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CASE REPORT

Epulis granulomatosa as an oral manifestation of Klippel-Trénaunay syndrome

Sebastian R. J. M. Kühl[1], Ralf K. W. Schulze[1], Andreas Kreft[2], Bernd d'Hoedt[1]

Departments of [1]Oral Surgery and [2]Pathology, Johannes Gutenberg-University, Mainz, Germany

The Klippel-Trénaunay syndrome (KTS) was first described by Klippel and Trénaunay in 1900. It is characterized by the triad of hemihypertrophy of soft and hard tissue, naevus flammeus and venous varicosity in the affected area. Though all oral tissues may be affected, only 5% of KTS show manifestations in the head and neck region. Only three cases are described with an oral manifestation, showing gingival overgrowth clinically and histologically corresponding to a pyogenic granuloma. It is still uncertain whether the combination of gingival fibromatosis and KTS is significant or coincidental. We report about a 25-year-old patient with KTS and recidivous gingival fibromatosis, clinically and histologically corresponding to an epulis fibromatosa in a case report. It is suggested that this occurrence is significant.
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Introduction

The Klippel-Trénaunay syndrome (KTS) is characterized by the triad of hemihypertrophy of soft and hard tissue, naevus flammeus and venous varicosity in the affected area (1–3). Identification of KTS can cause difficulties because of extreme degree of variability of symptoms (2, 4). Though all oral tissues may be affected, only 5% of KTS show manifestations in the head and neck region (5). It is still uncertain whether the combination of gingival fibromatosis and KTS is significant or coincidental. We report about a 25-year-old patient with KTS and recidivous gingival fibromatosis, corresponding to an epulis fibromatosa in a case report. It is suggested that this occurrence is significant.

Correspondence: Dr S. Kühl, Poliklinik für Zahnärztliche Chirurgie, Augustusplatz 2, D-55131 Mainz, Germany. Tel: +49 6131 175090, Fax: +49 6131 173434, E-mail: sebastiankuehl@web.de
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Case description

A 25-year-old man with KTS was referred because of gingival swelling in region 13/14 and 18/19 in 2001. Extraoral findings are typical for KTS. Venous varicosity and a partial gigantism especially affecting the acres were limited to the left side. Naeuvus flammeus was limited to the left midfacial region, especially affecting the periorbital region (Fig. 1). The tissue affected by naevus flammeus showed slight hypertrophy when compared with the non-affected side and additional hypertrophy of the left lower an upper lips was recognizable. Venous varicosities were only found at the left hand and feet.

Intra-oral examination revealed a soft swelling of 1×0.5 cm in region 13/14, and of 0.5×0.5 cm region 18/19, both limited to the buccal marginal interproximal gingiva (Figs 2 and 3). Surfaces showed signs of inflammation with clear margins. Bleeding was easily provocable by mechanical stimulus. Probing depths distally 13 was 10 mm with a loosening of grade II. A panoramic radiograph (OPTG) and a conventional X-ray image 13 (Fig. 4) show a vertical loss of bone interproximal 13/14 and 18/19 with clearly visible, enlarged periodontal space 13. A magnetic resonance image (MRI) and a computerized tomography (CT) of the head were advised which revealed hypertrophy of bony and soft tissue limited to the left side. No evidence for haemangioma of the maxillary and mandibulary was found.

A total excision of the hyperplasia 13/14 was performed during 2003 in local infiltration anaesthesia with 4% articaine and 1:200 000 adrenaline (Ultracain DS: Aventis Pharma, Frankfurt, Germany) using a CO_2 laser. Histopathological examination of the excised hyperplasia revealed a pyogenic granuloma with focal ulcerated inflammatory tissue (Fig. 5) corresponding to the clinical diagnosis of an epulis granulomatosa.

The patient was lost for follow up after suture removal 7 days post-surgery until 2005. At this time, the patient represented again because of a recidivous swelling in region 13/14 which had a more fibrous appearance. Size, colour, surface and consistence were equivalent to the removed tissue described before. Again

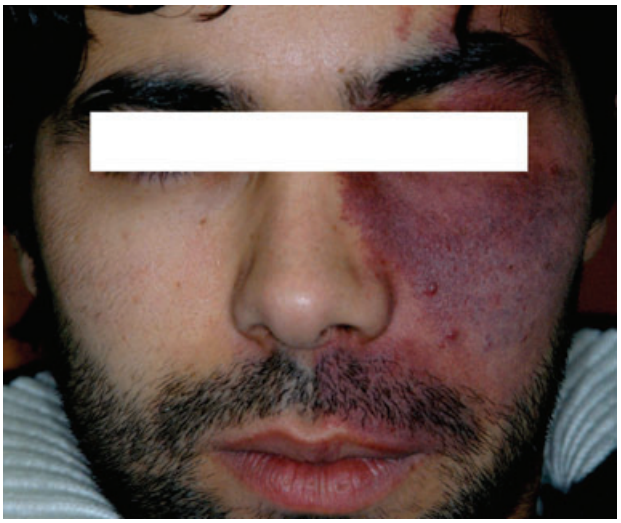

Figure 1 Midfacial naevus flammeus with slight hypertrophy of the affected tissue.

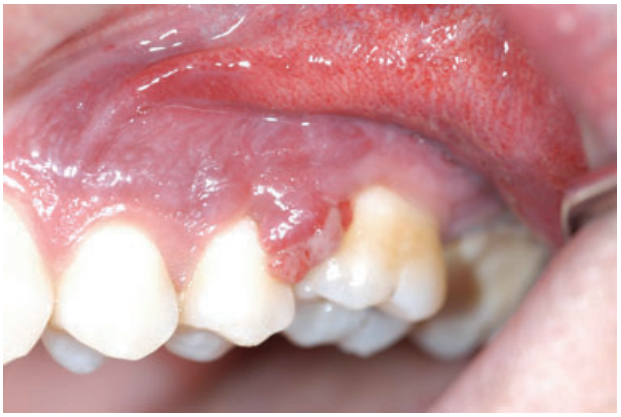

Figure 2 Epulis fibromatosa interproximal 13/14.

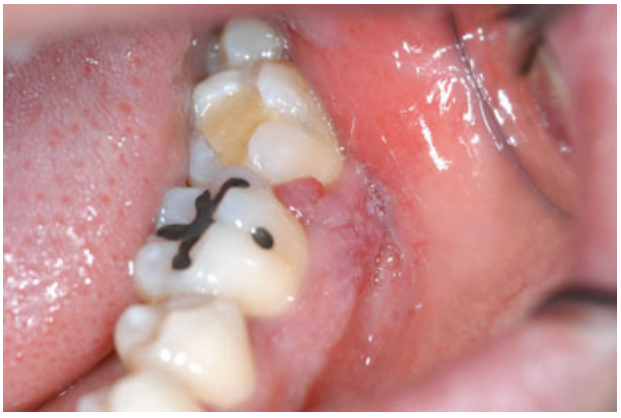

Figure 3 Gingival overgrowth interproximal 18/19.

a total excision of the swelling was performed in local anaesthesia, at this time without the use of laser but including a deep scaling of the root surfaces 13/14 under direct view. Suture removal was performed 7 days after

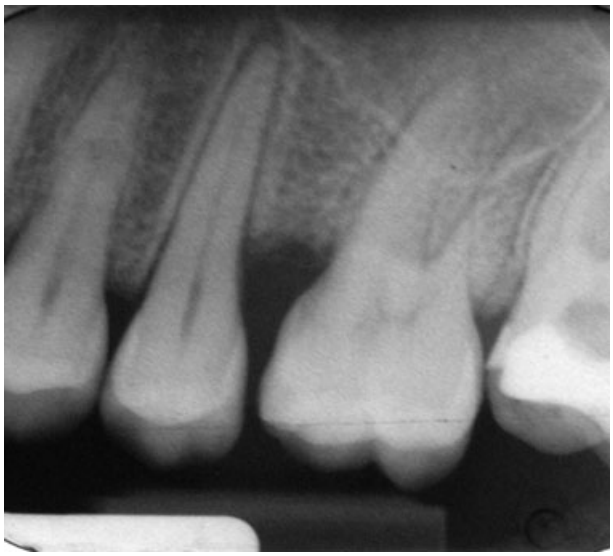

Figure 4 Conventional X-ray film 13/14 with horizontal interproximal 13/14 loss of bone.

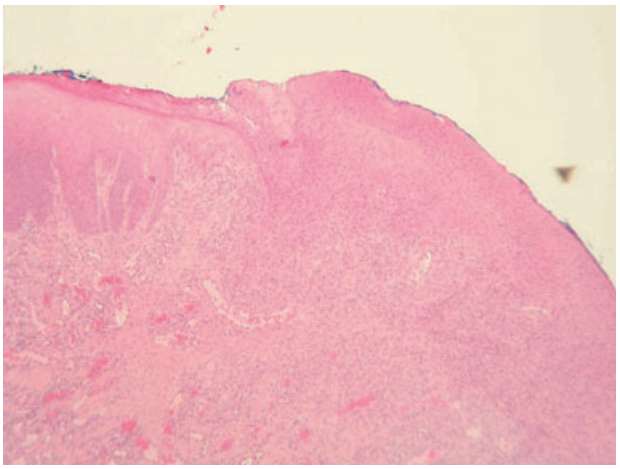

Figure 5 Excised hyperplasia with pyogenic granuloma with focal ulcerated inflammatory tissue region 13/14 (haematoxylin and eosin, H & E).

surgery and the excised tissue was analysed in the Department of Pathology. Histopathological examination revealed an inflammatory, lymphoplasma cell-rich tissue with fibrosis (Fig. 6) histologically and clinically corresponding to an epulis fibromatosa.

Again after sutural removal 7 days post-surgery the patient did not represent for further treatment and planning.

Comments

Though definition of the KTS is clear, the identification can cause difficulties because of the extreme degree of variability in localization and occurrence of the symptoms (2, 4). Auluck et al. (1) suggested that 5% of cases may involve the head and neck region (5). In our case, a naeuvus flammeus was affecting the left midfacial region and the left acres. The unilateral

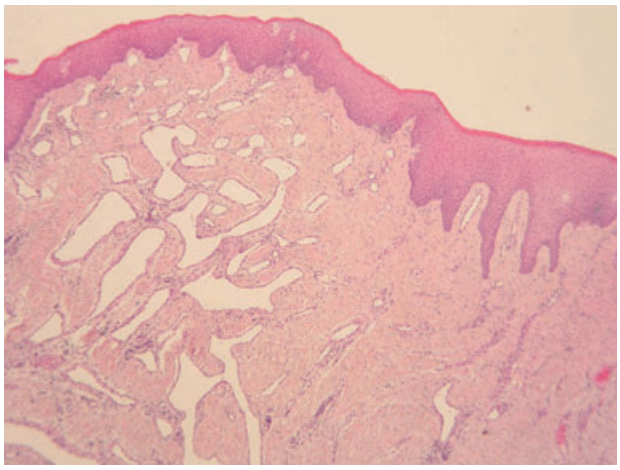

Figure 6 Recidive with inflammatory, lymphoplasma cell-rich tissue with fibrosation histologically and clinically corresponding to an epulis fibromatosa.

limitation of naevus is typical but bilateral naevi have also been described earlier (2). Venous varicosities are rarely found in the orofacial region as gravity seems to facilitate the venous drainage from the head and neck region when compared with lower extremities (2). This explains the absence of venous varicosity in the oral region in our case.

Limitation to one side concerns hypertrophy of hard and soft tissue (1). The hyperplasia can be attributed to increased vascularity resulting from vascular malformations. This may explain the slight hyperplasia of the tissues affected with naevus flammeus in our case. All tissues in the oral cavity, such as the tongue, cheek and lips can be affected. In contrast to the study of Auluck et al. (1), it had been shown that hypertrophy of soft tissue may occur bilaterally as described by Bathi et al. (2). In our case we only found hyperplasia of the lips and gingiva as an oral feature, limited to the left side. Additional findings as early tooth eruption, jawbone asymmetry and differences in the size of teeth between affected and non-affected areas as described by Sciubba and Brown (5) were not evident in our case. The appearance of gingival hypertrophy in our case limited to the same side as the extraoral hypertrophy reveals the question whether this is significant or coincidental. As Bathi et al. (2) and Hallett et al. (3) presented three cases with KTS and gingival overgrowth in the affected area, we support that this appearance is significant.

The diagnosis epulis can only be verified by histological analysis of the excised tissue. Epulis is caused by mechanical or inflammatory stress especially on the upper two-thirds of the affected root surface. Additionally, a non-specified predisposition is discussed, one of which could be KTS. Noticeable with regard to this hypothesis beside the appearance of three case reports with gingival overgrowth and KTS is the unilateral occurrence of epulis, affecting the same side as the extraoral findings. This leads to the suggestion that the appearance of gingival hypertrophy in form of an epulis is significant. Therapy includes total excision plus deep scaling of the root surfaces of affected teeth. Recidives are often noticed. In this case the extraction of affected teeth would remain the only therapy. The appearance of an epulis fibromatosa as a recidive of the resected epulis granulomatosa is contributed to the scarring process. To prove our hypothesis, further reports are needed on this particular rare entity.

References

1. Auluck A, Suhas S, Pai KM. Klippel-Trenaunay syndrome. Case report. *Oral Dis* 2005; **11**: 255–8.
2. Bathi RJ, Agarwal N, Burde KN. Klippel-Trénaunay syndrome (angioosteohypertrophy syndrome): a report of 3 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2002; **93**: 276–80.
3. Hallett KB, Bankier A, Chow CW, Bateman J, Hall RK. Gingival fibromatosis and Klippel-Trenaunay-Weber syndrome. Case report. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1995; **79**: 578–82.
4. Defraia E, Baccetti T, Marinelli A, Tollaro I. Biometric and magnetic resonance imaging assessment of dentofacial abnormalities in a case of Klippel-Trénaunay-Weber syndrome. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2004; **97**: 127–32.
5. Sciubba JJ, Brown AM. Oral-facial manifestation of Klippel-Trenaunay-Weber syndrome. Report of two cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1977; **43**: 227–32.

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