CASE REPORT

Rete mirabile associated with pial arteriovenous fistula: imaging features with literature review

Prabath Kumar Mondel, Rashmi Saraf, Uday S Limaye

Department of Interventional Neuroradiology, Seth G. S. Medical College and K. E. M. Hospital, Mumbai, Maharashtra, India

Correspondence to
Professor Uday S Limaye,
Department of Interventional Neuroradiology, Seth G. S. Medical College and K. E. M. Hospital, Acharya Donde Marg, Parel, Mumbai 400012, Maharashtra, India; uslkem@gmail.com

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SUMMARY

A rete mirabile is a vascular network of intercommunicating small arteries or arterioles that replace the definitive adult artery supplying the brain. It supplies the brain in lower mammals but is not seen in normal human embryological development. A 26-year-old man presented with worsening tinnitus that was interfering with his sleep. On CT and digital subtraction angiography he was found to have a temporal lobe pial arteriovenous fistula with bilateral carotid and vertebral rete mirabile. The patient was offered open surgical and endovascular treatment options for pial arteriovenous fistula but he refused both and opted for conservative medical management. At 6-month follow-up he continued to have pulsatile tinnitus but was otherwise neurologically normal. We present the first described association of rete mirabile with pial arteriovenous fistula and discuss its clinical presentation and imaging features, with a review of the literature for bilateral carotid and vertebral rete mirabile.

BACKGROUND

Rete mirabile (RM) has been associated with aneurysm, arteriovenous malformation, carotid cavernous fistula, vein of Galen aneurysmal malformation, pseudoxanthoma elasticum and Dieulafoy's lesion.^{1 2} This is the first documented association of RM with pial arteriovenous fistula (PAVF).

CASE PRESENTATION

A 26-year-old man presented with recent worsening of a right-sided pulsatile tinnitus that interfered with his sleep. He had a pulsatile swelling behind his right mandibular angle since childhood, a right-sided tinnitus for 1 year and left-sided tinnitus for

2 years. On examination, there was no focal neurological deficit and the patient was otherwise normal.

INVESTIGATIONS

A Doppler ultrasound of the mandibular swelling revealed multiple tortuous arteries. The patient was further evaluated with a CT angiogram (CTA). On CTA, both carotid canals were hypoplastic (figure 1A). A left temporal PAVF with bilateral carotid and vertebral RM was seen (figure 1B, C). The patient was further evaluated with a digital subtraction angiogram. RM was seen in distal V2 and V3 segments of both the vertebral arteries (VAs) (figure 2A, B). The vertebral RM was supplied by enlarged deep cervical, ascending cervical, occipital and muscular branches of the VAs (figure 2C, D). The temporal branches of the posterior cerebral and middle cerebral arteries supplied a multi-feeder, single-hole, posterior temporal PAVF. A variceal temporal cortical vein drained the fistula into the left transverse-sigmoid junction (figure 2E, F). Both the internal carotid arteries (ICAs) were hypoplastic (figure 3A, B). The ICAs showed RM in their cavernous segments (figure 3C, D). Bilateral supraclinoid ICAs were patent with bilateral dilated ophthalmic arteries (figure 3E, F). Enlarged branches were seen arising from the internal maxillary and ascending pharyngeal arteries to supply the carotid RM. There was extensive perinidal angiopathy adjacent to the fistula (figure 3).

TREATMENT

The patient was offered endovascular and open surgical options for further management. Transarterial

Figure 1 CT angiogram. (A) Axial section shows bilateral hypoplastic carotid canals (3.3 and 2.8 mm in diameter on left and right sides respectively) (arrows). (B) Axial maximum intensity projection (MIP) image shows a single-hole pial arteriovenous fistula (PAVF) supplied by temporal branches of the posterior cerebral artery (curved arrow) and middle cerebral artery. Note multiple phleboliths within the venous varix. (C) Sagittal MIP image shows rete mirabile involving the internal carotid and vertebral arteries (asterisks). The single-hole multiple feeder PAVF is supplied by the hypertrophied posterior temporal branch of the posterior cerebral artery (curved arrow) and temporal branches of the middle cerebral artery (thick arrow). A patent and dilated left ophthalmic artery is also seen (arrowhead).

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Figure 2 Digital subtraction angiogram of posterior circulation. (A, B) Left and right vertebral angiograms show rete mirabile (asterisks) involving distal V2 and V3 segments of the vertebral arteries (arrows). (C, D) Left and right subclavian angiograms show rete mirabile (asterisks) of the vertebral arteries. Enlarged ascending cervical (thick arrow), deep cervical (arrowheads) and vertebral arteries (thin arrow) supply the rete mirabile. (E, F) Left vertebral angiogram frontal and lateral views show a single-hole pial arteriovenous fistula supplied by an enlarged posterior temporal branch of the posterior cerebral artery (curved arrows).

embolization was not feasible through the RM. The options of open surgical disconnection and transvenous embolization were discussed.

OUTCOME AND FOLLOW-UP

The patient decided against flow disconnection and opted for conservative management. At 6-month follow-up he continued to have pulsatile tinnitus but is otherwise normal.

DISCUSSION

Congenital absence (hypoplasia, aplasia and agenesis) of ICA: definition and classification

Congenital absence of ICA is a rare anomaly with a prevalence of less than 0.01%. According to Lie *et al*, congenital absence of ICA can be classified into ICA agenesis, aplasia and hypoplasia. ICA agenesis is characterized by the absence of the carotid canal and ICA. ICA aplasia is a vestigial ICA remnant, frequently a fibrous cord, with a patent carotid canal. In ICA hypoplasia, the ICA lumen is patent but is uniformly small, as seen in this case.³

Embryology: RM and PAVF

Both the ICA and VA are derived from fusion of seven embryological segments. Lasjaunias *et al* have discussed their embryological origin in further detail. ICA is fully formed at 6 weeks of gestation. In case of any disruption prior to 6 weeks, circulation is re-established through embryonic collaterals (eg, primitive maxillary artery), whereas in disruptions beyond 6 weeks, collateralization is through the circle of Willis. RM is a normal vascular network seen in lower mammals that supplies the brain. However, RM does not occur at any stage of human embryogenesis. It has been postulated that RM seen in humans is a subencephalic transdural phenomenon seen in cases with embryonic ICA (cervical, petrous and cavernous) regression. Furthermore, it is argued that embryonic ICA regression should stimulate collateralization by embryonic arteries. However, if there is regression of embryonic collaterals, transdural RM

develops to provide the blood supply to the brain. Thus, RM is considered a secondary response to ICA regression and not an embryonic variation in humans. It is also known that specific regions of the arterial system have segmental vulnerabilities that can affect anatomically unrelated arteries simultaneously during development. In the present case, the presence of bilateral and symmetrical RM involving transdural ICA and VA with hypoplastic carotid canals suggests a congenital etiology caused by a trigger that could also have simultaneously caused a PAVF, as seen in the present case. This is supported by the multiple reported instances of RM associated with vascular malformations such as vein of Galen malformation, aneurysm, arteriovenous malformation, and carotid cavernous fistula.^{4 5}

RM: diagnostic criteria

In 1844, Quain *et al* were the first to describe RM in humans. RM is a fine network of interconnecting arteries or arterioles located in the cavernous sinus and suboccipital region.⁴ Araki *et al* proposed angiographic criteria to diagnose carotid RM:¹

1. Hypoplastic ICA
2. Arterial network supplied by internal maxillary artery and petrous segment of ICA
3. Dilated ophthalmic artery
4. Patent supraclinoid ICA supplied by maxillary and ophthalmic arteries
5. Bilateral lesions
6. No intradural arterial collaterals reminiscent of a *moya* *moya*-like angiographic picture.

The present case satisfied all these criteria.

PAVFs: definition, clinical features, classification, disease associations, prognosis and management

PAVFs are abnormal direct artery-to-vein communications in the absence of an intervening nidus or arterial plexus. They are superficial and cortical, located in the subpial space and drained by enlarged veins.^{6 7} PAVFs account for 1–5% of all intracranial vascular malformations.⁸ Neonates commonly present with

Figure 3 Digital subtraction angiogram of anterior circulation. (A, B) Left and right internal carotid angiograms show hypoplastic internal carotid arteries (ICAs). The ICAs are hypoplastic from their origin with contrast dilution from anastomosing rete mirabile at the cavernous and horizontal petrous segments, respectively. (C, D) Left and right selective external carotid angiograms show rete mirabile (asterisks) supplied by branches from the internal maxillary artery. There is faint opacification of the pial arteriovenous fistula (PAVF) through the posterior communicating artery and temporal branches of the middle cerebral artery (thick arrows). The supraclinoid internal carotid artery is patent and normal on both sides (*dotted arrow*). (E, F) Left and right common carotid angiograms show dilated ophthalmic artery (arrowheads) and patent supraclinoid ICAs (*dotted arrows*).

Table 1 Summary of salient features of previously reported patients with bilateral carotid and vertebrobasilar rete mirabile^{1 2 5 10 12–18}

Authors and year	Age/sex	Clinical features	Segments involved	Disease associations
Jones <i>et al</i> , 1970	43/M	Recurrent loss of consciousness	Cavernous ICA Distal basilar artery	None
Koo <i>et al</i> , 1972	20/M	Carotid cavernous fistula	Cavernous ICA V2 segment of VAs	Pseudoxanthoma elasticum, splenic arterial rete mirabile
Itoyama <i>et al</i> , 1993	40/M	Ischemic stroke	Cavernous ICA V2 segment of VAs	None
Hyogo <i>et al</i> , 1996	37/F	Hemorrhagic stroke (subarachnoid hemorrhage)	Cavernous ICA V3–V4 segments of VAs	None
Mahadevan <i>et al</i> , 2004	29/F	Hemorrhagic stroke (recurrent blindness and subarachnoid hemorrhage)	Cavernous ICA V4 segment of VAs	None
Kim <i>et al</i> , 2006	38/M	Incidentally detected	Cavernous ICA Vertebrobasilar junction	None
Li <i>et al</i> , 2006	57/F	Hemorrhagic stroke (intracerebral and subarachnoid hemorrhage)	Cavernous ICA V3–V4 segment of VAs	None
Sahin <i>et al</i> , 2010	32/F	Incidentally detected	Cavernous ICA Proximal basilar artery	Maxillary sinus anaplastic meningioma
Castro <i>et al</i> , 2010	34/F	Intermittent altered sensorium, visual changes and cognitive deficit	Cavernous ICA Distal basilar artery	None
Lee and Cha, 2011	28/M	Incidentally detected	Cavernous ICA V3 segment of VAs	None
Lu <i>et al</i> , 2014	18/F	Recurrent headaches	Cavernous ICA V3–V4 segment of VAs	None
Mondel <i>et al</i> , 2014	30/F	Recurrent speech slurring, proptosis, vomiting, loss of bladder and bowel control	Cavernous ICA V2–V3 segment of VAs	Vein of Galen aneurysmal malformation
Present case	26/M	Recurrent headaches and pulsatile tinnitus	Cavernous ICA V2–V3 segment of VAs	Pial arteriovenous fistula

ICA, internal carotid artery; VAs, vertebral arteries.

congestive cardiac failure, children <15 years with seizures and children >15 years with hemorrhage.⁷

PAVFs are classified into single-hole or multi-hole fistulas.⁶ They have been associated with hereditary hemorrhagic telangiectasia, Klippel–Trenaunay–Weber syndrome, Ehlers–Danlos syndrome, neurofibromatosis type 1, encephalocraniocutaneous lipomatosis and aortic coarctation.⁹ The prognosis of untreated PAVFs is poor with a mortality rate as high as 63%.⁸

The management of PAVFs involves flow disconnection. Flow disconnection can be achieved surgically or by endovascular techniques. Surgical flow disconnection is by clipping or cauterization whereas endovascular techniques involve embolization with liquids such as n-butyl cyanoacrylate or Onyx (Covidien, Mansfield, Massachusetts, USA) or coils via transarterial or transvenous routes. Adjuvant radiotherapy has been used to treat angiogenesis following venous pouch thrombosis.⁶ Surgical disconnection is associated with a high risk of morbidity when the fistula is located deep or in an eloquent location. Transarterial embolization may be unsuccessful in tortuous and multiple feeding artery PAVFs.⁷

Clinical and anatomical considerations in RM with PAVF

A review of all PubMed indexed literature for ‘bilateral carotid and vertebral rete mirabile’ showed the commonest clinical presentation as an ischemic or hemorrhagic stroke.^{1 2 10} None of the previously reported patients had tinnitus at presentation (table 1). Vascular causes of pulsatile tinnitus include arterial lesions like carotid atherosclerosis, arteriovenous fistulas and malformations, aberrant arterial anatomy and hypertension whereas venous lesions include benign intracranial hypertension, jugular bulb anomalies and hydrocephalus.¹¹ This patient’s bin-aural pulsatile tinnitus is related to RM. The presence of RM excludes the transarterial route of embolization. The patient could be treated by open surgical or endovascular transvenous flow disconnection. Surgical flow disconnection in the presence of RM could potentially cause excessive hemorrhage with an increased risk of cerebral infarction. Transvenous embolization would be associated with a higher risk of morbidity and mortality compared with transarterial techniques.

Contributors USL and RS managed the case. PKM identified, drafted and revised the paper. PKM and USL revised the draft paper. USL is the guarantor of the study.

Competing interests None declared.

Patient consent Obtained.

Ethics approval Local Ethics Committee.

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