CASE REPORT

Dental management of Klippel–Trenaunay Syndrome

Afnan Al-Saleem a, Asma Al-Jobair b,*

a Dental Department, Riyadh Military Hospital, Saudi Arabia
b Department of Pediatric Dentistry and Orthodontics, College of Dentistry, King Saud University, Saudi Arabia

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Abstract Klippel–Trenaunay Syndrome (KTS) is a rare complex congenital disorder, characterized by two or more of the following: (1) capillary malformations (port-wine stains), (2) varicose veins or venous malformations (3) soft tissue or bony hypertrophy (or both). It is usually diagnosed in early childhood and has severe long-term sequelae. The orofacial region is rarely involved; however, a broad scale of dento-orofacial abnormalities may require specialized dental and anesthetic management. We are reporting a case of a 3-year-old Saudi girl with KTS, reviewing the literature, and outlining medical complications and dental considerations of this syndrome.

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1. Introduction

Klippel–Trenaunay Syndrome (KTS) is a rare complex congenital disorder, and was first reported in 1900 by Klippel and Trenaunay.1 It is characterized by two or more of the following: capillary malformations (port-wine stains), varicose veins or venous malformations, soft tissue or bony hypertrophy (or both).2 Klippel–Trenaunay Syndrome’s exact cause is not known. Most cases are irregular, although there is literature that reported an autosomal dominant pattern of inheritance.3 There is no reported gender or racial predilection.4 The syndrome occurs rapidly during the peripubertal phase, and the changes do not develop after puberty.4 This syndrome might be associated with other features such as decalcification of involved bones, lymphatic obstruction, poor wound healing, ulceration, polydactyly, macrodactyly, hyperhidrosis, spina bifida, paresthesia, stasis dermatitis, chronic venous insufficiency, thrombosis, and emboli.2 Gloviczki et al.5 reported that 5% of cases may involve the head and neck region. The patients with this condition may present with facial hemangioma (nevus flammeus) in the oro-facial region; early tooth eruption; hemangiomata of oral mucosa, tongue telangiectase, oro-pharyngeal mass, gingival hyperplasia; hypertrophy of soft tissue such as enlarged lip, cheek, and tongue; and jawbone hypertrophy causing asymmetry, malocclusion, and anterior open bite.5–9 Mental retardation and seizures are common in patients with KTS, especially those with facial angiomata.10

* Corresponding author. Address: Department of Pediatric Dentistry and Orthodontics, College of Dentistry, King Saud University, P.O. Box 60169, Riyadh 11545, Saudi Arabia. Tel.: +966 14784524.
E-mail address: aaljobair@ksu.edu.sa (A. Al-Jobair).
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We reported a case of a 3-year-old Saudi girl with KTS, reviewed the literature, and outlined the medical complications that may be associated with the dental treatment.

2. Case report

A 3-year-old Saudi girl was seen at Pediatric Dentistry Clinics, Security Forces Hospital in Riyadh, Saudi Arabia. Her complaint was dental caries which need fillings. This was her first dental visit. Her parents were first degree cousins and youngest among two siblings. The mother had full term pregnancy and was delivered by caesarian section. There was no history of a similar family condition.

At birth, hemangioma was present on the right lower limb. At one month, red discolorations and brown hyperpigmentation started to appear bilaterally in the upper arms, legs and feet. In addition, hemi-hypertrophy started to be visible. After complete investigations and consultations, the case was diagnosed as KTS at 10 months. At 2 years of age, she had an abdominal swelling and she was admitted at the hospital due to an internal bleeding. At present, she is under recall neurological visits. Dietetic history was evaluated which includes frequent daily sugar intake.

The patient appeared to be healthy, stable, communicative and intelligent. Extra-oral examination revealed an extensive flat cutaneous hemangioma on the left half of the neck and trunk and on both arms, hands, legs, and feet. The left hand was of average size for the patient’s age, but the right hand was larger and darker in color (Fig. 1). All five fingers of the right hand were uniformly large. Her right foot was large in size but her left foot was abnormally large with macrodactyly and elongated toes (Fig. 2). The right side of the face was slightly larger than the left side but without severe facial asymmetry (Fig. 3).

Oral examination revealed no intraoral hemangioma, nor soft tissue enlargement. Complete primary dentition was present. The maxillary arch appeared enlarged with spaces noted between all maxillary teeth. Teeth were normal in shape and position with multiple carious lesions. The stage of eruption of the primary teeth was normal and according to the age. Oral hygiene was fair with visible plaque in anterior teeth and gingival inflammation was moderate. Radiographic examination revealed normal and symmetrical teeth developments on both sides of the primary and permanent dentitions (Fig. 4).

The girl was apprehensive during examination and refused dental treatment in the next visit. Due to the lack of cooperation and the need of multiple restorations, a decision was made to treat the patient under general anesthesia (GA). After consultations and complete investigations, clearance was made for the GA and the patient was operated under GA. Composite restorations, pulp therapy, and stainless steel crown restorations were performed according to the treatment plan (Fig. 5). The patient left the operating room in good condition and was discharged without any complication.

3. Discussion

Klippel–Trenaunay Syndrome is a term used to describe the combination of cutaneous capillary malformation, varicose veins, and hypertrophy of bone and/or soft tissue. At least two signs should be present to establish the diagnosis. The symptoms can be of varying degrees, ranging from minimal
This patient presented with hemangioma which was present at birth and skin discoloration in the lower and upper limbs, trunk and neck. Ninety-seven percent of KTS patients have hemangioma and the most common type of hemangioma is flat cutaneous hemangioma as it is manifested in the presented case. The onset of the first symptom, hemangioma in the majority of cases, varied from being present at birth to detection at the age of 6 years. In addition, hyperatrophy of the upper right limb and lower left limb was manifested in this patient which is extremely rare. The crossed-bilateral involving the upper and lower extremities occurs in only 2.5% of KTS cases. The varicosity was not significant as the patient is too young and future development of a clinically significant varicosity might still be expected to occur. It occurs in 62% of individuals of KTS.

Besides these main symptoms, additional common features are manifested in this case. The patient has macrodactyle of the second toe in the left foot. Digital deformity of hands or feet is considered to be a common finding in KTS patients. McGrory et al. found that macrodactyle of the second toe is the most common digital deformity in KTS patients. At 2 years of age, the patient suffered from internal bleeding which required hospital admission. This indicates vascular involvement of visceral organs. Visceral organs such as the GI tract, liver, spleen, bladder and kidney may be involved in the angiomatous malformations leading to a variety of manifestations depending on the organ involved. The well-known manifestations of this syndrome are hematochezia, hematuria, and spontaneous internal bleeding.

The right side of the face was slightly larger than the left side but without severe facial asymmetry, which might indicate maxillary hypertrophy. No oral manifestation of KTS was present in this case other than spacing between the maxillary teeth, as she presented to the dental clinic at a young age. Most of the reported cases with oral manifestations are for children at age of 5 years and above. Teeth eruption was according to the patient’s age. Radiographically, the developmental stage of permanent teeth was also according to the patient’s age. Mueller-lessmann et al. reported early eruption of permanent teeth in KTS patients. However, Bathi et al. reported retained deciduous teeth and impacted permanent teeth in some patients with KTS.

The patient was presented with multiple carious lesions which are mainly due to high sugar intake. Caries prevalence in KTS patients was not reported in previous studies, however, KTS patients are at a high risk of developing carious lesions. The presence of malocclusion and gingival hypergrowth is leading to difficulty in maintaining good oral hygiene and subsequently developing dental caries.

Although the patient was mentally fit, she did not show enough cooperation to be treated in dental chair under local anesthesia. Because of that, a decision was made to treat the patient under general anesthesia (GA). Several components of KTS may complicate anesthesia and dental intervention. A bleeding diathesis represents a potential hazard in the management of KTS. Persons affected may have thrombocytopenia due to platelet sequestration in oral hemangiomas. Patients with KTS may pose difficulty with airway management. Surgery related to severe KTS may be associated with massive hemorrhage and the anesthesiologist should anticipate...
the need for appropriate fluid resuscitation. To prevent possible postoperative bleeding problem in all patients with KTS, a detailed hematological investigation must be ordered by the dentist prior to any oro-surgical procedure even without oro-facial involvement. In this patient, the laboratory investigations (urinalysis, complete blood count, blood biochemistry) did not reveal any abnormality. There was no contraindication for GA after complete investigations and consultations. The patient was operated under GA using naso-tracheal intubation. No complication during or after GA and the patient was discharged in good condition.

Attention must be focused on the dental management of these patients. The three most common oral findings all have implications for the dentist. Early dental development and eruption necessitate orienting any dental treatment to the patient’s dental rather than chronological age. Hypertrophy of the bone and/or gingiva could create severe malocclusions and present difficulties in designing and utilizing prostheses. Hemangiomatic areas that need any surgical procedures require extra attention regarding hemorrhage control.

In conclusion, it is very important to evaluate patients with KTS at a young age to introduce preventive procedures and treat the teeth as early as possible to avoid extraction and subsequent excessive hemorrhage and delayed healing. Although KTS is rare, general practitioners and pediatric dentists should be aware of the general and oro-facial manifestations in patients who have this syndrome.

References