

## Case Report

# Large frontal osseous hemangioma with dural sinus involvement in a patient with Klippel-Trenaunay syndrome: A rare case report

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## Abstract

**Background:** We present one of the first documented cases in the literature of an adult with Klippel-Trenaunay syndrome (KTS) with a large frontal osseous hemangioma.

**Case Description:** A 30-year-old male presented with a rapidly enlarging frontal skull lesion that had developed in only 3 months. Radiological investigation revealed a highly vascular lesion attached to the frontal bone. The lesion was surgically resected with the patient making complete recovery. Histopathology was consistent with an osseous hemangioma.

**Conclusion:** We report the clinical presentation and surgical management of a rare presentation of osseous hemangioma in a patient with KTS.

**Key Words:** Hemangioma, Klippel-Trenaunay syndrome, Neuropathology

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## INTRODUCTION

Klippel-Trenaunay syndrome (KTS) is a rare congenital disorder characterized by the classic triad of capillary malformation, venous malformation, and limb overgrowth.<sup>[6]</sup> The eponymous syndrome was first described by French physicians Maurice Klippel and Paul Trenaunay in the early 20<sup>th</sup> century.<sup>[1]</sup> A well described entity of KTS is the development of peripheral vascular hemangioma, however, hemangioma arising from the skull is rare and to our knowledge, not described in the literature prior. We present the unique case of a 30-year-old male who presented with a rapidly expansile hemangioma arising from the frontal bone of the skull. Clinical presentation and surgical resection are described.

## CASE REPORT

A 30-year-old male with a known history of KTS presented with a rapidly enlarging subcutaneous central frontal mass, measuring 41 × 47 × 54 mm

since its formation just 3 months earlier causing pain and an obvious cosmetic defect. The patient had no demonstrable neurological signs on examination with normal pre-operative laboratory studies, however, had previous episodes of significant hemorrhage arising from the lesion as a result of minor trauma.

Clinically, the patient was dysmorphic, most notably severe soft tissue overgrowth, bilateral lower limb lymphoedema, and cellulitis. Despite this, he had no

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history of other complications arising as a result of his KTS such as malignancy or peripheral hemangioma.

Radiological findings initially began with computed tomography angiogram, which revealed a destructive expansile arterially-enhancing lesion arising from the frontal bone. Heterogenous contrast enhancement on magnetic resonance imaging [Figure 1] further defined the lesional dural involvement and invasion into the superior sagittal sinus sparring cerebral parenchyma.

Gross total resection consisted of a T-shaped incision followed by bilateral frontal craniectomies [Figure 2]. The vascular lesion was identified arising from frontal bone and within the superior sagittal sinus. The lesion was resected en bloc from calvaria. A custom-made titanium implant was used for cranioplasty [Figure 3].

Postoperatively, the patient had a full recovery. Histopathological hematoxylin and eosin stains revealed the lesion extending through bone, with a small foci of extramedullary hemopoiesis, consistent with hemangioma. There has been no evidence of recurrence 6 months post resection.

## DISCUSSION

Since its description in the early 20<sup>th</sup> century, prevalence has remained uncommon.

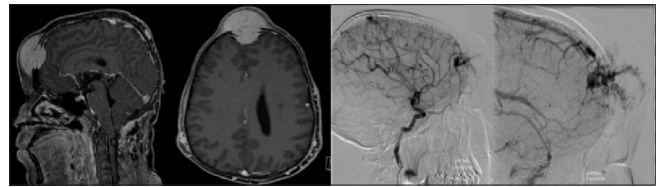
To our knowledge, vascular lesions arising from the skull in patients with KTS have not been previously described in the literature. Previous reports do highlight the increased prevalence and wide variability of malignancy in patients with KTS peripherally and in the spine.<sup>[5,7]</sup>

Hemangioma are benign vascular tumors, most commonly seen in pediatric age groups.<sup>[4]</sup> Rapid growth and sinus involvement both increase hemorrhagic risk.

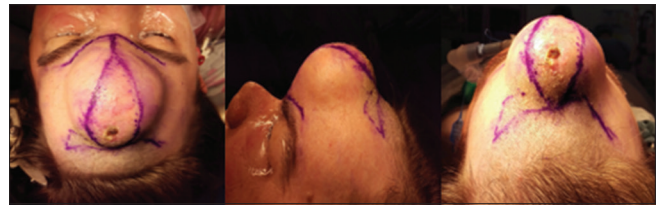
Intraoperatively, the tumor was seen to locally invade into calvaria with evidence of skull remodeling and destruction which is scarcely discussed as a characteristic of vascular hemangioma in past literature. Gross total resection is the gold standard of therapy and recommended by the authors. This minimizes recurrence and allows for tissue histopathological diagnosis in a syndrome known for predisposition to malignancy. The role for non-operative management in hemangioma including radiotherapy still exists.<sup>[2]</sup> The ambiguity of the lesion usually necessitates histopathological tissue diagnosis, with previous reports detailing the close resemblance to meningiomas and hemangiopericytoma.<sup>[3]</sup>

Postoperatively, the patient received regular follow-up with radiological screening for tumor recurrence.

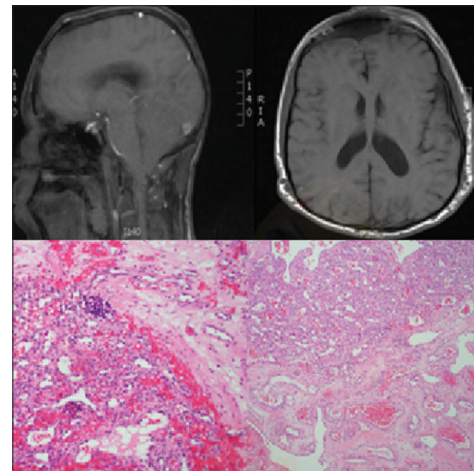
We, as the authors would like to highlight this unique case of an intracranial osseous hemangioma with frontal



**Figure 1:** T1 weighted post contrast magnetic resonance imaging depicting the destructive nature and invasion of the vascular lesion; although it is closely associated with dura and superior sagittal sinus, there is no evidence of parenchymal invasion. Digital Subtraction Cerebral Angiogram (right sided image) further characterizing the vascular lesion; arterial supply with fistulization from the ophthalmic artery and also some lesser arterial supply from the superficial temporal arteries bilaterally



**Figure 2:** These images detailing of the preparation and incisional marking. The lesion's sheer size can be appreciated



**Figure 3:** From Left to Right; Saggital and axial postoperative magnetic resonance imaging sections; H and E stains showing the mixed cavernous and capillary portions of the highly vascular lesion with invasion into bone

skull based origin in a patient with KTS, a rare syndrome with a predisposition to malignancy. There are associated dangers and complexities for the surgeon involved in resection of these uncommon lesions.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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