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Case report / Приказ болесника

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Asymptomatic "twig-like" middle cerebral artery embryological anomaly

Асимптоматска ембриолошка аномалија средње церебралне артерије налик гранчици

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SUMMARY

Introduction Anomalies of middle cerebral artery (MCA) are very rare. "Rete MCA", "twig-like MCA" (T-MCA), "aplastic MCA", "unfused MCA" and others, are all synonims for unilateral embriological anomaly of M1 segment of MCA, where, due to unknown cause, fusion of primordial arteries of M1 segment did not happen. As a result, M1 segment of the affected side consists of mesh of small arteries from which arise normal perforators and cortical branches. In differential diagnosis should be considered moyamoya disease, moyamoya-like syndrome, atherosclerotic steno-occlusive disease, vasculitis and dissection of MCA.

Case outline We present 60 years old female patient with twig-like left MCA, accidentally diagnosed because persistent headaches six days prior to admission. Non-contrast computed tomography (NECT) head exam was without peculiarities. Computed tomography angiography (CTA) showed network of small vessels in place of left M1 segment, bridging internal carotid artery (ICA) terminus with branches of MCA bifurcation and giving rise to lenticulostriate arteries. 14 months later, on physical exam, she was in good general condition, without a neurological deficit, with occasional episodes of headache no stronger than 3–4/10 on the visual analogue scale (VAS).

Conclusion We report a patient with extremely rare variation of M1 segment of the left MCA, accidentally diagnosed because headache.

Keywords: twig-like MCA; middle cerebral artery; CT angiography

САЖЕТАК

Увод Аномалије средње мождане артерије (МЦА) су веома ретке. "Рете МЦА", "twиг-лике МЦА" (Т-МЦА), "апластична МЦА", "нефузио-нисана МЦА" и други називи, су синоними за једностарну ембриоланлну аномалију М1 сегмента МЦА, где, из непознатих разлога, није дошло до фузије примордијалних артерија М1 сегмента. Као резултат, М1 сегмент захваћене стране се састоји из мреже малих артерија из које полазе нормални перфоратори и кортикалне гране. Диференцијално дијагностички треба разматрати болест моуамоуа, синдром сличан моуамоуа-и,атеросклеротичну стено-оклузивну болест, васкулитис и дисекцију МЦА.

Приказ болесника Приказујемо жену стару 60 година са МЦА налик "гранчици", случајно откривеној због перзистентних главобоља шест дана пре пријема. Безконтрастна компјутеризована томографија (НЕЦТ) главе није показала неуобичајености. Ангиографија на компјутеризованој томографији приказала је мрежу малих крвних судова уместо левог М1 сегмента, која је повезивала терминални сегмент унутрашње каротидне артерије (ИЦА) са гранама МАЦ бифуркације и лентикулостријатним артеријама. Четрнаест месеци касније, на контролном физикалном прегледу, пацијенткиња је била доброг општег стања, без неуролошког дефицита, са повременим епизодама главобоље не јачим од 3-4/10 на визуо-аналогној скали (ВАС).

Закључак Презентујемо пацијента са екстремно ретком варијацијом левог М1 сегмента МЦА случајно откривеном због упорних главобоља.

Кључне речи: Т-МЦА; средња мождана артерија; ЦТ ангиографија

INTRODUCTION

Anomalies of middle cerebral artery (MCA) are very rare. They are less commonly seen than those of other major intracranial arteries [1, 2, 3]. Typically, three MCA anomalies (variations) are described: duplication (D-MCA), fenestration (F-MCA), and the presence of an accessory branch (A-MCA) [4]. "Rete MCA", "twig-like MCA" (T-MCA), "aplastic MCA", "unfused MCA" and others, are all synonyms for unilateral embryological anomaly of M1 segment of MCA, where, due to unknown cause, fusion of primordial arteries of M1

segment did not happen [1, 2, 3, 5, 6, 7]. As a result, M1 segment of the affected side consists of mesh of small arteries from which arise normal perforators and cortical branches [7]. Interestingly, the variations or possible anomalies in morphology of terminal branches of the internal carotid artery, like fenestration of the anterior cerebral artery (ACA) and hypoplastic ACA, have been also described in healthy non-human primates, as well as the left/right asymmetry in morphology of the MCA [8].

CASE REPORT

A 60-year-old female patient was admitted to the Emergency Center, University Clinical Center of Serbia, due to persistent headaches six days prior to admission. Non-contrast computed tomography (NECT) head exam was without peculiarities. Computed tomography angiography (CTA) showed network of small vessels in place of left M1 segment, bridging internal carotid artery (ICA) terminus with branches of MCA bifurcation and giving rise to lenticulostriate arteries. Left MCA M2 branches, although somewhat "paler", appeared to be normally filled with contrast agent. Deep middle cerebral vein had anomalous drainage into left superior petrosal sinus. (Figure 1 – MIP and Figure 2 – VR)

Since this was an accidental finding, patient was discharged home with only symptomatic therapy for headache (paracetamol / acetaminophen). Also, antiplatelet therapy in the form of acetylsalicylic acid (ASA) was prescribed. Fourteen months later, on physical exam, she was in good general condition, without a neurological deficit, with occasional episodes of headache no stronger than 3–4/10 on the visual analogue scale (VAS).

Informed consent was obtained from the patient for this publication.

DISCUSION

The middle cerebral artery is the largest and most complex artery supplying the brain, vascularizing the largest territory of neocortex [9, 10]. MCA develops after anterior cerebral artery (ACA), when fetal plexiform network of multiple small arteries fuse and regress in order to form perforating branches of M1 segment and main trunk of MCA (M1 segment). Disruption of this process, by still unknown cause, leads to MCA developmental anomalies [7]. Fukuyama reported one case of Ap/T-MCA which is associated with RNF213 mutations, which was previously believed to be associated exclusively with moyamoya [11]. In "T-MCA", this plexiform network persists unilaterally in place of M1 segment, while cortical and perforating branches, although filled with contrast agent with discrete delay, appear to be normal [6, 7]. Of all the MCA anomalies, T-MCA is the least commonly seen. Reports range from 0,1% to 4 % prevalence, while Viso stated prevalence of 0,088% in their cohort which included over 10,000 patients [3].

Possibility of hypoperfusion and, eventually, ischemic events has been described [6]. Uchiyama reported intracerebral hemorrhage in patient two years after TIA and diagnosed T-MCA as culprit [12]. Also, there is increased risk of aneurysm formation, due to hemodynamic stress and network vessels fragile histological architecture [5], which can lead to rupture and hemorrhage [3, 6, 13]. Sakai reported rupture of a "de novo" formed aneurysm arising from the twig-like network (TLN) of an anomalous collateral artery associated with aplastic or twig-like middle cerebral artery (Ap/T-MCA) in a patient who had ruptured aneurysm on A1 segment four years earlier [14].

In differential diagnosis should be considered moyamoya disease, moyamoya-like syndrome, atherosclerotic steno-occlusive disease, vasculitis and dissection of MCA [1, 3, 15].

Therapy options may vary depending on patient symptoms and angiographic findings, but no universal treatment has been established to this day [5]. If T-MCA is asymptomatic, coincidental finding, patient should to be counseled and warned about nature of the anomaly. Vessels in the mesh are functional and also fragile, so no intervention should be performed unless necessary [1]. It is still unclear if microsurgical superficial temporal artery bypass is beneficial in cases of recurrent ischemic events. In their case report, Matsunaga stated that postoperative magnetic resonance angiography showed a decrease of blood flow in aberrant network indicating that this approach may improve perfusion of affected MCA territory and lower hemodynamic stress in aberrant network [6]. On the other side, Matsuo stated that there is no evidence that revascularization is effective approach in preventing stroke on affected side. Further studies on this anomaly are necessary to understand its nature and provide adequate therapy [16]. Aneurysms in anomalous MCA network have high risk of rupture and should be treated surgically or by endovascular embolization. Open surgery is more commonly used due to higher risk of endovascular approach through these fragile vessels [7].

Although uncommon, clinicians should recognize this vascular entity in order to avoid misdiagnosis and unnecessary treatment which can lead to catastrophic adverse events, especially in the era where mechanical thrombectomies became every days practice, and this entity could make confusion because of simulation of thromboembolic event. Less experienced neuroradiologist could easily overlook subtle vessel network between ICA and distal part of MCA.

In our opinion, the patient should be on lifelong preventive antiplatelet therapy (ASA), in order to avoid consequences of steno-occlusive and thromboembolic events. A follow-up physical examination, by a neurologist/neurosurgeon, should be performed every two years, while neuroradiological examination is reserved only for patients with hemorrhagic or ischemic symptoms.

Conflict of interest: None declared.

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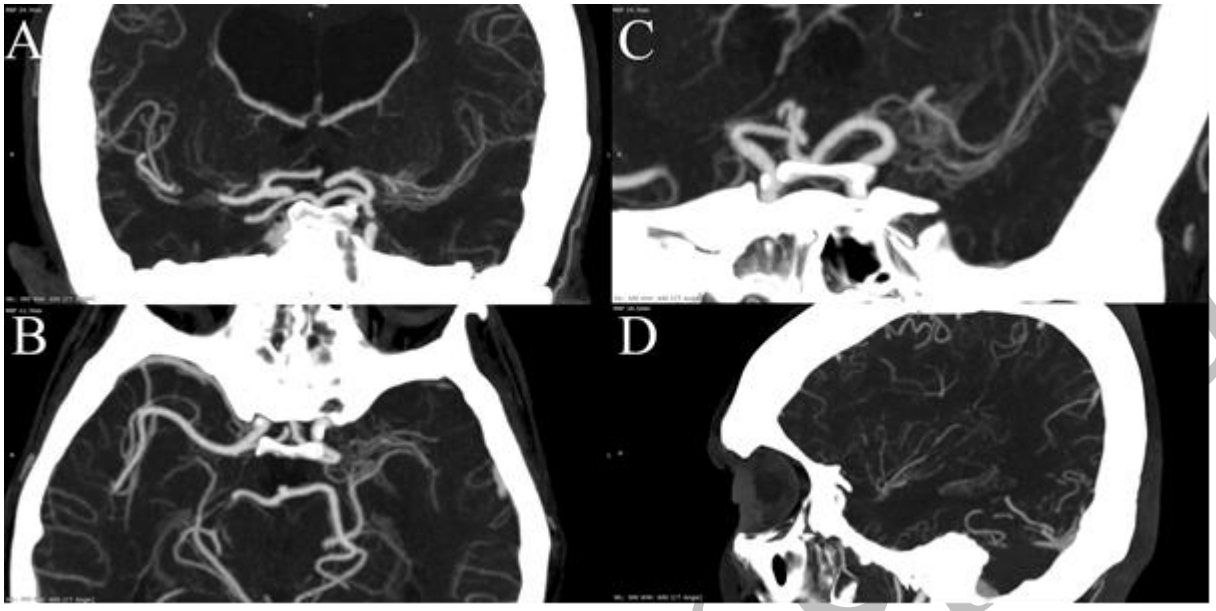

Figure 1. Computed tomography angiography maximum intensity projection reconstructions, A – coronal, B – axial, and C – oblique projections show multiple "twig-like" arteries arising from terminal part of left internal carotid artery, forming network in place where M1 should be, D – sagittal projection demonstrates normal arborization of the left middle cerebral artery with branches that are slightly less filled with contrast agent

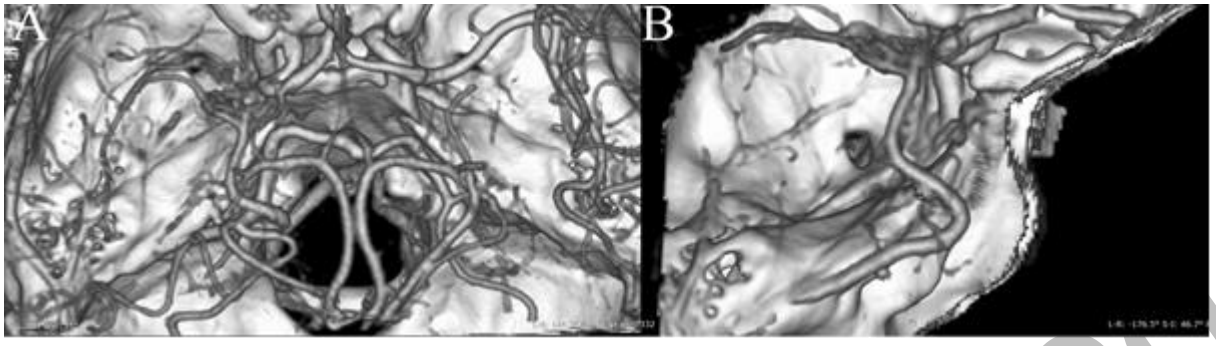

Figure 2. Computed tomography angiography VR; A and B show deep middle cerebral veins anomalous drainage into superior petrosal sinus

