Klippel-Trénaunay Syndrome Manifesting as Gingival Overgrowth and Teeth Agenesis

Hakan Ozdemir * / Ismail Marakoglu ** / Melih Akyol ***/ O Fahrettin Goze **** / Ulvi Kahraman Gursoy *****

Objective: *Klippel-Trénaunay syndrome (KTS) is characterized by triad of venous varicosity, naevus flammeus, and soft/hard tissue hypertrophy. Manifestations of the syndrome in the head and neck region are rare, but in some cases hemangioma of the lips, tongue, and gums, open bite and cross bite, and early tooth eruption are associated with the disease.* ***Study design:*** *We report a 12-year-old KTS patient with gingival hyperplasia, congenital missing teeth, and increased mucosal vascularization as oral manifestations.* ***Results and conclusion:*** *All manifestations of the KTS were observed on the same side of the head, except bilateral missing teeth. Histological examination showed several vascular enlargements in enlarged gingiva. It is suggested that oral manifestations of the syndrome are generally related to the severity of the disease, but they do not always present in the same pattern.*

Keywords: *Klippel-Trenaunay syndrome, congenital missing teeth, gingival overgrowth*
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INTRODUCTION

Klippel-Trenaunay syndrome (KTS) is a congenital anomaly classically defined as the triad of vascular stain, soft tissue and/or bone hypertrophy, and venous varicosities. KTS patients usually have these three features together, however diagnosis can be made with two of them.[1] The occurrence of KTS is rare in head and neck (5%), and most common oral features are hemangiomas of the lips, tongue, and gums; hypertrophy of the jaws; a deep palatal vault; open bite and cross bite; and early tooth eruption.[2, 3]

In this case report we present a KTS patient with vascular stains, soft tissue hypertrophy, gingival overgrowth, and congenital missing teeth in the absence of venous varicosities.

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Case Report

A 12-year-old Caucasian male diagnosed with KTS by the dermatology department was referred to dental department of Cumhuriyet University because of gingival swelling and caries problems.

Extraoral examination showed diffuse port wine-colored stains on all over the left side of his body including face, chest, and upper and lower extremities. Medical history confirmed that the stains were congenital. Stains were not protuberant, and neither achy nor itchy. However, lower lip and left thumb were swollen and covered with stains. Also, left thumb had abnormally developed fingernail. Patient was otherwise systemically healthy, and had no mental or developmental retardation (Fig. 1). All possible systemic diseases were ruled out after laboratory analysis of peripheral blood and physician consultation. No other family member of the patient was diagnosed with KTS. Consultation with a geneticist proved further genetic tests unnecessary.

Intra-oral examination showed class II canine and super class I molar dental relation on the right side and class II canine and class I molar dental relation on the left side and severe anterior deep bite. Panoramic radiograph showed that the patient had permanent dentition with an impacted canine (d. 13), a retained deciduous canine (dd. 53), and missing premolars (dd. 35, 45). Crowns and roots of the teeth were anatomically normal, and well developed. There were also accompanying caries. A secondary caries under an old amalgam filling on the mandibular left first molar tooth (d. 36) caused periapical inflammation. Endodontic treatment for this tooth was completed one day before the patient was first seen in periodontal clinic; therefore, radiolucency on the root apex was still visible on radiograph (Fig. 2). Maxillary

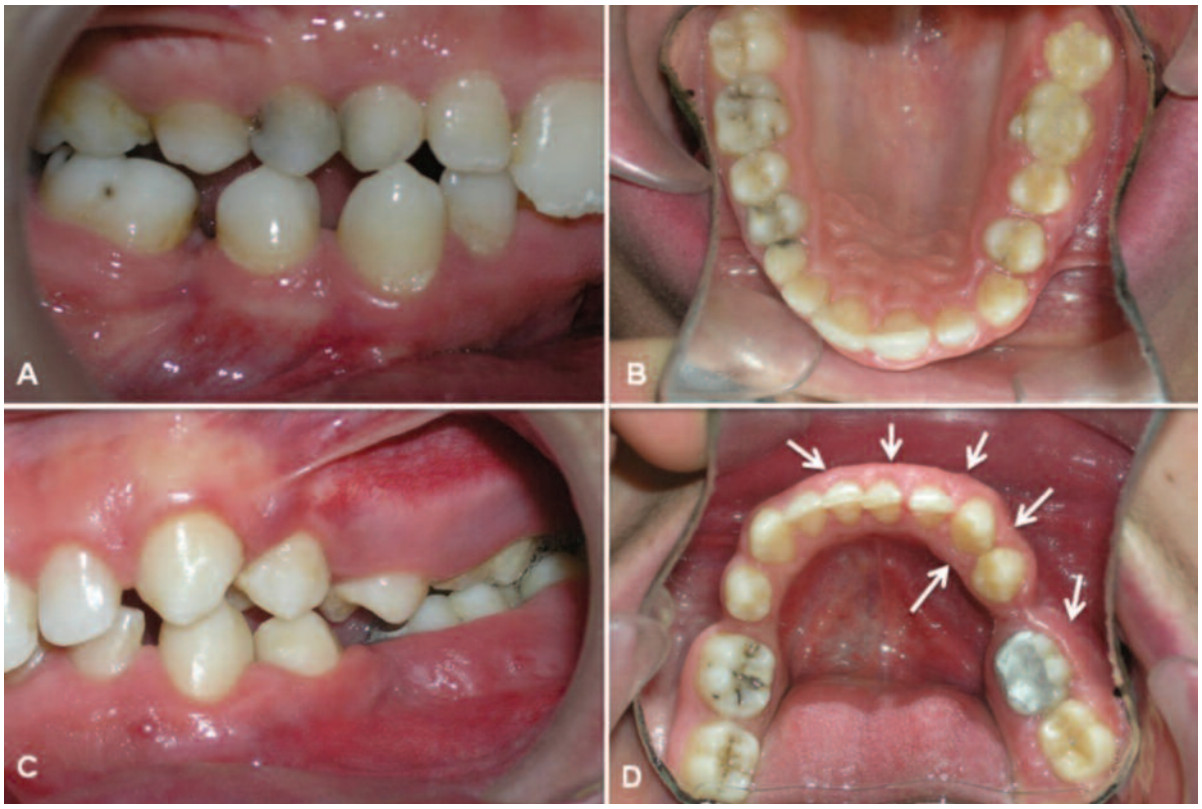

Figure 1. Oral findings of a Klippel-Trénaunay syndrome patient. Even though the dental malformations were bilateral, gingival enlargements (arrows) and increased mucosal vascularizations was prominent on the left side of the mouth. A: right buccal, B: maxilla, C: left buccal, D: mandibule

mucosa was clinically normal and healthy but mandibular gingiva was overgrown and had increased vascularization that was especially localized on the anterior left side. Gingival overgrowth was fibrous, pink colored, and had a firm structure. There was overgrowth starting from the distal region of the canine (d. 43) and extending to the distal area of second molar (d. 37). Overgrowth was prominent particularly on the buccal/labial side of the mandible. Gingiva had no tendency to bleed and no probing depth more than 3mm was measured. Vascularization was prominent on the lower lip, involving the same region with overgrowth, and also affecting the base of the mouth. Mucosal area had no tendency to bleed but had clear red to purple color (Fig. 3). Patient was not receiving any immunosuppressive,

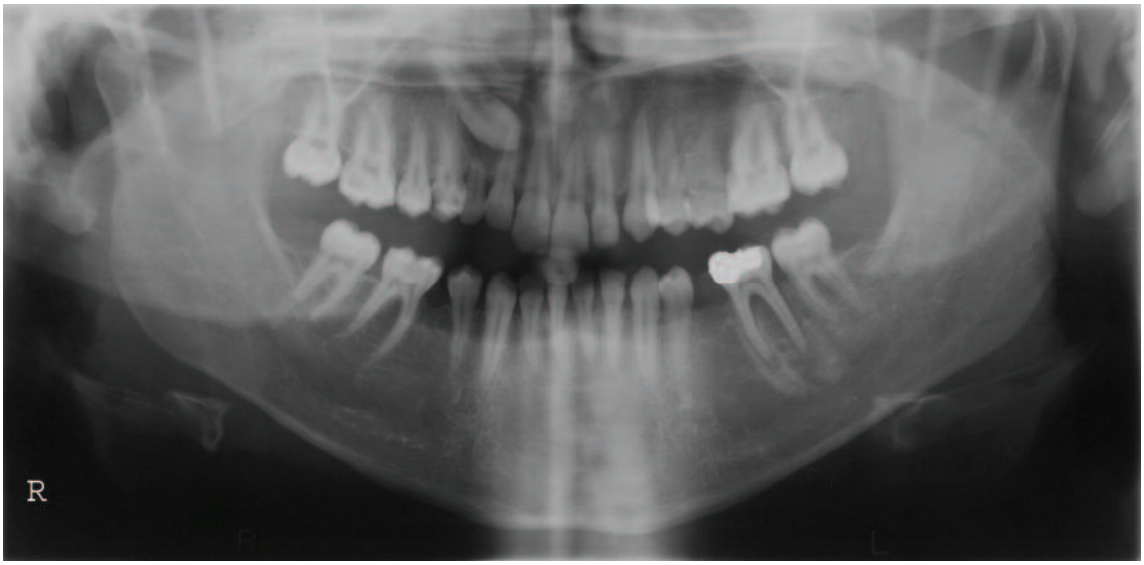

Figure 2. Panoramic radiograph from the initial visit. Mandibular premolars were missing and maxillary canine was impacted (R: right).

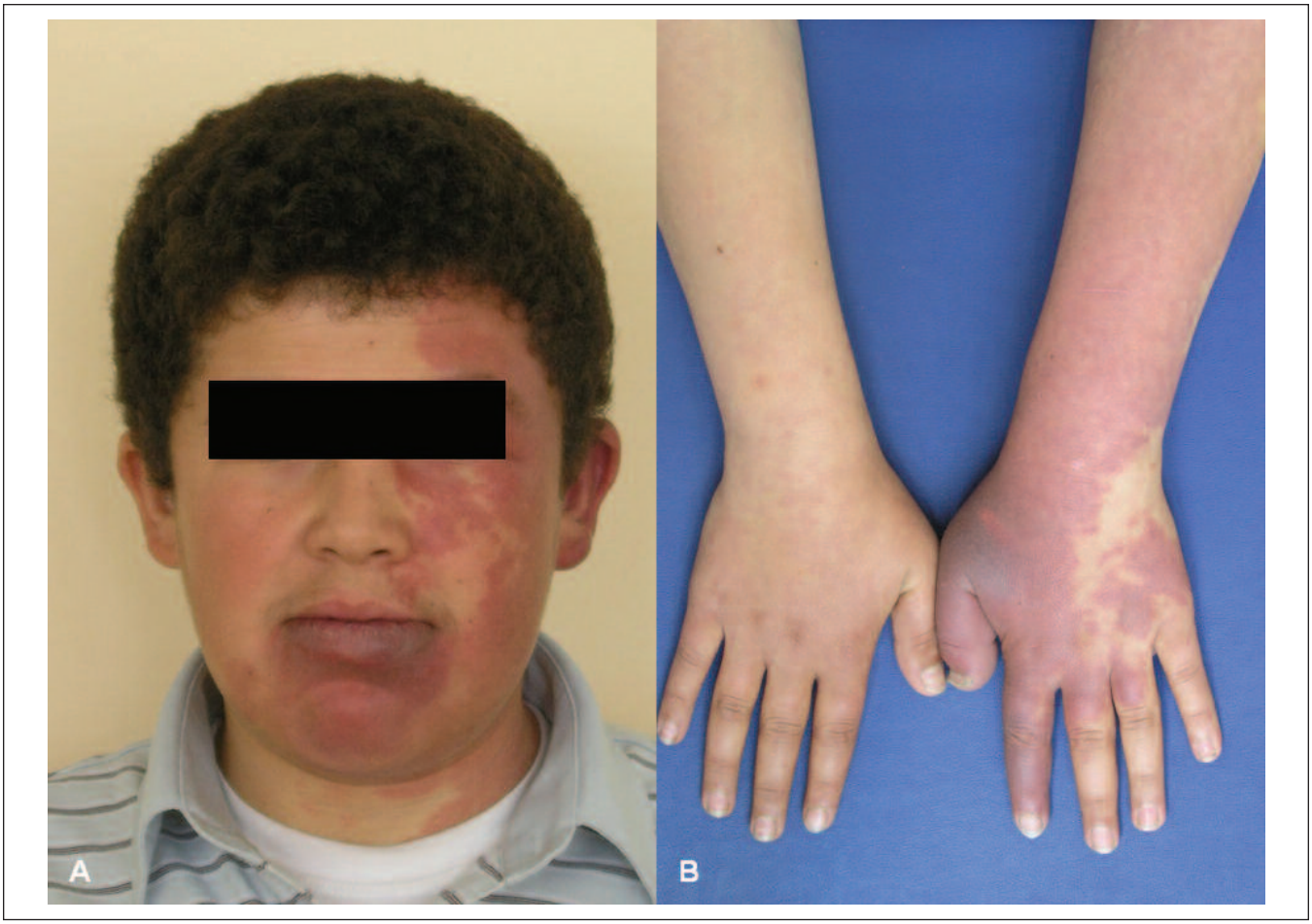

Figure 3. Diffuse Port-wine stains on the left side of the patient's body and soft tissue enlargements on his lip and thumb.

antiepileptic, or antihypertensive medications that were known to cause gingival overgrowth. Also, local factors such as mouth breathing were not contributory.

All teeth with caries were treated before the patient received any periodontal therapy. After establishing proper oral hygiene, hypertrophic gingiva was excised by gingivectomy and gingivoplasty techniques in order to restore ideal gingival form. Gingivectomy was performed under local infiltration anesthesia with 4% articaine (Ultracain DS, Aventis Pharma, Germany) using Kirkland 15/16 gingivectomy blade. Excised tissues were analyzed histologically. Histological examination showed thick epithelial rete pegs elongating into connective tissue, capillary vascular proliferation deeply extending into lamina propia, and partly band shaped inflammatory infiltration rich in plasma cells in subepithelial area that was intensified in perivascular areas (Fig. 4).

Simultaneously with the dental treatments, medical treatments were initiated to eliminate port-wine stains. However, the patient did not want to continue the medical treatment, due to the discomfort he felt during the therapy.

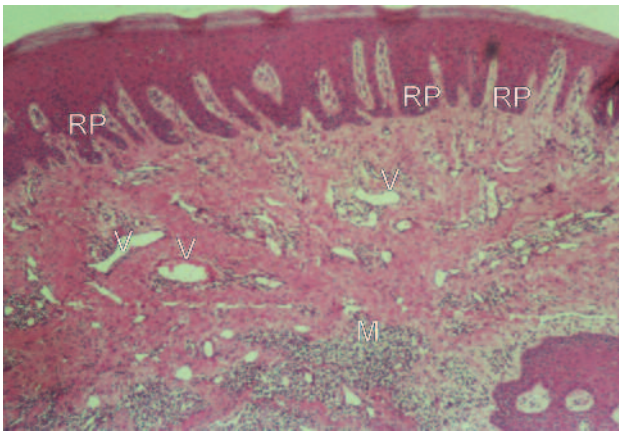

Figure 4. Histological view from gingiva. Increase in vascularization and enlargement of veins were prominent, along with elongated epithelial extensions (Rete pegs) into connective tissue. (H&E X10). (V: vein, RP: rete peg, M: mast cell)

DISCUSSION

Geoffroy-Saint Hilaire was the first to report on a case of KTS in 1832 and in 1900, Klippel and Trenaunay gave the first description of the triad as a specific clinical entity. Klippel-Trenaunay syndrome and Klippel-Trenaunay-Weber syndrome both appear interchangeably in the literature, along with the term angioosteohypertrophy.[2]

Orofacial localization of hyperplasia can occur in KTS. Increase in the dimensions of both hard and soft tissues,

including lips, cheeks, tongue and teeth, has been reported.[3] The present case had diffuse vascular stains (Port wine stains) and soft tissue hyperplasia, particularly in the thumb, lip and gingiva. Varicose veins and hard tissue hypertrophy were not found. One of the explanations for finding the venous malformations on mandibular lip of KTS patients is gravity, as suggested by Bathi *et al*.[3] Venous drainage malformations might also be the cause of gingival hyperplasia, as we found various enlarged veins in histological analysis of gingiva. Additional findings in KTS can be jawbone asymmetry, early tooth eruptions and dental abnormalities.[3,4] Our patient had retained primary canine and impacted maxillary permanent canine, anterior deep cross-bite, malocclusion in the posterior area, and missing second mandibular premolars. Premolars were missing on both sides. This evidence may allow us to suggest that there are no typical dental consequences of KTS, but it is related to several dental abnormalities.

Even though the literature suggests clinicians to be cautious in dental managements of the KTS patients because of excessive hemorrhage, we observed normal hemorrhage after gingivectomy procedures. We think that excessive hemorrhage seen in KTS patients might be related to venous varicosities, which our patient did not have. But we still accept the suggestion of providing the highest care of KTS patients during dental treatments.

KTS should be differentiated from other congenital vascular anomalies such as Parkes-Weber and Sturge-Weber syndromes. Parkes-Weber syndrome has similar features with KTS, but has additionally arterio-venous fistula.[3-5] In Sturge-Weber syndrome there are craniofacial angiomatosis and port-wine nevus along the branches of trigeminal nerve, with cerebral calcification, sensory and paramotor paralysis, and calcification of vessel walls.[3] Our patient had none of these additional features. Peripherally, KTS should be distinguished from Recklinghausen's disease, Maffuci's syndrome, Proteus syndrome, and Wiedemann-Beckwith syndrome; since they display prominent differences.[4] After evaluation of the clinical, radiographic, and histological findings of this case, we suggest that soft tissue enlargements related to KTS may also involve gingiva, and malformations in vascular structure can be the reason for this outcome, and congenitally missing teeth may be associated with the syndrome. However, the severity of the oral findings may increase with the involvement of varicous veins and hard tissue hypertrophy.

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