# KLIPPEL-TRENAUNAY SYNDROME: A RARE BLEEDING RISK

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## Introduction

Klippel-Trenaunay syndrome (KTS) is a rare congenital malformation of blood vessels, soft tissues and bones[1,2]. It has an incidence of 1 in 20-40,000[3]. The syndrome has three classic features: port-wine stains, venous malformations (e.g. varicose veins) and overgrowth of soft tissues and bone, most commonly affecting the legs or torso during infancy.

Most cases of KTS are caused by somatic mutations in the *PIK3CA* gene resulting in increased cellular proliferation within affected tissues[4]. The condition is not known to be inherited and there is no curative treatment. Management is conservative to reduce risk of complications, such as bleeding and thromboembolism.

## Presentation

A 26-year-old female was referred by her General Dental Practitioner to the Oral Surgery department for extraction of a grossly carious right mandibular third molar (tooth 48) with a potentially close relationship to the Inferior Dental Nerve (IDN).

Background medical history included KTS and associated thromboembolic disease, depression and anxiety. The patient was a non-smoker. On examination, there was a bilateral lower facial overgrowth and a port-wine stain affecting the chin and neck (Figure 1).

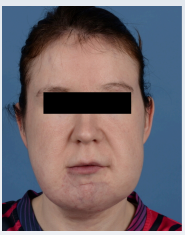

***Figure 1***

There were clusters of intra-oral vascular bundles, including a highly vascularised, erythematous section of mucosa immediately adjacent to the unrestorable mandibular right third molar (Figure 2). There was limited tongue movement and evidence of haemangiomas (Figure 3).

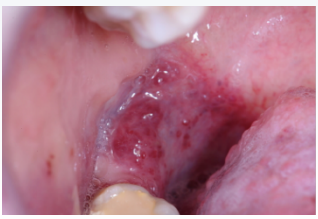

***Figure 2*** – *Highly vascular mucosa adjacent to LR8*

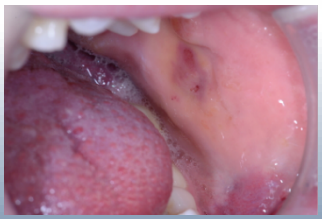

***Figure 3*** – *Haemangioma left buccal mucosa*

## Special Investigations

A panoramic radiograph revealed the grossly carious right mandibular third molar with conical roots and a potentially close relationship to the IDN, as exhibited by interruption of the superior border of the ID canal.

CBCT was not justified as coronectomy was contraindicated .

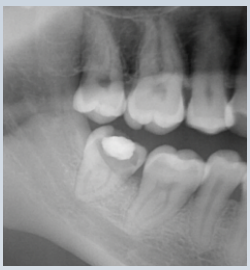

***Figure 4*** – *Panoramic radiograph showing tooth 48 and its relationship with the ID canal.*

Review of historic MRI scans revealed the extent of the vascular malformation present within the head and neck (Figure 5, 6).

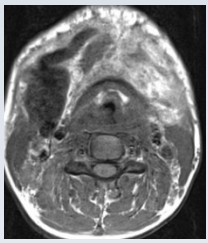

***Figure 5*** – *extensive venous varix inferior to right submandibular gland.*

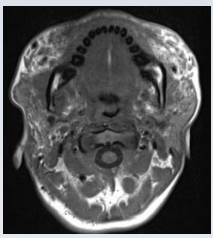

***Figure 6*** – *Lymphovascular malformations clearly illustrated in buccal regions.*

## Discussion

Patient safety is a priority in clinical practice. The option of coronectomy was contraindicated due to the extent of caries within the tooth and the morphology of the roots increases the risk of root mobilisation during a coronectomy procedure.

The patient was referred to a tertiary Oral and Maxillofacial unit for safe extraction of the lower right third molar in an environment with inpatient and emergency facilities.

## Conclusion

Although this is a rare condition, it is of great importance to surgical practitioners; and it is essential to recognise intra-oral vascular malformations which may increase the risk of significant peri and post-operative haemorrhage.

## References

1. Sung HM, Chung HY, Lee SJ, et al. Clinical Experience of the Klippel-Trenaunay Syndrome. *Arch Plast Surg.* 2015;42(5):552-558. doi:10.5999/aps.2015.42.5.552

2.Klippel-Trenaunay Syndrome (KTS) Information Page. *National Institute of Neurological Disorders and Stroke.* 2019. https://www.ninds.nih.gov/Disorders/All-Disorders/Klippel-Trenaunay-Syndrome-KTS-Information-Page

3. Klippel-Trenaunay syndrome. *Gosh.nhs.uk.* 2015. https://www.gosh.nhs.uk/conditions-and-treatments/conditions-we-treat/klippel-trenaunay-syndrome

4. Klippel-Trenaunay syndrome | *Genetic and Rare Diseases Information Center (GARD) – an NCATS Program.* Rarediseases.info.nih.gov. https://rarediseases.info.nih.gov/diseases/3122/klippel-trenaunay-syndrome