doi:10.5368/aedj.2010.2.4.180-182.pdf

PROMINENT OROFACIAL FINDINGS IN KLIPPEL-TRENAUNAY SYNDROME.

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ABSTRACT

Klippel-Trenaunay syndrome is a triad of congenital anomalies characterized by nevus flammeus, varicosities, and unilateral bony and soft tissue hypertrophy. Orofacial manifestations include facial asymmetry, jaw enlargement, and malocclusions as well as premature tooth eruption. Presented here is an illustrative report of Klippel Trenaunay syndrome in a 30 year old male patient showing all the characteristic findings.

***KEY WORDS*: Klippel Trenaunay syndrome, KTS, Nevus flammeus, Facial Hemi hypertrophy**

INTRODUCTION

Klippel-Trenaunay syndrome (KTS),first described in 1900 by two French physicians Klippel and Trenaunay, is a rare, congenital, vascular disorder described by the presence of a combined vascular malformation of the capillaries, veins, and lymphatics; congenital venous abnormalities; and limb hypertrophy. It is characterized by a triad of port-wine stain, varicose veins, and bony and soft tissue hypertrophy involving an extremity[1,2]. This is a rare sporadic condition and there are no available figures for its annual incidence or prevalence in the population[1].

Case report

A 30 year old male reported to our department with chief complaint of facial discoloration on the left side since birth along with abnormally large left part of upper lip since past few years. No significant medical, dental, personal or family history was present.

On general examination, all the vital signs of the patient were normal. Port wine stain were present on the left side of the face extending from philtrum region, involving left lateral part of the nose and reaching up to the left periocular region **(Fig.1).** Intraorally, the port wine stain was showing characteristic ipsilateral distribution affecting left buccal mucosa, soft palate and gingiva **(Fig. 2).** Gingival hypertrophy was present on the affected side with moderately deep periodontal pockets and downward placement of upper left posterior teeth **(Fig.3).** Increased height of the alveolar ridges and marked increase in the depth of the buccal vestibule was appreciated in the left quardrants. Hard tissue examination revealed marked proclination of upper incisor teeth along with spacing due to arch length and tooth size discrepency. There was downward and buccal placement of left maxillary teeth resulting in canting of occlusion **(Fig.4).** Grade I mobility was noticed in lower anterior teeth.

On radiographical examination, orthopantomogram showed left maxillary hypertrophy. Three dimensional reconstruction of CT confirmed the hypertrophy of maxilla on affected side. A diagnostic search for a clinical condition presenting with a triad of port-wine stain and bony and soft tissue hypertrophy confirmed the case as Klippel Tranaunay syndrome.

Discussion

KTS is a rare sporadic condition with no racial or geographic predisposition. Males and females are affected equally, and no racial predominance exists. Most patients demonstrate all three signs of the clinical syndrome: port-wine stain, varicose veins, and bony and soft tissue hypertrophies[1-3]. The capillary hemangioma or port-wine stain usually presents first having a distinct, linear border that respects the midline. It is often noted on face or lateral aspect of the limb having variable depths limited to the skin or extends deeper to subcutaneous tissue, including muscle and bone.

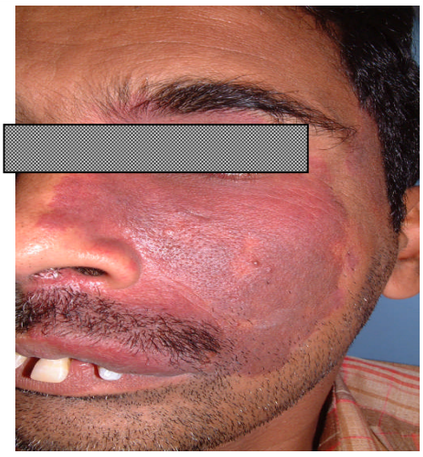

Fig.1.Extra oral portwine stain

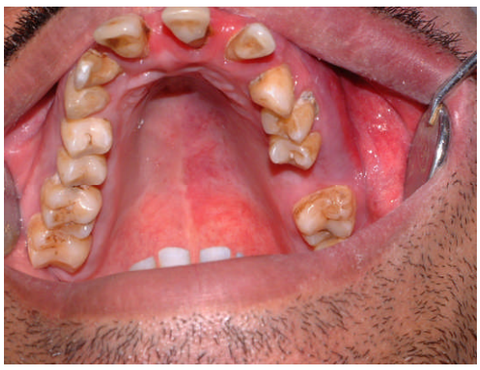

Fig.2.Involvement of palate, buccal mucosa and gingiva

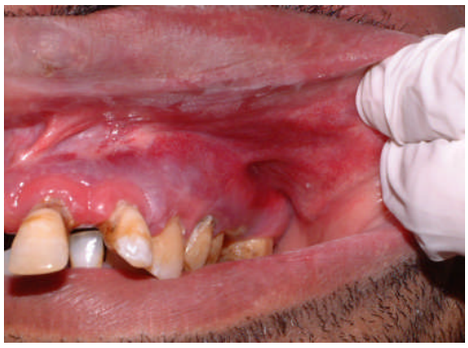

Fig.3. Gingival hypertrophy with moderately deep periodontal pockets and downward placement of upper left posterior teeth

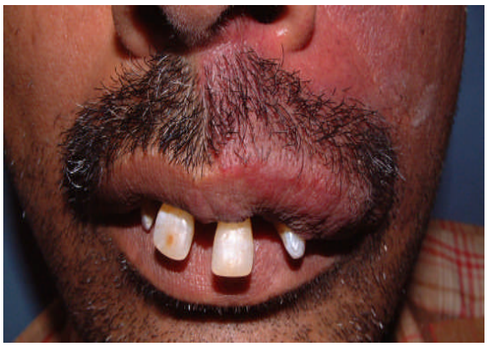

Fig.4.Malocclusion with hypertrophied left part of upper lip.

It is typically of the nevus flammeus type, but cavernous hemangiomas or lymphangiomas may also occur. Nevus flammeus is a salmon pink patch, sometimes with a verrucous quality, which evolves to a deep purple color with time[1-3].

Varicose veins in KTS are congenital. Bony and soft tissue hypertrophies are the third sign of KTS. Other features include lymphatic obstruction, spina bifida, hypospadias, polydactyly, syndactyly, oligodactyly, hyperhidrosis, hypertrichosis, paresthesia, decalcification of involved bones, chronic venous insufficiency, stasis dermatitis, poor wound healing, ulceration, thrombosis, and emboli. There has also been a reported case of portosystemic encephalopathy and a cerebral haemangiopericytoma[1,4].

Orofacial findings correspond to the bony and soft tissue hypertrophy of the upper and lower jaw resulting in facial asymmetry and malocclusion. Typical features include hemangiomas of the lips, tongue and oral mucosa along with premature eruption and accelerated growth of teeth on the affected side.[2,3] In our case, the hemihypertrophy of the left side of the maxilla was clinically visible. Malocclusion in our patient corresponds to anterior proclination along with spacing between the teeth due to arch length- tooth size discrepancies.

Differential diagnosis of this syndrome include *Parkes-Weber syndrome* (where there is a high-flow arteriovenous malformation rather than capillary haemangioma), *Sturge-Weber syndrome* (facial port-wine stain, epilepsy and hemiparesis), *Maffuci syndrome* (rare dysembryoplasia causing cartilage and vessel tumors), *Proteus syndrome* (rare hamartomatous disorder causing asymmetrical hypertrophy of a range of tissues') [1,3].

Evaluation of the deep venous system can be completed with duplex scanning contrast venography, ultrasonography, contrast venography and arteriography, and nuclear MRI studies. Arteriography is especially helpful in the diagnosis of an arteriovenous fistula. MRI is also helpful in imaging the soft tissue hypertrophy. In addition, magnetic resonance angiography can be very helpful in identifying and defining vascular malformations. Prenatal diagnosis by ultrasonography has been reported. Orthopantomograms and lateral cephalograms show the jaw and occlusal abnormalities[1,4,5].

Most patients with KTS can be treated conservatively with compression stockings or pneumatic pumps which decrease edema, act as a barrier for minor trauma, and reduce venous insufficiency. Medications that can be prescribed are antiplatelet agents like aspirin and corticosteroids. Surgical intervention which have been successfully tried are resection or ligation of abnormal blood vessels and intravenous sclerotherapy[1,4,6,7].

CONCLUSION

Oral findings in Klippel-Trenaunay syndrome are common and thus oral physicians and dentists must be vigilant and aware of the potential complications of dental or surgical procedures. The major problems associated with dental management of patients with this syndrome are excessive haemmorrhage from any oral hemangiomatotic lesions and delayed healing of surgical wounds.

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