagulation tests for the blood inside lesions may have different results with the one in peripheral blood in KTS patients. In addition, manipulation of intramedullary hemangioma could result in extensive bleeding as abnormal veins are not able to spasm and start coagulation (16). The presented patient had no coagulative malfunction based on pre-surgical laboratory tests and no excessive bleeding occurred during surgeries. However, complete excision of vascular malformations was not possible as scar tissue formation as well as repeated bleeding and excessive pain could occur and compromise the treatment. This method prevented complete removal of the lesion. Hence, comprehensive reconstruction of facial contours was not achieved. Treatment outcome, however, was acceptable for the patient and improved his social life.

Conclusion

KTS involving the orofacial tissues is a scarce syndrome and no previous report on facial corrective surgery in patients with KTS was found in the literature. Surgical management of these patients may be complicated by extensive bleeding. In the current study we presented surgical intervention of a case of KTS with hemifecial hypertrophy. During surgery vascular malformations were not entirely excised and mostly fibrous and adipose in soft tissue were removed, however, the outcome was satisfactory for the patient.

Conflict of Interest: 'None declared'.

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