Lethal subarachnoid haemorrhage in a patient with rete mirabile network of the posterior circulation with associated aneurysm

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SUMMARY

Rete mirabile (RM), an arterial network normally existing in some vertebrate animals interconnecting the extracranial and intracranial arterial circulation, can rarely be found in humans whether asymptomatic or presenting with cerebral ischaemia or haemorrhage. Encompassing diverse angiographic characteristics and similarities with other arterial malformations, proper diagnosis and differential diagnosis is challenging. We hereby describe an unusual RM case variant, presenting to us with lethal subarachnoid haemorrhage owing to a ruptured small aneurysm associated with the RM network. Angiography disclosed an absent P1 segment of the posterior cerebral artery (PCA) and an RM network anastomosing the basilar apex with the normal distal PCA. Brain death was confirmed on the fifth day after admission and attributed to the severity of subarachnoid haemorrhage (SAH). This is an exceedingly rare case representing an intradural-to-intradural RM anastomosis in a patient presenting with lethal SAH. RM epidemiology, pathophysiology, presentation, angiographic findings and prognosis are reviewed.

BACKGROUND

In animals, rete mirabile (RM) is a normal transdural arterial anastomosis interconnecting the internal carotid artery (ICA) with the external carotid artery. It is considered to serve thermoleguratory,[1] pressure absorption[2] or blood flow regulation purposes.[2] It is normally found in lower mammals, such as pigs, cows and sheep.[3]

When identified in humans, RM encompasses a congenital malformation involving a collateral, usually transdural, network between two arteries. The most commonly implicated intracranial vessels are the petrous or cavernous ICA,[4 5] receiving blood flow from extracranial arteries such as the maxillary artery[6 7] or the ascending pharyngeal artery.[7] The term carotid RM (CRM) has been used when ICA is implicated in the anastomosis. When the vertebral artery is also implicated, the term carotid and vertebral RM has been used.[3] An abundance of cases with bilateral RM has been reported.[3 8–10] RM of the posterior circulation is an even more rare variant, with only a few cases reported over the years.[3 11] Most of the RM vessels tend to occur extradurally providing a connection with a vessel across the dura.[4 12] Nevertheless, the term RM has been used quite liberally in the literature to describe an abnormal, usually reticular-like, artery-to-artery anastomosis, since there has been no strict

angiographic definition or criteria developed thus far. We identified, including our case, multiple cases of intradural-to-intradural anastomosis[3 8–11 13–17] and also a single extradural-to-extradural anastomosis case.[18] Finally, a single acquired CRM case has been reported.[19]

CASE PRESENTATION

We present a middle-aged patient who was admitted with 'worst headache of life'. Initial Glasgow Coma Scale (GCS) was 12 but the patient quickly deteriorated to GCS 4. An initial non-contrast CT scan of the head showed diffuse subarachnoid haemorrhage (SAH) with intraventricular extension, complicated by acute hydrocephalus (figure 1). SAH was assessed as grade 5 on Hunt and Hess Scale.

TREATMENT AND OUTCOME

The patient was intubated and a frontal external ventricular drain (EVD) was placed. Angiography revealed complete absence of the proximal part of the P1 segment of the right posterior cerebral artery (PCA). An RM network was identified interconnecting the basilar terminus with the existing P1 part of the PCA (figures 2–4). Posterior circulation angiography (figure 3) and microcatheter-based angiography with the microcatheter tip at the basilar terminus (figure 4) are available in 3D reconstruction. Bilateral thalamic and midbrain perforators emerged from this anastomosing small calibre RM network (figure 4). A small pseudo-aneurysm was detected at the dorsal part of the network, measuring $2.2 \times 1.9 \times 8$ mm. The patient was deemed non-amenable to endovascular or surgical treatment and hence was placed on aminocaproic acid. In another angiogram performed 2 days later, the aneurysm was found to be smaller, measuring 1.63×1.33 mm. An additional EVD system was placed on the second day of hospitalisation. Electroencephalogram showed diffuse low-voltage ($<20 \mu V$) waves with continuous diffuse severe slowing, and coma.

OUTCOME AND FOLLOW-UP

The patient had developed decerebrate rigidity by the second day of hospitalisation and progressively deteriorated developing intracranial hypertension, cerebral oedema and brain herniation. On the fifth day of hospitalisation, he met brain death criteria and was pronounced deceased.
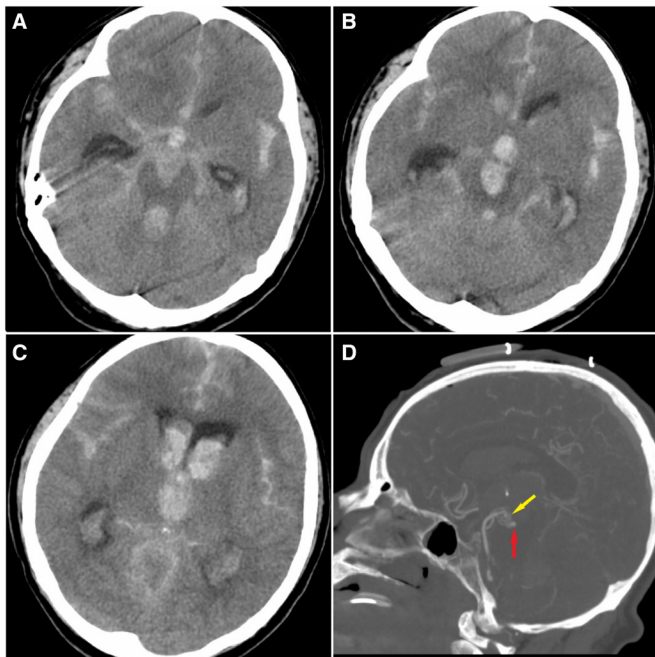
DISCUSSION

The first RM case was reported in 1966.[20] RM is usually associated with ICA[12] or vertebral[3]

Figure 1 (A) Non-enhanced CT scan revealing severe acute cisternal subarachnoid haemorrhage, especially in the interhemispheric and sylvian compartments, as well as acute brainstem intraparenchymal haemorrhage. (B,C) More rostral slices of the same CT scan demonstrating intraventricular haemorrhage in the third ventricle and frontal horn of the lateral ventricles. (D) CT angiography demonstrating an abnormal vascular network at the tip of the basilar artery and possibly a small aneurysm.
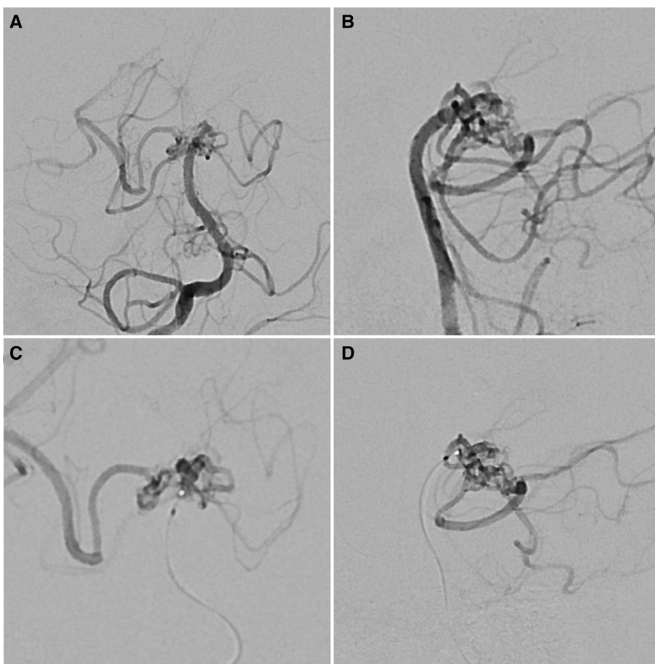

Figure 2 Posterior circulation angiography demonstrating the RM network in anteroposterior (A) and lateral (B) views. Microcatheter angiography of the RM network, with the tip of the microcatheter being at the basilar artery tip. Anteroposterior (C) and lateral (D) views showing no definite normal connection between the basilar artery and the right posterior cerebral artery. RM, rete mirabile.
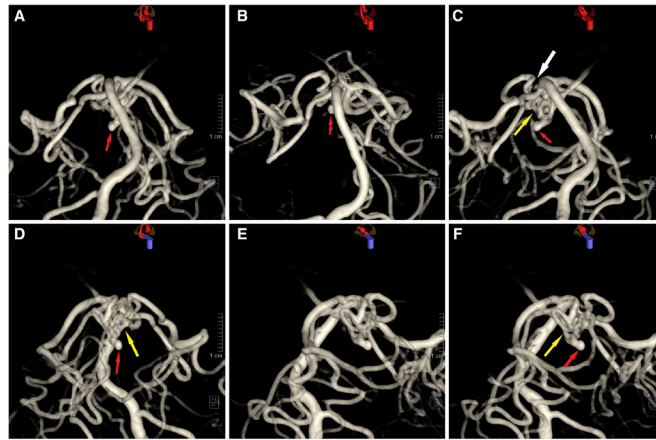
Figure 3 3D reconstruction of the posterior circulation in anteroposterior (A,B,D) and lateral (C,E,F) views. Left-sided posterior cerebral artery (PCA) has an embryologic origin. On the right side, there is no existing normal connection between the PCA and the basilar tip (C, white arrow). Instead, there is a rete mirabile (RM) network (C,D,F, yellow arrow) interconnecting these vessels. The right PCA demonstrated delayed filling. The aneurysm (red arrow) is associated with the RM network and protrudes caudally and anteriorly.

hypoplasia/aplasia/segmental occlusion, with the RM being responsible for supplying the areas lacking normal supply by the absent vessels.[6] Hypothetically hypoplasia/aplasia promotes primordial vessels to create new 'salvage' anastomosing networks to irrigate ischaemic regions, during embryogenesis.[9] The PCA's proximal segment derives from ICA's posterior division, while the cortical branches stem from the part of the anterior choroidal artery lying next to lateral geniculate body.[21] A defect during embryogenesis could explain the RM formation in our case. These events are likely promoted by ischaemia due to the occlusion.[11] RM appearance has been described in prior ipsilateral ICA ligation.[19]

The angiographic prevalence of RM was reported as 0.01% within a single institution, until 1997.[22] However, prevalence has been reported higher in the Asian population,[9] being as high as 0.67% in angiographies.[23] However, true prevalence and what
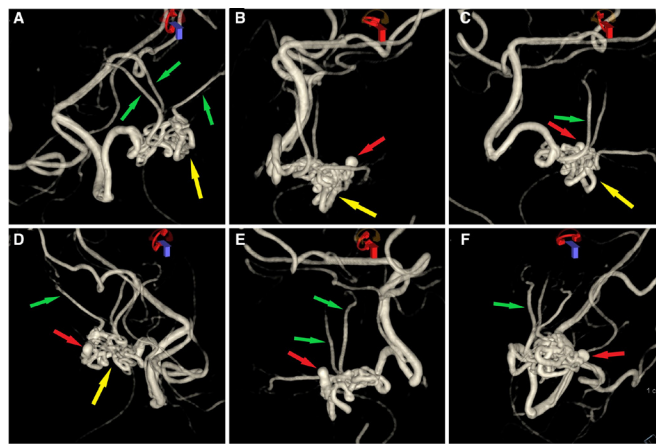
Figure 4 3D reconstruction of microcatheter-based angiography demonstrating the RM network. Findings are the same as in figure 3. Normal thalamoperforators (A,C–F) are seen emerging form the RM network (green arrows). Aneurysm (red arrow) and its relationship with the RM network (yellow arrow) is demonstrated (B–F). RM, rete mirabile.

percentage becomes symptomatic are currently unknown. RM has been identified in patients with haemorrhagic or ischaemic manifestations.[24 25] In a 2015 review, 34% of the identified RM cases (12/35) presented with SAH,[24] the first of which was reported in 1967.[26] SAH in patients with RM is thought to be caused not only by ruptured aneurysms[3 7 24] but also by haemodynamic stress[22] or rupture of the anastomosing vessels,[12] since an aneurysm was identified only in a small subset of patients. Haemodynamic changes related to the abnormal anastomotic pattern may lead to mechanical stress on vessels, thus promoting aneurysm formation.[3] In a case described by Henkes *et al*, RM was associated with fibromuscular dysplasia, as shown by biopsy, and the presence of a ruptured aneurysm.[27]

Diverse RM angiographic features have been reported as follows: ICA hypoplasia/agenesis,[12] transdural anastomotic arterial plexus between cavernous ICA and maxillary artery (usually small calibre[27]), ophthalmic-to-maxillary artery anastomosis, ophthalmic artery and RM plexus supplying a normal supraclinoid ICA, bilateral RM and RM network resembling intradural moyamoya disease. Another study classified RM as moyamoya type or nidus type, depending on whether the vessels resembled stages 3 and 4 of moyamoya disease or the nidus of an arteriovenous malformation, respectively.[22] Moyamoya type anastomotic vessels (ethmoidal pattern) have been found in a case of ICA occlusion.[5] RM may at times be confused with an arteriovenous malformation or fistula.[11] RM constitutes an artery-to-artery anastomosis. Selective angiography could exclude these diagnoses by showing the absence of a draining vein.[11] Differential diagnosis from moyamoya is challenging. The authors considered this case to be an RM one and not moyamoya disease, since the abnormal network seemed to be reconstituting the gap between the PCA and basilar artery while it did not resemble the small and thin 'puff of smoke'-like vessels of moyamoya.

Prognosis of patients with RM is usually good, based on the available reports from the literature. Notably, there has been another lethal SAH case in a patient with RM,[19] like ours. Lethality in our case is related to the severe SAH presentation (Hunt and Hess grade 5). Different management approaches have been endeavoured in patients with RM who presented with SAH, including aneurysm clipping.[24]

Learning points

- ► Rete mirabile is an artery-to-artery anastomosis, found normally in vertebrate animals, but rarely found in humans, associated with arterial agenesis/hypoplasia/severe stenosis.
- ► Most common pattern is an extradural-to-intradural anastomosis between the internal carotid artery and the anterior part of circle of Willis.
- ► Diverse angiographic features can be demonstrated including resemblance to arteriovenous malformations (AVMs) or moyamoya vessels.
- ► Angiographic prevalence is 0.01%, most of the patients are asymptomatic, but haemorrhagic or ischaemic phenomena might ensue.
- ► Experience is required for proper diagnosis and differential diagnosis and the therapeutic approach should be tailored on a case-by-case basis depending on the type of presentation.

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