

Open surgical management of a ruptured intracranial aneurysm in Klippel-Trenaunay-Weber (KTW) syndrome

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SUMMARY

A 24-year-old man with a history of Klippel-Trenaunay-Weber syndrome presented with severe headache and neck pain. Work-up revealed subarachnoid hemorrhage and evidence of multiple intracranial aneurysms. The patient was treated with open surgical clipping of his ruptured aneurysm and is currently doing well.

BACKGROUND

Klippel-Trenaunay-Weber (KTW) syndrome is characterised by asymmetric limb hypertrophy, cutaneous haemangiomas and varicosities. There have been few reports of KTW patients harbouring intracranial aneurysms. There have been only two prior cases reported of open surgical management of intracranial vascular lesions in KTW patients.^{1 2} This is the first known report of successful open clipping of a ruptured aneurysm in an adult patient with KTW.

CASE PRESENTATION

A 24-year-old man with a history of KTW syndrome presented with severe headache and neck pain after falling out of bed. He had a brief loss of consciousness at that time. He was then brought to the emergency room where he showed nuchal rigidity and no neurological deficit.

INVESTIGATIONS

Initially a CT scan revealed a subarachnoid haemorrhage. This later prompted a cerebral angiogram

which revealed multiple intracranial aneurysms including a 4 mm left ophthalmic artery aneurysm, a 5 mm left anterior choroidal artery aneurysm, a blister aneurysm of the left supraclinoid internal carotid artery and a right posterior communicating artery blister aneurysm (figure 1).

TREATMENT

He was taken to the operating room for surgical clipping of the left anterior choroidal and ophthalmic artery aneurysms and wrapping of the blister aneurysms. Intraoperatively, a several millimetre segment of the supraclinoid internal carotid artery (ICA) appeared irregular and diseased.

OUTCOME AND FOLLOW-UP

His postoperative course was complicated by right hemiparesis due to a left anterior choroidal artery infarct likely due to vasospasm as well as an upper gastrointestinal bleed. He ultimately recovered, returning to his neurological baseline.

DISCUSSION

KTW syndrome is characterised by asymmetric limb hypertrophy, cutaneous haemangiomas and varicosities. There have been few reports of KTW patients harbouring intracranial aneurysms. There have been only two prior cases reported of open surgical management of intracranial vascular lesions in KTW patients (figure 2).^{1 2}



Figure 1 Cerebral angiogram, left lateral internal carotid artery injection.



Figure 2 Extremity soft tissue and bone hypertrophy.

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Learning points

- ▶ Klippel-Trenaunay-Weber syndrome is a relatively rare condition characterised by asymmetric limb hypertrophy, cutaneous haemangiomas and varicosities.
- ▶ Rarely, this patient population can harbour intracranial aneurysms.
- ▶ Open surgical treatment is a viable option for acute rupture of intracranial aneurysms in this patient population.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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