Unilateral Rete Mirabile as a Result of Segmental Agenesis of the Ascending Petrous Segment of the Internal Carotid Artery: Embryology, Differential Diagnosis and Clinical Implications

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Summary

We describe an extremely rare case of unilateral rete mirabile in a young woman and discuss its significance in embryological development as well as its radiological aspects, differential diagnosis and potential clinical implications.

Introduction

With the widespread use of noninvasive neurovascular imaging such as magnetic resonance (MR) and computed tomography (CT) angiography, anatomical variations in the cervico-cranial vasculature are encountered with increasing frequency. While rare, congenital collateral networks may be found during the course of the neurological workup and are vital to restore anterograde flow in the vascular territory of congenitally diseased, aplastic or hypoplastic vessels. Understanding the roles and significance of these vessels in embryological development is important. This paper describes an extremely rare case of unilateral rete mirabile and discusses its development as well as its potential clinical significance and differential diagnosis.

Case Report

The patient is a 21-year-old woman referred for evaluation of right facial paresthesias and dizziness. MR angiography (MRA) revealed segmental agenesis of the ascending petrous portion of the right ICA which was replaced by multiple small vessels reconstituting the right ICA in the cavernous segment (Figure 1). The MRA also showed a large clival branch from the ascending pharyngeal artery entering the skull base through the hypoglossal canal reconstituting the right cavernous ICA (Figure 1). CT scan demonstrated that the right carotid canal was dysplastic, likely representing an embryological/congenital abnormality (Figure 2). Cerebral angiography was then obtained for better characterization of the cranial arterial supply and revealed antegrade flow in the right ICA with focal absence of a portion of its ascending petrous segment and distal reconstitution at the level of the cavernous segment through collaterals from the internal maxillary artery and a large clival branch from the ascending pharyngeal artery that enters the skull base through the hypoglossal canal (as seen in the MRA) and anastomosis with the meningo-hypophyseal trunk (Figures 3 and 4). The pa-

Figure 1 MR angiography (MRA) of the head demonstrating segmental agenesis of the ascending petrous portion of the right ICA which is replaced by multiple small vessels reconstituting the right ICA in the cavernous segment. The MRA also shows a large clival branch from the ascending pharyngeal artery entering the skull base through the hypoglossal canal reconstituting the right cavernous ICA.

tient's young age, the lack of atherosclerotic plaques, her clinical presentation, and angiographic findings makes it more likely that the collateral network seen is a rete mirabile. As her symptoms spontaneously improved she did not require further medical or surgical treatment.

Discussion

Collateral networks of the arterial supply in humans are infrequent and may be seen in cases where there is aplasia, hypoplasia or narrowing of the native vessels. One of these rare networks includes the carotid rete mirabile (RM), which is defined as involvement (segmental agenesis) of the epidural portion of the ICA with a compensatory arterial network most often supplied by ECA branches such as the

branches of the maxillary artery and ascending pharyngeal artery. While rare in humans, the RM is frequently found in other vertebrates. Their circle of Willis is often incomplete and is frequently supplied by the maxillary artery and rarely the vertebral arteries. During embryological development of the RM in certain mammals, the ICA likely undergoes secondary atrophy leading to reconstitution of distal ICA flow by embryological arteries including the ascending pharyngeal, basilar, trigeminal, primitive maxillary and internal maxillary arteries¹. As a result of the late regression of the ICA, the RM is a physiological network in lower mammals including pigs, cows, sheep and goats that serves to replace the arterial supply to the circle of Willis. This common adaptation is thought to facilitate heat exchange and regulate intracerebral blood pressure and flow². In addition, as formation of the RM is frequently bilateral it

Figure 2 CT head scan using a bone algorithm demonstrates a dysplastic underdeveloped aspect of the distal portion of the right carotid canal.

Figure 3 A) AP and B) lateral views of the right ICA angiogram reveal antegrade flow in the right ICA with focal absence of a portion of its ascending petrous segment which has been replaced by multiple small vessels reconstituting the right ICA in the cavernous segment.

Figure 4 Lateral view of the right ECA angiogram reveals distal reconstitution of the right ICA at the level of the cavernous segment through collaterals from the internal maxillary artery and from a large clival branch from the ascending pharyngeal artery that enters the skull base through the hypoglossal canal, and anastomosis with the meningohypophyseal trunk.

has been postulated that there is a unique signal trigger during embryological development that results in RM development³. Comprehending the formation of the RM in lower vertebrates helps to elucidate the development of the rare carotid RM and even more infrequent vertebral RM in humans.

The embryological development of the ICA in humans was described by Padget and helps to illustrate the unique anatomical relationships during the embryonic stages for the origins of the ICA, common carotid artery (CCA) and external carotid artery (ECA). Lie et al. helped to define six pathways that may develop in collateral circulation for those with internal carotid artery (ICA) occlusions. Of the six collateral circulations, type F best represents the RM and demonstrates transcranial anastomoses from the internal maxillary artery of the ECA to the distal ICA⁴. While it is more common to find case reports of the RM involving bilateral epidural internal carotid arteries, less common variations include unilateral vertebral rete mirabile (junction of extra and intradural portions) and ophthalmic rete mirabile. Associated symptoms and diagnoses of RM include loss of balance, dizziness, subarachnoid hemorrhage with associated aneurysms, carotid fistulas, pseudoxanthoma elasticum, and stroke⁵. In our case, it is unclear if the symptoms of right facial paresthe-

sias and dizziness were related to the RM. In addition, while there are case reports and reviews of bilateral carotid RM, unilateral involvement has not been as frequently reported.

In contrast to the rete mirabile that is thought to develop due to pathological changes during embryological development, the vasa vasorum is a collateral network that becomes prominent due to pathological changes later in life. The vasa vasorum provides microcirculation to the adventitia and outer third of the media of mid to large arteries including the carotid artery. While not seen on normal imaging of vessels, for those patients with noted atherosclerotic plaques, the vasa vasorum can become prominent through neovascularization and neoangiogenesis. In these cases, the vasa vasorum can be visualized on studies including carotid duplex ultrasound and may be misinterpreted as recanalization. Confirmation may only be obtained with selective angiography of the vessels.

For the carotid arteries, the vasa vasorum contributes to the formation of a dense network within the plaque and has been found to

originate from the superior thyroid and ascending pharyngeal arteries⁶. While it are also commonly seen in proximal vessels, the vasa vasorum is less likely to be found in distal intracranial arteries such as the middle and anterior cerebral arteries. The vasa vasorum can become significant in the rare cases where it provides anterograde flow in fully occluded right ICA due to surgery or atherosclerosis. Management of these patients should involve a modification of the risk factors associated with atherosclerosis as it has been shown that further development of the vasa vasorum is closely linked to growth in atherosclerotic plaques⁶.

Conclusion

We described a rare case of unilateral rete mirabile presenting in a 21-year-old woman. It is important to understand its significance in embryological development as well as its radiological aspects, differential diagnosis and future clinical implications.

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