Basilar Artery Dissection in an Adult Patient Presenting with De Novo Moyamoya Disease

Sir,

Spontaneous intracranial artery dissection (ICAD) is an important cause of ischemic stroke in young adults, more frequently affecting the Asian population. The exact pathophysiology is unknown, and various underlying conditions, connective tissue disorders, and moyamoya disease (MMD) were proposed as predisposing factors.[1,2] Acute basilar artery dissection (BAD) is an uncommon event, with significant morbidity and mortality rates.[3] Moyamoya disease is a rare and progressive steno-occlusive disorder of supraclinoid segments of the internal carotid artery (ICA), resulting in the formation of a bilateral network of collaterals at the base of the brain.[4] MMD most frequently manifests with transitory ischemic attacks and ischemic or hemorrhagic strokes within anterior circulation.[4,5] We present an extremely rare occurrence of basilar artery dissection with de novo diagnosis of moyamoya vasculopathy, which according to our knowledge is only the second reported case so far.

A 38-year-old Caucasian female was transferred to the Stroke unit of the Emergency center, with right-sided hemiparesis and speech disturbance as symptoms of the stroke. Her past medical history was unexceptional apart from subtotal hysterectomy and left-sided ovariectomy 4 years ago, due to a benign tumor. The family history was unremarkable, other than the presence of hypertension and onset of a stroke at a younger age. On the admission her general physical examination showed no abnormalities and the initial laboratory investigations, lumbar puncture, chest X-ray, and echocardiogram were normal. Neurological examination revealed bilateral miosis, divergent strabismus, and bilateral cortical blindness with transitory psychomotor agitation while the National Institutes of Health Stroke Scale (NIHSS) was 15. The first noncontrast head computed tomography (CT) scan showed extensive regions of cortical-subcortical hypodensity, mostly in the vascular territory of posterior circulation, which was confirmed by magnetic resonance imaging (MRI) performed on the second day of hospitalization [Figure 1a]. In addition, magnetic resonance angiography (MRA) displayed marginal irregularities of Willis circle arteries [Figure 1b].

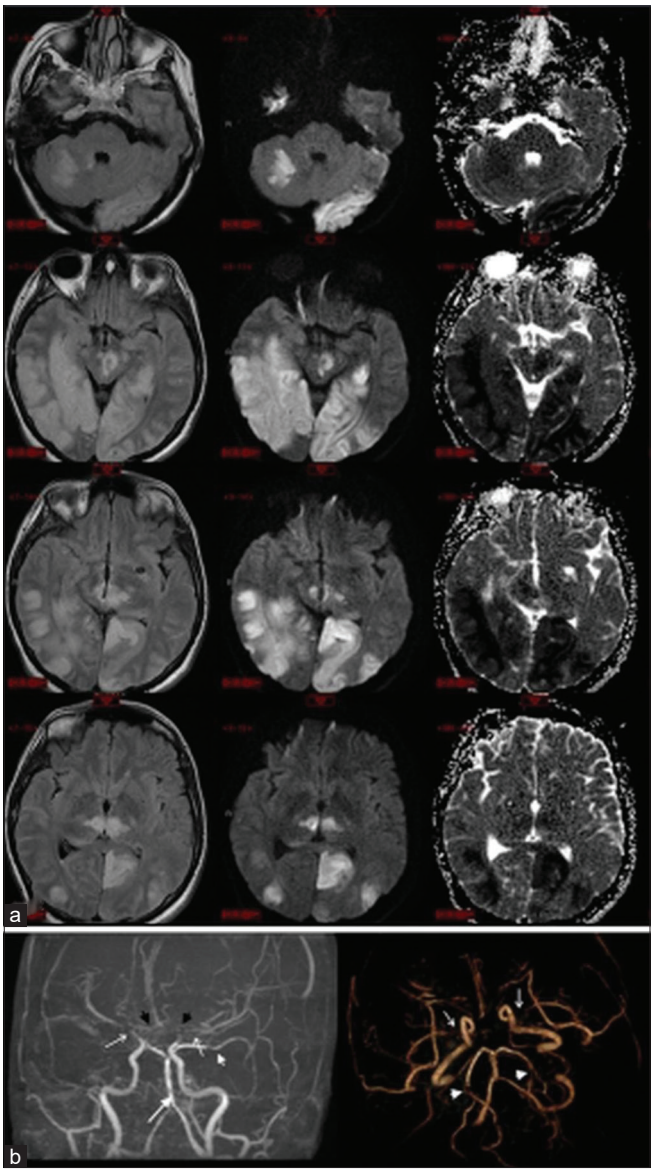

Figure 1: (a) MRI [(FLAIR), (DWI), (ADC) tomograms]: areas of ischemia in right cerebellum (superior cerebellar artery – SCA), mesencephalon, medial basal ganglia bilaterally (basilar artery (BA) perforators and P1 segment of posterior cerebral artery – PCA), and in occipital and basal temporal areas bilaterally (PCA). (b) MRA and volume rendering technique (VRT): bilateral occlusions of terminal ICA segments, M1 of middle cerebral artery – MCA (white arrows) and A1 of anterior cerebral artery – ACA (black arrowheads), with stenosis of BA (big white arrow) and both PCA (white arrowheads)

On the fourth day of hospitalization, digital subtraction angiography (DSA) revealed dissection of BA with the flap extension to the left posterior cerebral artery (PCA) [Figure 2a and b], as well as the network of collateral vessels via the middle meningeal artery (MMA) on the left [Figure 2c and d] and ophthalmic artery on the right [Figure 2e].

Defining the etiology of moyamoya vasculopathy was one of our priorities. Considering her previous medical history, CT and MRI of the abdomen were done, which revealed de novo benign tumor of right ovarium with normal levels of serum tumor markers. Laboratory studies showed dyslipidemia with strong atherogenic potential (Lp (a) > 0.9 g/L) and insulin resistance. Extended coagulopathy workup was within normal limits as well as the patient's thyroid function tests. We completed genetic studies for the determination of increased risk factors for thromboembolism and hypercoagulability:

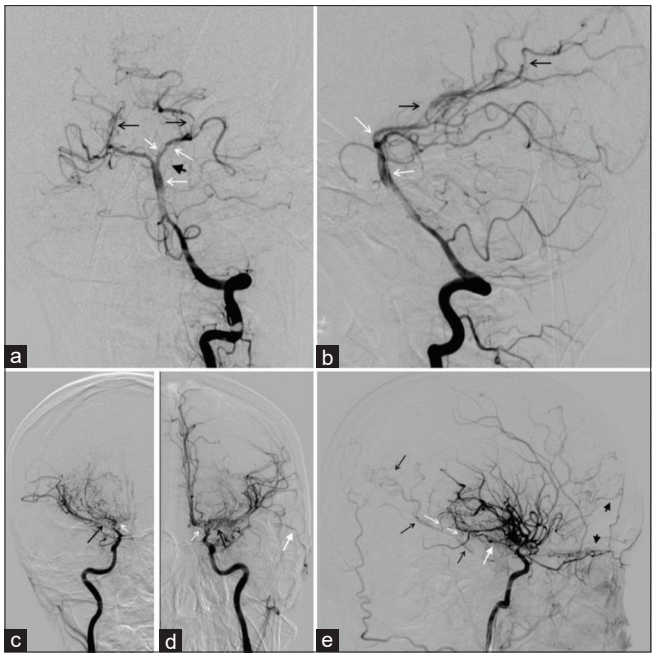

Figure 2: (a, b) DSA of left vertebral artery (VA): dissection of BA with flap extension to left PCA (white arrows), stenosis of both PCA (black arrows), and absence of left SCA (black arrowhead). (c, d) DSA of both ICA: occlusions of terminal ICA, M1 of MCA (black arrows), A1 of ACA (white arrows), with collaterals via left MMA (big white arrow). (e) DSA of right ICA: right anterior choroidal artery (big white arrow) branching (smaller white arrows) towards posterior choroidal artery fills the right PCA (black arrows); collaterals via right ophthalmic artery (black arrowheads)

Factor V Leiden G1691A, Factor II GA20210, and MTHFR A1298C and C677T mutations, which were all normal. Genetic examination for mutated variant c.14429G >A in the RNF213 gene was negative. Two weeks after the admission, control neuroimaging study confirmed previously noted dissection of BA on DSA [Supplementary Figure 1], and we registered a typical "brush sign" of MMD, which correlated with impaired hemodynamics of the affected brain areas [Supplementary Figure 2].

Since the appearance of new neurological symptoms several days after the admission (left third nerve palsy), clopidogrel has been added to the previously established therapy with aspirin and a hypolipemic agent while revascularization surgery was not considered. At the discharge, our patient remained on dual antiplatelet therapy, her cortical blindness persisted with moderate right-side hemiparesis, with NIHSS 9, and modified Rankin scale (mRS) 3. She was transferred to a regional center for the physical rehabilitation program, and after 3 months, the secondary prevention treatment for ischemic strokes was reduced to aspirin only. At the last follow-up visit after a year and a half, her clinical status remained unchanged.

According to literature, the incidence of BAD is approximately 4.5%–7% of all intracranial vessel dissections.[3] Dissections usually tend to occur when intrinsic fragility together with underlying arteriopathies cause structural weakening of arterial walls.[1,2] Definitive diagnosis is based on the combination of neuroimaging methods and DSA remains the gold standard.[1] After the first MRA study, aside from most probable signs of moyamoya vasculopathy in our patient, the diagnosis of BAD was not conclusive enough. Both basilar artery dissection and moyamoya disease were confirmed by subsequent DSA.

It is widely known that MMD has a bimodal distribution of age at onset and a slight female gender predomination.[5] While the most of moyamoya patients have lesions in areas of anterior circulation, combined steno-occlusive changes in both anterior and posterior circulations, such were in our patient, are extremely rare.[4,6] We performed a detailed laboratory workup, regarding the etiology, and the possibility of moyamoya syndrome was excluded.

The first case of BAD occurrence in a patient with MMD was reported by Abe *et al.*,[7] but the exact pathological process behind this extremely rare association has not been elucidated yet. Processes of intimal hyperplasia and thinning of tunica media are well-known characteristics of MMD.[4,5] Moreover, the abberrant vasculogenesis and excessive production of extracellular matrix proteins contribute to the advancing fragility of blood vessels.[4,5] Previous histopathological studies of intracranial vertebral artery dissections (VAD) confirmed connection with the degeneration of the internal elastic lamina and subsequent rupture of the medial layer.[8] Taken together, these events could most probably lead to the uncommon appearance of ICAD[9] and even BAD in our patient. Some genetic studies reported a smaller prevalence of RNF213 c.14576G > A polymorphism in cases of intracranial VAD compared to MMD, suggesting that it may be preferentially related to strokes in anterior circulation.[10] Other gene mutations affecting the biosynthesis of extracellular matrix and smooth muscle cell contractile system, might have a role in ICAD[10] but were unavailable for us to detect.

Taken all into account, we presented an extremely rare case of spontaneous BAD associated with de novo diagnosis of late-stage MMD, along with multiple stenotic changes in posterior circulation and with satisfactory functional outcome. Whether BAD and MMD occurred coincidentally or share common pathological processes is still uncertain. Future studies are expected to give more conclusive proof of the etiology and mechanism of intracranial dissections in patients with underlying moyamoya disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Aleksandra Lucic Prokin[1,2], Vladimir Galic[1,2], Nikola Boban[2,3]

[1]Department of Neurology, University Clinical Center of Vojvodina, Novi Sad, [2]Faculty of Medicine, University of Novi Sad, Novi Sad, [3]Center for Radiology, University Clinical Center of Vojvodina, Novi Sad, Serbia

Address for correspondence: Prof. Aleksandra Lucic Prokin, Hajduk Veljkova Street 1-9, 21000 Novi Sad, Serbia.
E-mail: aleksandra.lucic-prokin@mf.uns.ac.rs

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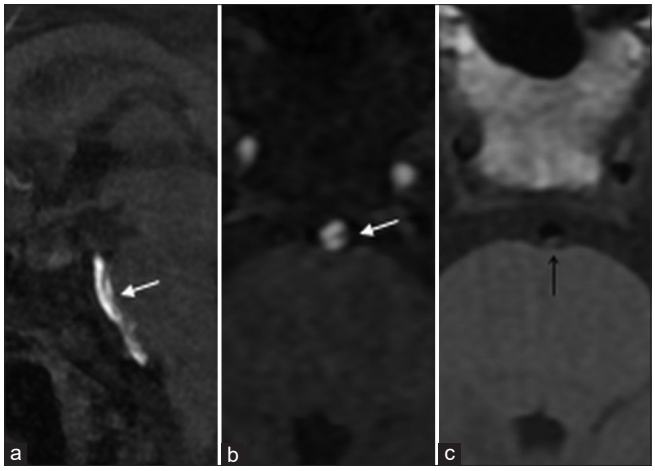
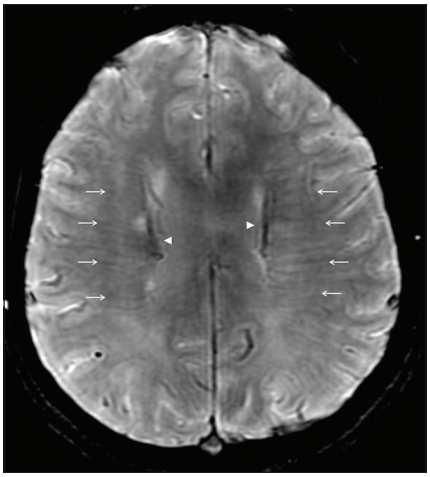


Supplementary Figure 1: (a, b) MRA: axial and sagittal T1W tomograms at the level of pons show defect in the flow signal – dissection flap in the BA (white arrows). (c) Axial T1W tomogram displays hyperdensity in the posterior aspect of BA, which represents intramural hematoma (black arrow)



Supplementary Figure 2: MRI [(SWAN) sequence]: numerous, deep medullar veins (white arrows) at the level of corona radiata, which drain towards subependymal veins (white arrowheads) – "brush" sign