CASE REPORT

Bilateral carotid and vertebral rete mirabile with vein of Galen aneurysmal malformation: an unreported association

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SUMMARY

Rete mirabile is a fine meshwork of anastomosing vessels that replace the parent artery. A 30-year-old woman complained of slurring of speech, right eye proptosis, recurrent vomiting, and loss of bladder and bowel control, followed by drowsiness lasting 30–40 min, for the past 6 months. On cross sectional imaging and angiography, the patient was found to have a vein of Galen aneurysmal malformation, with bilateral carotid and vertebral rete mirabile. The patient was offered both endovascular and open surgical options but she refused any form of surgical treatment and opted for conservative management. At the 6 month follow-up, she continued to have occasional episodes of headache and vomiting but was otherwise normal. We describe the clinical, cross sectional, and

angiographic features of this patient. A comparison with other patients with bilateral carotid and vertebral rete mirabile is also reported.

BACKGROUND

The term ‘rete mirabile’ (RM) is used for a meshwork of multiple fine arteries or arterioles freely intercommunicating that reconstitute the absent or hypoplastic segments of the parent artery. RM commonly involves the internal carotid artery (ICA) and the vertebral artery (VA), with the exception of a single case involving the posterior cerebral artery.^{1 2} RM has been described in association with aneurysms, arteriovenous malformations,

Figure 1 Non-enhanced CT and MRI scans of the brain. (A) Multiple, hyperdense, serpiginous structures in the interpeduncular, pontine, and cerebellopontine cisterns (arrows). (B) Enlarged and hyperdense median vein of prosencephalon (*) with intramural calcification with straight sinus stenosis (arrow). (C) Prominent chunky calcification is seen at the rostral and superior end of the median vein of prosencephalon (arrowhead). (D) Multiple serpiginous small flow voids are seen in the suprasellar, interpeduncular, pontine, and cerebellopontine cisterns (arrows). An enlarged basilar artery is seen posteriorly (arrowhead). (E) MRI section at the level of the body of the lateral ventricle shows an enlarged median vein of prosencephalon (*) with prominent feeding arteries opening directly into the median vein of prosencephalon with straight sinus stenosis (arrow). (F) The cavernous segments of both internal carotid arteries are replaced by groups of multiple small flow voids (arrows). Both vertebral arteries are enlarged with an enlarged basilar artery (arrowhead).

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Figure 2 Cerebral digital subtraction angiogram. (A) Right subclavian angiogram in frontal view shows rete mirabile involving the distal V2 and V3 segments of the right vertebral artery. Enlarged ascending cervical (arrow) and vertebral arteries (arrowhead) supply the rete mirabile. (B) Left subclavian angiogram in lateral view shows rete mirabile involving the distal V2 and V3 segments of the left vertebral artery. Enlarged ascending cervical (block arrow), artery of the cervical enlargement (arrows), and vertebral arteries (arrowhead) supply the rete mirabile. (C, D) Right and left internal carotid artery (ICA) angiograms in lateral view show rete mirabile replacing the absent ICAs in their Debrun C3 cavernous segment (arrowhead). Enlarged ophthalmic arteries (*) supply rete mirabile with enlarged and tortuous anterior choroidal arteries opening into the anterior fistulous (choroidal) communication of the vein of Galen aneurysmal malformation (VGAM) (arrow). (E) Right external carotid artery (ECA) angiogram in lateral view shows dural branches arising from the frontal and parietal middle meningeal branches of the ECAs that form a network in the inferior part of the median vein of prosencephalon prior to opacifying the VGAM (arrowhead). Flow related aneurysms are also seen along the course of the dural branches of the middle meningeal artery. (F) Left ICA angiogram in a frontal oblique view shows both the anterior (arrow) and inferior (choroidal subtype) (arrowhead) communications. (G) Right vertebral angiogram in Towne's view shows bilateral persistent marginal and occipital sinuses (arrowheads).

carotid-cavernous fistulas, pseudoxanthoma elasticum, and Dieulafoy's lesion.³ To our knowledge, this is the first report of bilateral carotid and vertebral RM in association with vein of Galen aneurysmal malformation (VGAM).

CASE PRESENTATION

A 30-year-old woman presented with intermittent slurring of speech, right eye proptosis, recurrent vomiting, loss of bladder and bowel control, followed by drowsiness lasting 30–40 min, for the past 6 months. On examination, there was mild right eye proptosis. Extraocular movement and visual acuity in both eyes were normal.

INVESTIGATIONS

On non-enhanced CT, there were multiple, hyperdense serpiginous structures in the cavernous sinus and subarachnoid cisterns (figure 1A). An enlarged median vein of prosencephalon (MVOP) was seen. The MVOP showed calcification (figure 1B, C). The bony carotid canal was uniformly narrow and hypoplastic on both sides and measured 2–3 mm. An MRI showed multiple flow voids in the cisterns (figure 1D). A few tortuous arteries converged on MVOP and were suggestive of VGAM (figure 1E). The right and left ICAs were of normal caliber until their petrous segment, with complete absence mid-way through the cavernous sinus where they were replaced by a meshwork of

arteries. The ICAs were reformed in their supraclinoid segment. The intradural VAs and basilar artery were enlarged (figure 1F).

On digital subtraction angiography, there was bilateral carotid and vertebral RM. Both VAs showed occlusion of the distal V2 and V3 segments. There was reformation of the V4 segment by RM. Ascending cervical, occipital, and vertebral branches supplied the vertebral RM with reinforcement on the left by the artery of cervical enlargement (figure 2A, B). Both ICAs were occluded at the Debrun C3 cavernous segment. The RM reformed the ophthalmic arteries (figure 2C, D). Branches from the internal maxillary, accessory meningeal, middle meningeal, and anterior tympanic arteries supplied the carotid RM (figure 2E). The dilated ophthalmic arteries reformed the supraclinoid ICAs. Both ICAs then supplied the middle cerebral arteries (figure 2C, D). Two distinct fistulous sites were present, anterior and inferior. Both the anterior and inferior communications were of the choroidal subtype. Multiple dural branches from the ICA and the external carotid artery supplied the malformation. The malformation drained through the straight sinus. There was bilateral moderate jugular bulb stenosis with venous drainage through the marginal and occipital sinuses (figure 2F, G). Bilateral enlarged and tortuous posterior-medial, posterior-lateral choroidal, and thalamoperforator arteries supplied the choroidal subtype of VGAM (figure 3A–C). There was unilateral Sylvian capture of the veins draining the normal cerebral parenchyma with outflow through the right

superior ophthalmic vein, bilateral inferior petrosal sinus, and pterygoid venous plexus (figure 3D). There was proptosis of the right eye due to venous congestion.

TREATMENT

The patient was offered endovascular and open surgical options for further management. The fragile and tortuous RM excluded a transarterial route of embolization. The option of transvenous or transthoracic embolization and open surgery with a higher risk of morbidity and mortality was discussed with the patient and her relatives.

OUTCOME AND FOLLOW-UP

The patient decided against any procedure and opted for medical management. At the 6 month follow-up, she continued to have occasional headaches and vomiting but was otherwise normal.

DISCUSSION

Segmental embryological origin of ICA and VA

The ICA is a composite of seven distinct embryological segments. According to Lasjaunias *et al*,⁴ secondary regression or absence of segment 5 of the ICA would result in RM formation. The VA is derived embryologically by fusion of six consecutive intersegmental anastomoses between the cervical intersegmental arteries.⁵

Rete mirabile

In humans, RM is not seen at any stage of embryonic development. It is believed that an unidentified trigger causes secondary regression of both the ICA and VA that stimulates carotid and vertebral RM.⁵ Hence RM is formed in the late fetal or perinatal period when embryological precursors of the ICA, VA, and their anastomoses have regressed.⁵

Clinical features and review of literature for RM

A review of the literature for patients with bilateral carotid and vertebrobasilar RM showed a total of 10 patients.¹ Acute ischemic or hemorrhagic stroke was the commonest presentation (50%). The remaining patients were detected incidentally on imaging, except for a single case of carotid-cavernous fistula. Additionally, six cases of carotid RM were associated with pseudoxanthoma elasticum.⁶ In the present case, the patient presented with features of cerebral venous ischemia. This is the commonest clinical picture in patients with VGAM with late presentation and patent sinuses.⁷

Embryology of VGAM

VGAM constitute 30% of all pediatric vascular malformations. VGAM occurs between 6 and 11 weeks of gestation. Abnormal arteriovenous shunts between the choroidal arteries and the MVOP of Markowski leads to VGAM.⁸ The shunts prevent normal involution of the prosencephalic vein with ballooning of its anterior segment that forms the aneurysmal component of VGAM.⁹

Figure 3 Cerebral digital subtraction angiogram. (A–C) Right vertebral angiogram in left oblique, right oblique, and lateral views shows enlarged posterior choroidal (arrowheads) and thalamoperforator arteries (arrow) supplying the inferior fistulous network of the vein of Galen aneurysmal malformation. (D) Left internal carotid artery angiogram in lateral view shows Sylvian capture of venous drainage of the normal cerebral parenchyma (block arrow). There was subsequent venous outflow through the right superior ophthalmic vein, bilateral inferior petrosal sinus (arrowhead), and pterygoid venous plexus.

Cardiac and vascular anomalies associated with VGAM

VGAM has been associated with congenital cardiac diseases such as atrial septal defect, ventricular septal defect, atrioventricular canal defects, aortic coarctation, tetralogy of Fallot, transposition of the great arteries, and partial anomalous pulmonary venous drainage.¹⁰ Lasjaunias *et al* reported a Moya-Moya type of arterial pattern in three premature babies with VGAM. All died over a period of 48 h. Thus the presence of a Moya-Moya type of vasculature is a contraindication to treatment as it worsens the prognosis by the added insult of arterial deprivation to venous ischemia.⁷

Anatomical and clinical importance of RM with VGAM

In the present case of the choroidal subtype of VGAM, the presence of bilateral carotid and vertebral RM reduced cardiac preload and prevented development of congestive cardiac failure. However, this predisposed the patient to develop cere-

bral venous ischemia. Our patient had a better prognosis because RM does not necessarily decrease cerebral perfusion although it may decrease perfusion pressure.⁶ Thus the presence of RM in a patient with VGAM may not cause significant arterial ischemia and should not in itself be considered a contraindication to therapy.

The presence of this rare association excluded transarterial embolization and required consideration of transvenous or transthoracic embolization or open surgery. Transvenous and transthoracic embolization are associated with a higher morbidity. Surgical management of VGAM is associated with uniformly poor results and mortality rates of 38–91%.⁷

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.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Bilateral carotid and vertebral rete mirabile in association with vein of Galen aneurysmal malformation is a previously unreported association.
- ▶ Both internal carotid arteries were occluded at the Debrun C3 cavernous segment and the vertebral arteries were occluded at the distal V2 and V3 segments with distal reconstitution.
- ▶ The presence of rete mirabile seen in association with vein of Galen aneurysmal malformation may not in itself constitute a contraindication to endovascular or open surgical management.
- ▶ The presence of carotid and vertebral rete mirabile with vein of Galen aneurysmal malformation excludes the firstline treatment option of transarterial embolization and requires consideration of transthoracic or transvenous embolization or open surgery for further management.

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