

Rete Middle Cerebral Artery Anomaly Presenting with a Large Intracerebral Hemorrhage: A Case Report

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J Clin Interv Radiol ISVIR 2022;6:147–149.

Abstract

Rete MCA anomaly is extremely rare and has been seldomly discussed in literature. Embryologically MCA develops by fusion of multiple twigs which form a single channel. If there is failure to fuse a plexiform network of vessels persists known as rete MCA. This web like network of vessels is at high risk of haemorrhagic stroke and therefore for patients presenting incidentally correct diagnosis is paramount. Here we report a case of rete MCA anomaly in a 35-year-old gentle man who presented with a large haemorrhagic stroke in emergency. DSA showed unilateral involvement of proximal MCA, and otherwise normal intra-cranial vasculature. With this report we aim to correctly diagnose this rare condition and avoid misdiagnosis leading to unnecessary treatments.

Keywords

- ▶ rete middle cerebral artery
- ▶ rete MCA
- ▶ twig-like middle cerebral artery

Introduction

Middle cerebral artery (MCA) anomalies are extremely rare and include accessory, duplicated, and fenestrated MCA. Rete MCA has been seldomly reported in literature under various names like “rete”/ “unfused”/ “twig-like”/ “aplastic” MCA. It is a congenital anomaly resulting secondary to nonfusion of rete like network/twig-like arteries of primitive MCA.

We report a case of 35-year-old gentleman who presented to the emergency with a large intracranial bleeding. The purpose of this case report is to help recognize the clinical and angiographic features and correctly diagnose this entity as it is at a very high risk of hemorrhagic stroke and to avoid misdiagnosis leading to unnecessary treatments.

Case Report

A 35-year-old man was brought to our hospital with a sudden onset of headache and vomiting with left-sided weakness. On admission, he presented with impaired consciousness with a Glasgow Coma Scale (GCS) score of 6 (E2V1M3). His past medical history was not remarkable. He had no previous

history of hypertension. A head computed tomography (CT) showed large intra-parenchymal hematoma. CT angiography (CTA) demonstrated severe stenosis of the right M1 segment of MCA with normal caliber of distal MCA branches. A diagnosis of arterial dissection was made on CTA based on irregular narrowed appearance of M1 segment of MCA associated with large hematoma (→Fig. 1). Digital subtraction angiography was performed that revealed a plexiform

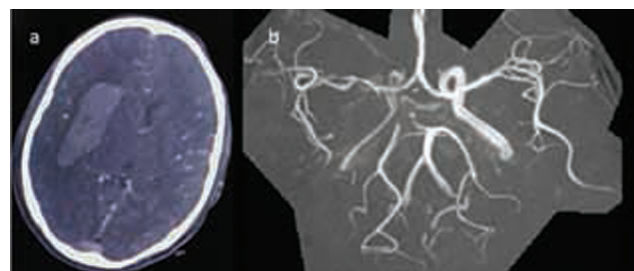


Fig. 1 (A) Noncontrast computed tomography (CT) scan showing a large intraparenchymal hematoma in right basal ganglia region (B) Reformatted CT angiography image showing irregular and

published online
November 12, 2021

DOI <https://doi.org/10.1055/s-0041-1732846>.
ISSN 2457-0214.

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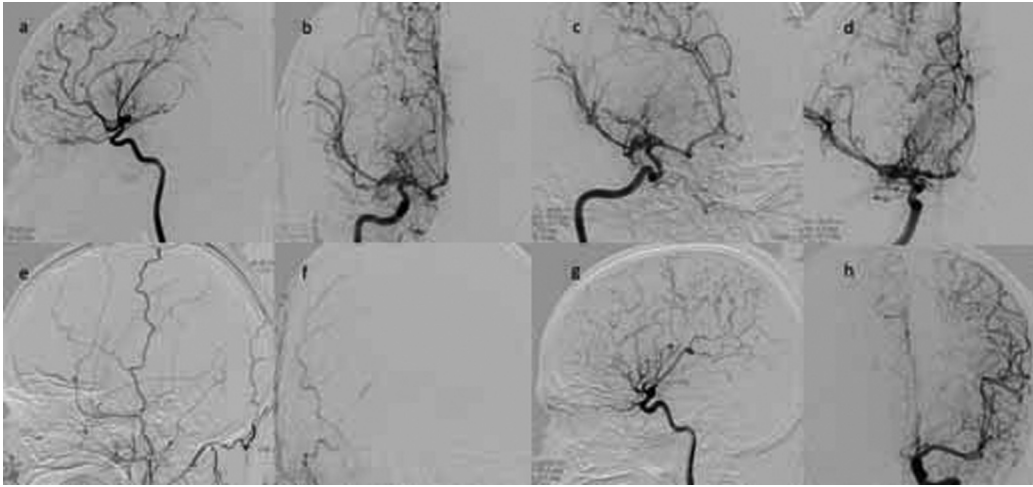


Fig. 2 (A–D) Right internal carotid artery angiogram demonstrates middle cerebral artery trunk replaced by a plexiform network of vessels. Lenticulostriate arteries are arising from the plexiform arterial network. (E–F) Right ECA angiogram demonstrates normal course of ECA branches. (G–H) Left ICA angiogram demonstrates normal course of ICA, ACA, MCA and their branches.

arterial network at the ipsilateral MCA trunk (►Fig. 2). Lenticulostriate arteries arose from the plexiform arterial network. His intracranial large vessels had no evidence of arteriosclerotic degeneration, and a transdural anastomosis was not detected.

Patient was intubated and managed conservatively in neurosurgical intensive care unit (ICU). However, on day 2, his conscious level decreased with a GCS score E1VtM2. A head CT revealed enlarged ventricles, implying hydrocephalus, without additional intracranial hemorrhage. On day 3 of admission, patient became unresponsive with nonrecordable vital signs. Despite all resuscitative efforts, he could not be revived.

Discussion

Anomalies of MCA are relatively rare and encompass rare instances of fenestrations, duplication arising from the internal carotid artery, an accessory MCA from the anterior cerebral artery and “rete”/ “twig-like”/ “unfused” MCA.

Rete MCA is extremely rare anomaly with a reported prevalence of 0.67% (13/1937) by angiographic examination by Cho et al.¹ A high prevalence has been reported in east Asian countries.

Embryologically during 7 to 12 mm fetal stage, MCA is first recognized as multiple twigs branching from the cranial division of internal carotid artery. Coalescence of these twigs into a single branch to form MCA proper occurs later at 16 to 18 mm stage and reaches adult configuration at 40 mm stage. Persistence of twig can explain the anomaly of rete MCA.²

Clinically patients present with both hemorrhagic and ischemic stroke as well as incidentally without any symptoms. Mechanism of hemorrhagic stroke includes either rupture of plexiform vessels or formation of flow-related aneurysm in fragile plexiform network of vessels or outside this network. Plexiform vessels are fragile in nature because of immature arterial wall that leads to formations of flow-

related aneurysms secondary to hemodynamic stress.³ Exact incidence of hemorrhagic stroke is not known; however, reported cases show ~50% incidence of hemorrhage in patients with rete MCA.^{2,3} Our case was not associated with an underlying aneurysm.

Digital subtraction angiography is the mainstay of diagnosis and shows features of plexiform network of vessels replacing the MCA trunk with the lenticulostriate arteries arising from this network. The distal branches of MCA beyond this rete are normal in caliber with antegrade flow.

Diagnosis of rete MCA by CTA/MRA is difficult and may be misdiagnosed as “moyamoya,” atherosclerotic stenooclusive disease, AVM, or as dissection as in this case.⁴ Moyamoya disease characteristically affects internal carotid artery terminus with possible involvement of proximal anterior cerebral artery, MCA, and posterior cerebral artery. There is formation of prominent basal collateral giving “puff of smoke” appearance as well as “transdural” collateral from external carotid artery. Typically, moyamoya disease is bilateral and symmetrical, whereas rete MCA is almost always unilateral and lacks transdural collaterals.⁵ Atherosclerotic stenooclusive disease is an acquired disease with atheromatous changes seen in multiple vessels.⁵ Post-varicella arteriopathy of childhood is another cause of unilateral MCA stenosis. However, most common age group involved in 1 to 2 years with brain imaging shows ischemic infarcts in lenticulostriate vessels territory.

Conclusion

Rete MCA is an extremely uncommon variation with lack of sufficient literature that needs to be recognized by medical practitioners to avoid misdiagnosis and unnecessary treatments. This uncommon variation is associated with flow-related aneurysm that is vulnerable to bleed. Patients who are incidentally discovered should be closely followed due to high risk of hemorrhagic stroke.

Funding
Self.

Conflict of Interest
None.

Acknowledgments
None.

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