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 telangectasia, and visceral AVM
- History otherwise non-contributory
- Work-up revealed left ophthalmic and anterior choroidal artery aneurysms

Asymmetric Limb Hypertrophy

Cutaneous Telangectasia

CT Imaging
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

Thank You