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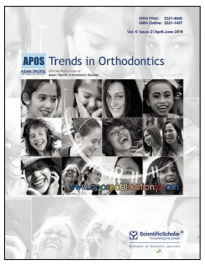

Case Report

Klippel-Trenaunay syndrome: A case report of orthodontic-surgical treatment

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ABSTRACT

Introduction: Klippel-Trenaunay syndrome (KTS) is a congenital disorder characterized by a clinical triad of capillary malformation; varicose veins; and soft tissue and/or bony hypertrophy. This rare syndrome has an estimated incidence of about 2–5/100,000 live births. KTS usually affects the extremities but occasionally can manifest in the craniofacial region, including the oral cavity.

Case Presentation: Our patient, a 17-year-old girl, presented with large areas of port wine stain, unilateral hypertrophy of the facial region, and varicose veins. She was diagnosed with KTS. Orthodontic treatment followed by orthognathic surgery was performed to address her complaint of facial asymmetry and poor chewing efficacy. Treatment resulted in significant improvement in her appearance and dental occlusion and our patient was very satisfied with the results.

Conclusion: This report illustrates an orthodontic and surgical treatment approach of a case with KTS. Effective management of orthodontic and facial manifestations of KTS considerably enhances the patient's function, appearance, and overall quality of life.

Keywords: Klippel-Trenaunay syndrome, Orthodontic management, Orthognathic surgery

INTRODUCTION

Klippel-Trenaunay Syndrome (KTS) is a congenital condition with the main characteristics of capillary malformation, varicose veins, and asymmetric overgrowth. The diagnosis of this syndrome is confirmed by the presence of any two of the mentioned characteristics.[1] The estimated incidence of this rare syndrome is about 2–5/100,000 live births. Both genetic basis and environment have been proposed as etiological factors for KTS; however, the exact etiology remains obscure.[2,3] If KTS is located in the orofacial region, the manifestations may include hemangioma, gingival hyperplasia, premature tooth eruption, soft-tissue hypertrophy, jaw enlargement, facial asymmetry, and malocclusion.[4,5] In this article, we present a case with KTS who received orthodontic treatment followed by orthogathic surgery.

CASE PRESENTATION

A 17-year-old lady presented to the orthodontic department of Shahid Beheshti dental school, with complaints of open bite causing poor chewing efficacy, facial asymmetry, and aesthetic concerns [Figure 1].

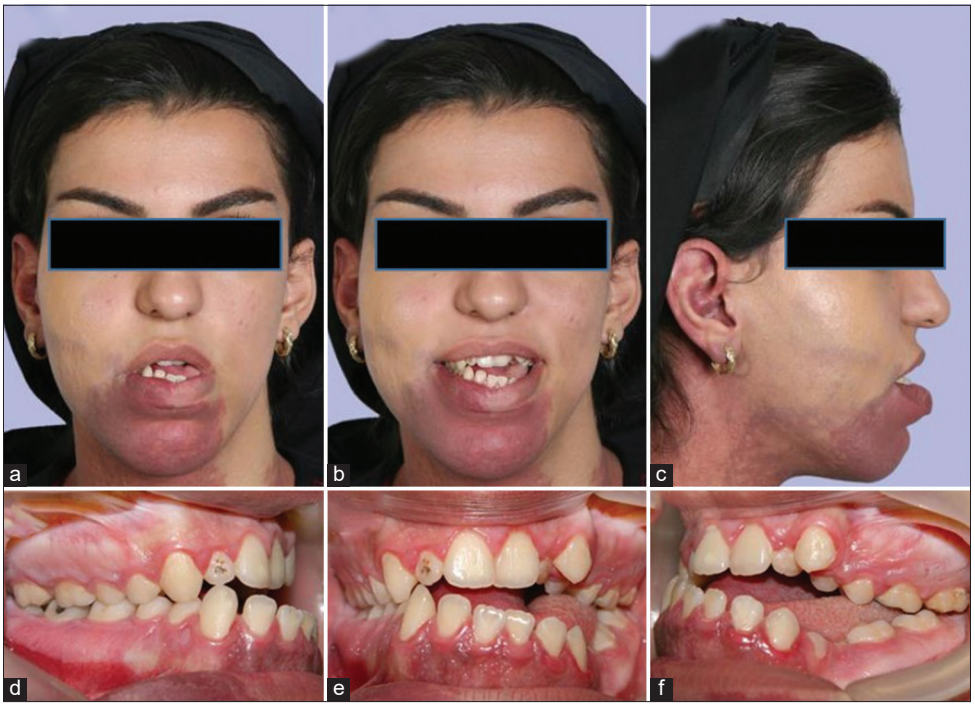

Figure 1: (a) Pretreatment frontal extraoral photograph. (b) Pretreatment smiling extraoral photograph. (c) Pretreatment lateral extraoral photograph. (d) Pretreatment right intraoral photograph. (e) Pretreatment frontal intraoral photograph. (f) Pretreatment left intraoral photograph.

At clinical examination, port-wine stain was apparent on her right ear extending to the lower face, the entire chin area, and the neck. The right side of her face appeared fuller than the left side and the chin was deviated to right. She had a long face, straight profile, pronounced mentolabial sulcus, incompetent lips, and an everted lower lip.

The patient mentioned areas of port-wine stain on her body which was apparent since birth and varicose veins over the femoral region of her right lower limb.

Intraoral findings included asymmetric mild enlargement of the right aspect of the tongue, soft palate hemangioma on the right side, inflamed gingival margins, full permanent dentition, cl III dental relationship, and a severe open bite from the right lateral incisors extending to left molars. Dental midlines were deviated to left [Figure 2].

The posteroanterior cephalogram confirmed the asymmetry, showing hypertrophic right condyle and ramus [Figure 3].

The analysis of lateral cephalogram revealed a vertical growth pattern, slightly retrusive apical bases, class III relationship, maxillary, and mandibular incisors with normal inclination and a protrusive position.

Following the presence of port-wine stains, varicose veins, and asymmetric overgrowth, our patient was diagnosed with KTS.

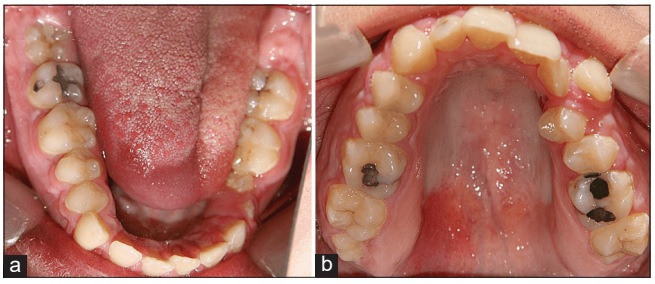

Figure 2: (a) Pretreatment occlusal view of mandibular arch. (b) Pretreatment occlusal view of maxillary arch.

Treatment objectives

The main objectives were to address the patients' chief complaint by correction of the severe open bite and facial asymmetry. Treatment also aimed to resolve crowding and obtain a stable class I dental relationship.

Treatment alternatives

Due to the severity of jaw base discrepancy and open bite, alongside with the absence of growth potential, orthognathic surgery accompanied with fixed orthodontic treatment was inevitable.

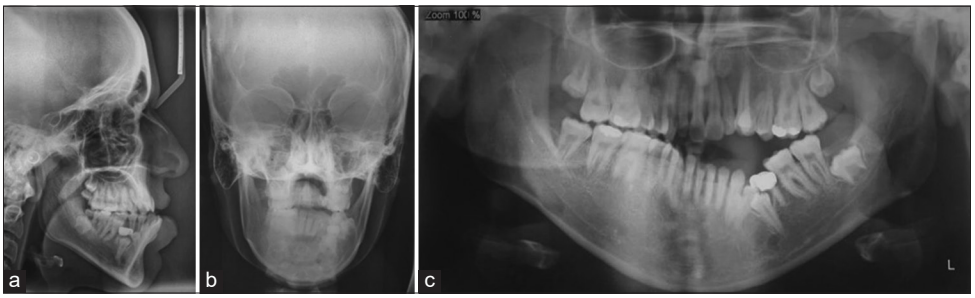

Figure 3: (a) Pretreatment lateral cephalometry. (b) Pretreatment posteroanterior cephalometry. (c) Pretreatment panoramic radiograph.

Treatment progress

After delivering thorough oral hygiene instructions to the patient, a fixed unilateral TPA-tongue guard was cemented on the upper arch to intrude maxillary first molars and prevent tongue thrust. Simultaneously, 0.022 × 0.028–inch fixed appliances with Roth prescription were installed on both arches. Leveling and alignment was initiated by 0.014 inch nickel-titanium wire, and followed by the sequence of 0.016- and 0.018-inch nickel-titanium wires, up to 0.018-inch round stainless steel wires. At this stage, the upper right first premolar was extracted to realign the midline. Archwire sequencing was followed by 0.016 × 0.022 and finally 0.018 × 0.025-inch stainless steel wire.

The aim of the pre-surgical orthodontic phase was to relieve crowding, align the dental midline of each jaw to the skeletal midline, and to achieve an arch coordination which would allow for a stable repeatable occlusion during operation. Evaluation of pre-surgical radiographs and hand articulation of the pre-surgical dental casts approved that the goals were obtained. The mandibular arch was partially leveled at this stage, with the remaining curve of spee on the left side to be corrected post-surgically.

During the pre-surgical orthodontic phase, the patient received multiple sessions of gingivectomy to control gingival hyperplasia, extraction of the wisdom teeth of the right quadrants of both jaws and a rhinoplasty surgery. She was then referred for orthognathic surgery [Figure 4].

Before surgery, complete blood count and coagulogram tests were performed and showed normal results. To rule out the presence of intraosseous hemangioma, a cone-beam computed tomography was obtained from the head and neck region, which revealed no lesion in the area. Arteriography for our case detected low-flow capillary lesions, excluding the presence of arteriovenous malformation.

To correct the unilateral open bite and class III jaw base relationship in this patient, a combination of maxillary impaction, mandibular setback, and rotation was planned. Following a Lefort I osteotomy, 3 mm maxillary posterior impaction of the left side was performed. In the mandible,

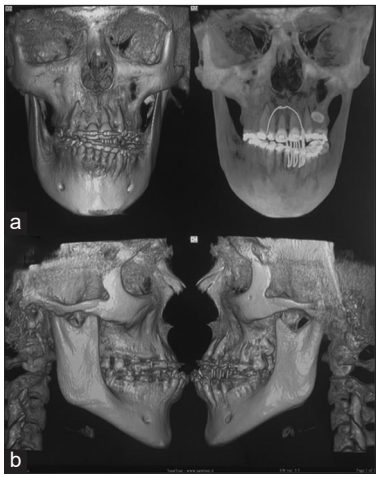

Figure 4: (a) Presurgical coronal view of cone beam computed tomography (CBCT) image. (b) Presurgical lateral view of CBCT image.

asymmetric setback of 2 mm on the right side and 5 mm on the left was carried out after bilateral sagittal split osteotomy. The surgical procedure was uneventful and no unusual bleeding was noted.

The remaining post-operative posterior open bite was closed using posterior box elastics and an 0.018-inch round stainless steel arch wire in place of the mandibular surgical arch wire. Post-surgical orthodontic phase was completed in 6 months.

After debonding, a Hawley appliance with a lateral tongue guard on the left side was fabricated to prevent open bite relapse, and a fixed retainer was bonded in the lower arch and the patient was instructed to wear the retainer full-time for 4 months except for eating and oral hygiene care.

Although significant improvement of facial esthetics was achieved at this time, the patient was concerned with the remaining asymmetry of her lips and increased facial height. Therefore, lip resection and genioplasty surgery was planned. Throughout this second surgery, 8 mm impaction and 2 mm advancement of the chin was performed and soft tissue was resected from the right side of the lower lip.

Treatment results

Our patient was very satisfied with the outcome of the treatment. Her facial asymmetry was corrected and occlusion improved substantially with open bite closure [Figures 5-7]. Canine and molar positions ended in cl I relationship except for the right molars which ended in a stable full cusp cl II occlusion. Cephalometrically, lower anterior facial height was decreased and the jaw bases were brought into cl I relationship. Subsequent follow-up appointments were scheduled every 6 months. Stable results confirmed the patient's compliance in wearing the removable retainer. On the fourth follow-up session, our patient was referred to cosmetic and restorative dentistry service and received direct composite veneers on anterior teeth [Figure 8]. The lower fixed retainer was removed before the procedure, and a new fixed retainer was bonded afterward, the Hawley appliance was adjusted and a mandibular Essix was also delivered to the patient. A timeline of the patient's care is shown in [Figure 9].

DISCUSSION

KTS was first reported in 1900 by two French physicians Klippel and Paul Trenaunay.[6] KTS is characterized by the triad of vascular nevus, congenital varicosities, and overgrowth of the same body part. The diagnosis of this syndrome is confirmed by the presence of any two of the mentioned features.[7] Imaging is advised to assess the underlying soft tissue and bone hypertrophy, venous/lymphatic abnormalities, and disease progression. To evaluate varicosities, venous malformation, or the presence of thrombus development, a Color Doppler ultrasound may be the first step.[8] Although the definition of KTS seems clear, identification can be difficult due to the extreme variability in occurrence of the symptoms.[9] The rarity of this condition also leads to scarce information and limited awareness about it, which may explain why our case was not diagnosed with KTS until referral at the age of 17.

KTS is still frequently misdiagnosed as Parkes-Weber syndrome, which is characterized by the same trio of abnormalities in addition to high-flow arteriovenous fistula. Both syndromes can be identified by clinical examination, but additional tests can help to confirm the diagnosis. Differentiating between these syndromes can benefit from the use of radiological investigations, such as ultrasound, computerized tomography scan, magnetic resonance imaging, and vascular examinations (arteriography and venography).[10] In our case, conventional arteriography revealed low-flow capillary malformation, and the presence of arteriovenous fistula was ruled out, confirming the clinical diagnosis of KTS.

Furthermore, KTS should be differentiated from other overgrowth syndromes that also present with

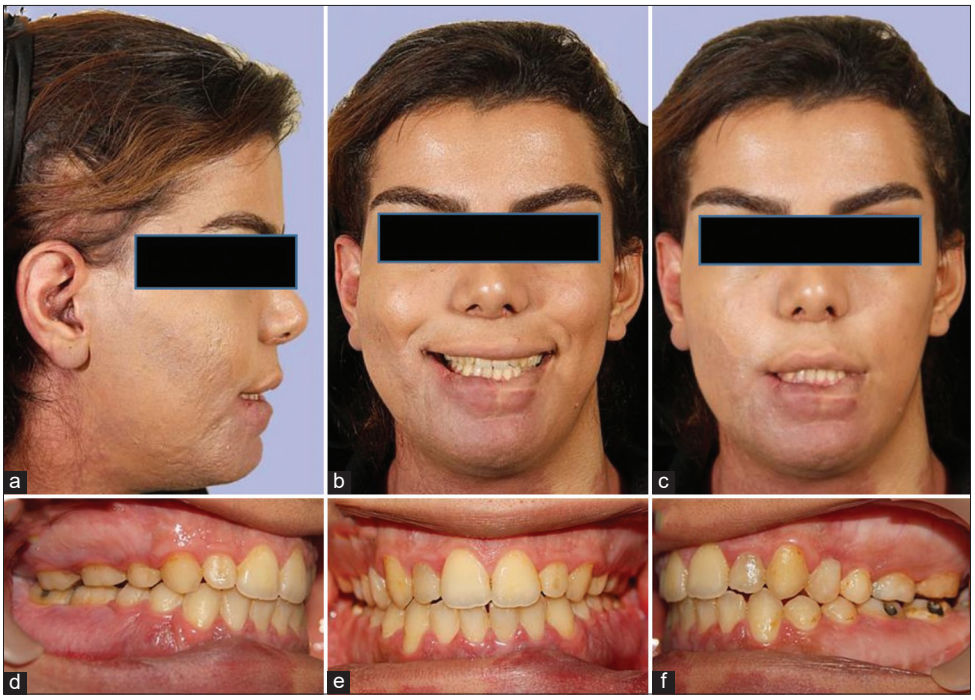

Figure 5: (a) Post-treatment lateral extraoral photograph. (b) Post-treatment smiling extraoral photograph. (c) Post-treatment frontal extraoral photograph. (d) Post-treatment right intraoral photograph. (e) Post-treatment frontal intraoral photograph. (f) Post-treatment left intraoral photograph.

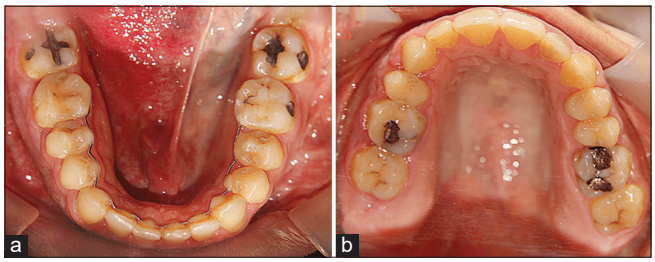

Figure 6: (a) Post-treatment occlusal view of mandibular arch. (b) Post-treatment occlusal view of maxillary arch.

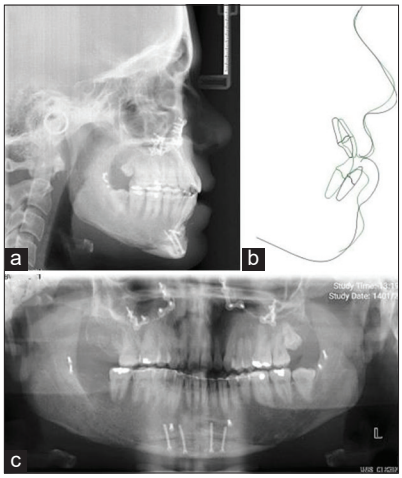

Figure 7: (a) Post-treatment lateral cephalometry. (b) Superimposition of pre-treatment (black) and post-treatment (green) cephalometric tracings. (c) Post-treatment panoramic radiograph.

hemihypertrophy and cutaneous lesions, such as CLOVES (congenital lipomatous overgrowth, vascular malformations, epidermal nevis, and skeletal anomalies), megalencephaly-capillary malformation syndrome, diffuse capillary malformation with overgrowth, fibroadipose overgrowth, proteus syndrome, Beckwith-Wiedemann syndrome, Maffucci syndrome, and plexiform neurofibromatosis in neurofibromatosis 1.[8]

Clinical signs and symptoms of KTS patients range widely, from mild hypertrophy, port wine stains, and a few venous varicosities to debilitating symptoms related to massive limb overgrowths, venous malformations, and bleeding disorders.[11] According to Auluck *et al.*, 5% of KTS cases might involve the orofacial region.[3] All of the aforementioned KTS characteristics can manifest in the orofacial region; however, varicosities are rare in this region as gravity facilitates venous drainage from the head and neck.[4] This explains why our case presented with varicose veins on her right thigh and not in the head and neck region.

A characteristic unilateral hypertrophy may affect tissues of the facial region. The resting pressure of an enlarged tongue may lead to the development of open bite, as with our case which presents with hypertrophy of the right half of the tongue but the left side lateral open bite, because the tongue lies toward the left. To allow for the closure of the open bite and prevent relapse, a permanent tongue guard was utilized during the pre-surgical phase, and a removable Hawley device with a tongue guard was used during retention.

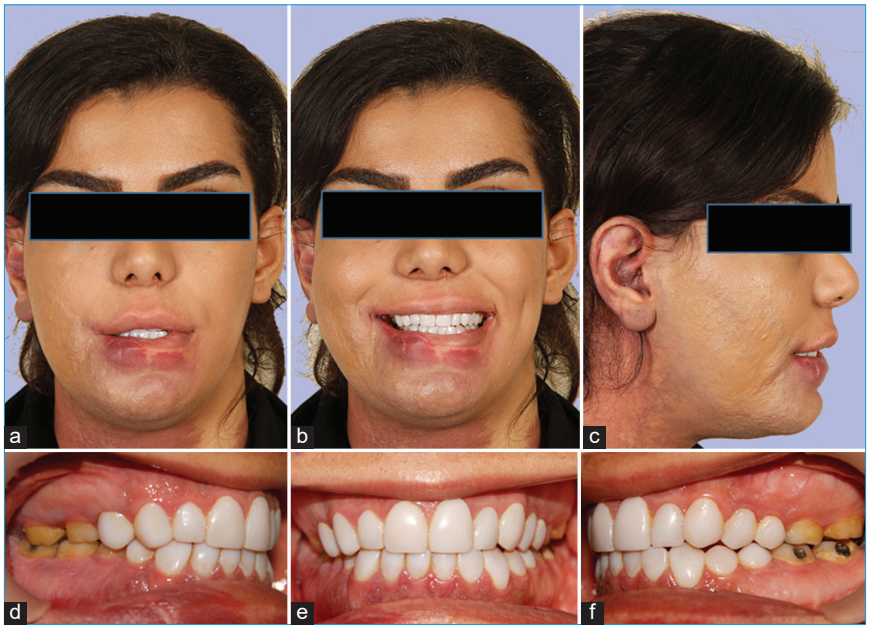

Figure 8: (a) Follow-up frontal extraoral photograph. (b) Follow-up smiling extraoral photograph. (c), Follow-up lateral extraoral photograph. (d) Follow-up right intraoral photograph. (e) Follow-up frontal intraoral photograph. (f), Follow-up left intraoral photograph.

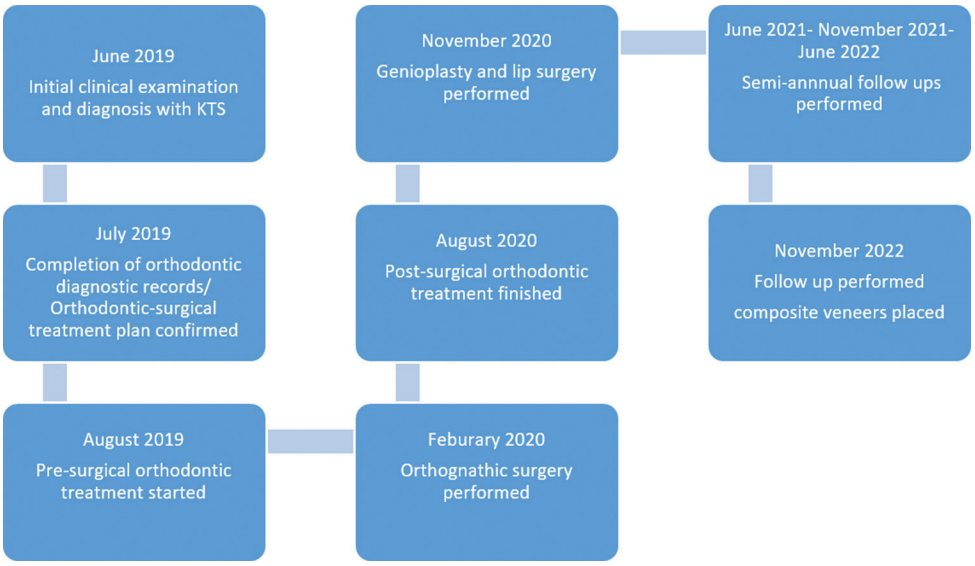

Figure 9: Timeline of patient's care.

The capillary malformation is the most frequently observed feature in KTS causing cosmetic concerns for the patient and is caused by dilated capillaries close to the skin's surface. Our patient presented with large areas of birthmarks over her body. Pulsed-Dye Laser (PDL) is currently the treatment of choice for these capillary malformations. The degree to which these lesions may improve, however, varies widely, and complete remission is uncommon.[12] For the birthmarks on her face, our patient underwent 4 sessions of 595-nm PDL treatment with 4 week intervals, which resulted in significant lightening of the lesions.

Considering vascular disorders among the characteristic of KTS, and few previous reports of prolonged bleeding in this syndrome, dentists, and oral surgeons must pay careful attention during practice on the highly vascularized area of the head and neck.[9,13] However, as in most reported cases of KTS, our case did not show any unusual bleeding or eventful healing.

CONCLUSION

Patients with KTS presenting with orthodontic problems can be managed with specific precautions. A successful treatment significantly improves the patients function, appearance, and overall quality of life.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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None.

Conflicts of interest

There are no conflicts of interest.

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