**Case Report**

# Nasopharyngeal angiofibroma in Klippel Trenaunay syndrome: a possible association

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## ABSRACT

Klippel-Trenaunay Syndrome (KTS) is a congenital vascular disorder consisting of vascular malformation and disturbed growth of bone or soft tissue. KTS is generally confined to the limbs but may involve the head and neck region. We present a case of 16- year-old adolescent male who presented with epistaxis and was diagnosed to have Juvenile nasopharyngeal angiofibroma. He also had features of KTS. (Rawal Med J 2011;36:66-68).

**Keywords:**

Klippel-Trenaunay Syndrome, angiofibroma, vascular malformations.

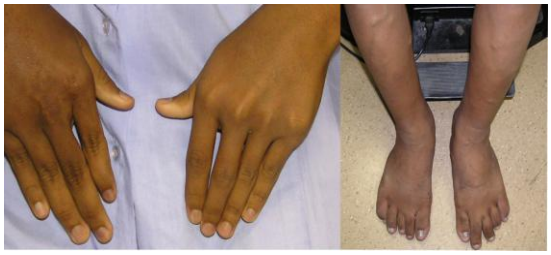
## INTRODUCTION

Klippel Trenauney syndrome (KTS) is uncommon and consists of bone/soft tissue hypertrophy, varicosities/venous malformation or capillary malformation and head and neck involvement ranges from localized craniofacial hemihypertrophy to associations with vascular anomalies or even neoplasm. Vascular anomalies of the head and neck region particularly vascular neoplasm in association with KTS are exceedingly rare. To our knowledge, KTS with juvenile nasopharyngeal angiofibroma has not been reported in literature.

## CASE PRESENTATION

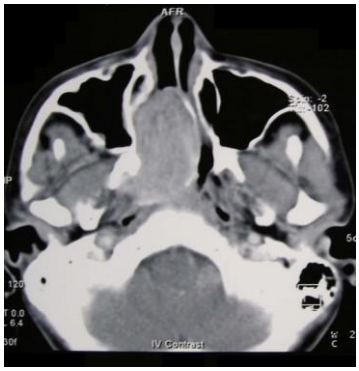
A 16-year-old adolescent male presented with intermittent right epistaxis with right nasal blockage for 5 years which had progressively worsened and required multiple blood transfusions. Furthermore, he had hematochezia for the past 3 months but no hematuria. His parents noticed his feet were bigger than other children since he was a toddler which then progressively grew disproportionately bigger with age.

**Fig 1. Limb hypertrophy in both hands and feet. Note also the venous varicosities of both lower limbs.**



His mother has a strawberry hemangioma on her left cheek. General examination revealed varicose veins of both lower limbs with bilateral limb hypertrophy (Fig 1). Capillary malformation on both soles of foot and both thumbs was present. Examination of the nose revealed a firm mass occluding the right nostril. CT scan of the paranasal sinuses revealed a soft tissue mass in right posterior choana measuring 6.0 x 2.5 cm extending into the nasopharynx with no bony destruction (Fig 2).

**Fig 2. An axial view of the CT scan showing a soft tissue mass occupying the right posterior choana extending into the nasopaharynx.**



Cerebral angiogram with embolization of the feeding vessel i.e. right internal maxillary artery was done one day prior to surgery. Preoperative hemoglobin level and coagulation profiles were normal. The tumour was excised en bloc via extended lateral rhinotomy approach. Intraoperative findings revealed a mass arising from the sphenopalatine foramen. There was no evidence of intranasal arteriovenous malformation (AVM). Histopathology confirmed the diagnosis of juvenile nasopharyngeal angiofibroma (JNA). The patient recovered well with no post operative complications. In view of haematochezia, colonoscopy and abdominal angiogram was done but did not reveal pelvic hemangimas or AVM. Subsequent follow up did not show any recurrence of angiofibroma.

## DISCUSSION

KTS was originally described as unilateral limb hypertrophy, venous varicosity and hemangioma without AVM. Mulliken and Glowacki classified vascular anomalies into 1) vascular tumours characterized by proliferating cells and slow involution (hemangioma) and 2) vascular malformation which are dysplastic vessels without spontaneous involution and is now widely accepted.[1] The nomenclature 'hemangioma' originally described in KTS is now recognized to be a vascular malformation most commonly seen as port wine stain. The vascular malformations are most famously confined to skin, but may involve muscle, bone or even visceral organ such as spleen, pleura, bladder or colon.

In the ensuing years a wide spectrum of this disease ranging from limb hypertrophy to hypotrophy, vascular anomalies which included lymphatic malformation and even clinically non significant AVMs have been reported.[2] The most recent proposed diagnostic criteria requires 2 major features (at least 1 from group a and should always include either capillary or venous malformation and at least 1 from group b).[2] Group (a) consist of vascular malformation which are 1) capillary malformation, 2) venous malformation, 3) small AVM

and 4) lymphatic malformations. Group (b) comprises disturbed growth of either 1) bone or 2) soft tissue in length or girth. KTS is associated with additional findings that are non obligatory but still in concert with this syndrome most commonly hip dysplasia and syndactyly. Other associated features that have been reported are hip dislocation, talipes, skin atrophy and hyperhydrosis.

Location of disturbed growth typically occurs in the extremities which may be present in half the body or single limb. However, there have been few case reports on KTS with craniofacial disturbed growth and a 12-year-old boy with right hand hypertrophy associated with bilateral lower limb lymphedema who also manifested right facial hypertrophy has been reported.[3] Another two cases described hemi-facial hyperplasia of soft and hard tissues with vascular malformation in the oral cavity with disturbed growth confined to the craniofacial region.[4,5] KTS has also been associated with overgrowth of bony cochlea in the middle ear causing mixed hearing loss in a patient with unilateral upper limb and facial hypertrophy.[6] In the head and neck region, intracranial vascular malformations leading to repetitive cerebral bleeding in the form of aneurysm and AVM have been reported.[7]

There was also one reported case of sensorineural hearing loss due to venous malformation in both cerebral hemispheres.[8] On the other hand, vascular tumours are exceedingly rare in KTS. There was an isolated case of oral vascular proliferative patch reported in a patient with KTS but it had no documentation of histopathological confirmation.[9] To our knowledge, only one case of vascular tumour in the head and neck region have been associated with KTS.[10] Nevertheless, KTS have been associated with other neoplasm in the head and neck region such as orbital rhabdomyosarcoma, meningioma, astrocytma and cerebral hemangiopericytoma.[10] However, to our knowledge this is the first report of association of KTS with nasopharyngeal angiofibroma.

Juvenile nasopharyngeal angiofibroma (JNA) is a vascular tumour effecting 0.05% of all head and neck neoplasms. It occurs almost exclusively in adolescent males and is rare after the age of 25 years old. Its etiology remains unclear but postulated to be due to hormonal influence and genetic defects. JNA have been postulated as a specific form of hemangioma, vasoproliferative malformation and even ectopic vascular tissue.[11] It has been shown that vascular architectural features of JNA in fact represent vascular malformation.[12] The presence of JNA in our case is in keeping with his age and gender. Nevertheless the presence of a vascular malformation in the nasal cavity in an essentially vascular syndrome raises the possibility of an association. In summary, KTS is a syndrome with a wide spectrum of manifestations and association. Our case demonstrates the possible vascular anomalies that may be associated with this syndrome.

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