

## Open Surgical Management of a Ruptured Intracranial Aneurysm In an Adult Patient with Klippel-Trenaunay-Weber Syndrome

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## Introduction

- Klippel-Trenaunay-Weber (KTW) Syndrome first described in 1900
- Cutaneous capillary malformations, venous malformations, asymmetric limb hypertrophy, arteriovenous fistula
- Etiology unknown, but mesodermal disturbance suspected
- Few cases of intracranial aneurysms previously reported in the literature

## Case Presentation

- 24 y/o male s/p fall
- Hunt-Hess 2 – Severe HA and Nuchal Rigidity
- Fisher 3 – Stellate SAH in basal cisterns w/o IVH
- PMHx – KTW, patient with asymmetric limb hypertrophy, cutaneous telangiectasia, and visceral AVM
- History otherwise non-contributory
- Work-up revealed left ophthalmic and anterior choroidal artery aneurysms

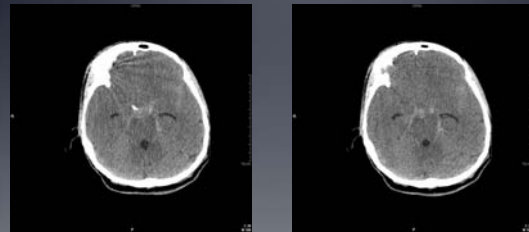
## Asymmetric Limb Hypertrophy



## Cutaneous Telangiectasia



## CT Imaging



## Angiogram



## Clinical Course

- PBD 1: Taken to angio, then OR for clipping
- Post-op R hemiparesis(3/5); MRI evidence of L AChorA ischemia attributed to intraoperative vasospasm
- PBD 6: R hemiplegia, taken to angio for IA verapamil for severe L MCA vasospasm
- PBD 16: Vasospasm resolved
- PBD 19: Patient develops GI bleed, GI consult obtained
- PBD 24: Patient discharged to inpatient rehab in good condition (4/5 right hemiparesis)

## Associated Complications

- Chronic Coagulopathy, particularly after trauma
- Increased DVT/PE risk
- Risk of GI bleeding from associated vascular malformations
- Distal thromboembolic events from intracranial aneurysm

## Prior Cases

- 7 prior reports of intracranial aneurysms in KTW
- 1/7 treated with clipping (8 y/o M)
- One prior case report of AVF clip ligation in a 12 week old with KTW

## Conclusions

- Open surgical management is a viable option for cerebral vascular lesions in KTW patients – our patient has made a full recovery
- KTW patients are at high risk for DVT/PE and should be treated as such
- Understanding of disorders such as KTW may shed light on pathogenesis of cerebrovascular lesions

## References

1. al-Salman MM. Klippel-Trenaunay syndrome: clinical features, complications, and management. Surg Today 1997;27:735-740.
2. Star A, Fuller CE, Landas SK. Intracranial aneurysms in klippel-trenaunay/weber syndromes: case report. Neurosurgery 2010;66:E1027-1028; discussion E1028.

Thank You

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