Acta Scientific Clinical Case Reports

Volume 2 Issue 11 November 2021

Case Report

An Interesting Case of Diffuse ICA Collapse in the Neck - The "Champagne Bottleneck" Sign

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Received: September 28, 2021

Published: October 23, 2021

Abstract

We herein report a 26-year young female who experienced sudden onset loss of consciousness and unilateral limb weakness (side not recalled) on her postpartum Day 1, of her first pregnancy 2 years back, followed by gradual recovery of her motor weakness, but with residual deficits in speech, comprehension, cognition and memory. Her neuroimaging now shows old large bilateral middle and posterior cerebral artery territorial infarcts. Angiogram revealed non-visualisation of bilateral internal carotid and extensive "smoke-like" collateral Moya-Moya circulation in brain. However, interesting additional findings were bilateral extracranial (cervical portion) narrowing of proximal internal carotids up to carotid bulb, the so called "Champagne bottle neck" sign.

Keywords: Postpartum; Bilateral Large Infarcts; Moya-Moya; Extracranial ICA Narrowing; Carotid Collapse; Champagne Bottle Neck Sign

Case Report

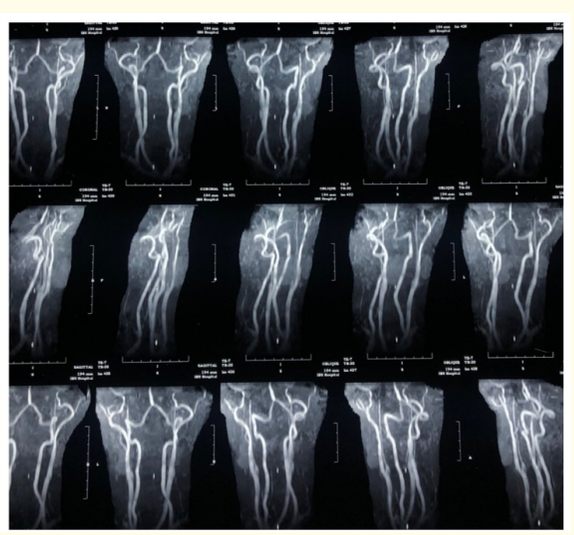
Patient is a 26-year female, married, without history of any chronic illness, and presented with complaints of altered behaviour, not understanding vocal commands, unable to recognize family members and unable to perform daily routine activities without help for the last 2 yrs. Patient was apparently well before 2 years, when she was admitted at some local hospital at full term pregnancy and planned for LSCS. There was no history of prior miscarriages or foetal loss. After she was received in ward post procedure, she didn't regain her consciousness for the next 3 days, after which investigations were performed to look into the matter. A CT Brain performed at that time showed bilateral Parieto-temporo-occipital hypo intensity, which later in MRI Brain revealed restricted diffusion in this region. This was interpreted as bilateral MCA Territory Infarcts, with other possibilities of Posterior Reversible Encephalopathy syndromes and remotely Encephalitis according to the medical record documents. Patient was managed conservatively with nasogastric feeding, bladder catheterisation, antiplatelets and iv fluids. CT angiography brain, as per documents, revealed "puff of smoke" appearance of vessels due to extensive collaterals. With deterioration in general condition post operatively, patient was taken home by family members. Over time, she gradually recovered and was able to speak, move about, and walk independently over next 6 months, however there were residual abnormalities in speech in form of slurred unclear speech, not understanding vocal commands, behaviour in form of erratic acts of shouting, physical violence, emotional incontinence in form of inappropriate laughing, along with non-recognition of family members. Besides, she used to walk with dragging of bilateral lower limbs. Occasional bladder and bowel incontinence was seen. However, no associated history of fever, seizures, residual limb weakness, facial weakness, or swallowing difficulty were present.

Presently, patient was brought with non-improving static nature of these complaints. History was suggestive of post stroke residual deficit due to involvement of bilateral temporo-occipito-

parietal lobes. Previous MRI was discussed and interpreted as bilateral massive infarcts. PRES was unlikely with residual gliosis in subsequent films. Clinical scenario and normal CSF at that time grossly ruled out infective etiologies like encephalitis. Also, the pre-existing presence of extensive collaterals in cerebral angiography was suggestive of a pre-existing chronic ischemia, and hence infarcts were more likely precipitated rather than caused by the postpartum state.

Her investigations including full blood count, electrolytes, renal and liver function, erythrocyte sedimentation rate, retroviral status, homocysteine, HbA1c, Antineutrophil cytoplasmic antibody, antinuclear antibody, double stranded DNA, complements, thyroid function test, rapid plasma reagin, lupus anticoagulant, thrombophilia screen tests, cerebrospinal fluid examination and echocardiogram were all normal. Therefore, a repeat neuroimaging was planned.

MRI Brain (Figure 3) revealed T1 hypo intense, T2 and FLAIR hyper intense lesions involving bilateral temporal, parietal and occipital lobes. There was no active diffusion restriction, apparent diffusion coefficient was hyper intense in same territory, suggestive of old gliosis. Angiography (Figure 1,2) of neck and brain vessels interestingly showed extensive collaterals in brain filling from bilateral posterior cerebral arteries (PCA), giving the well-known Moya-Moya or puff of smoke appearance. This was also suggested by non-visualised bilateral internal carotids, and angiography neck revealing diffusely tapered ICA distal to the carotid bulb on both sides, with normal calibre of external carotids and bilateral vertebral arteries. PCA territory infarctions can be explained due to P1 segment cut-off on left more than right side, and possible steal phenomenon.



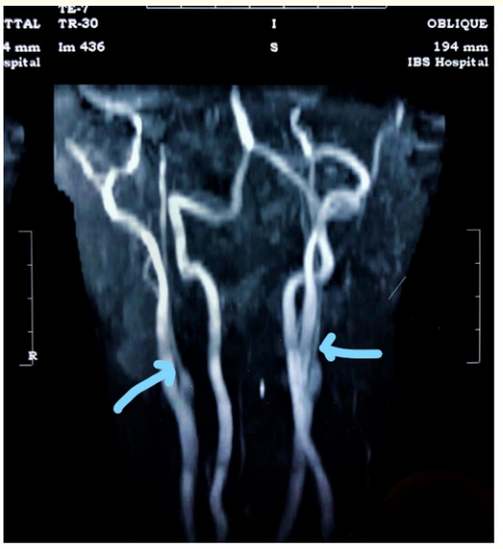


Figure 1

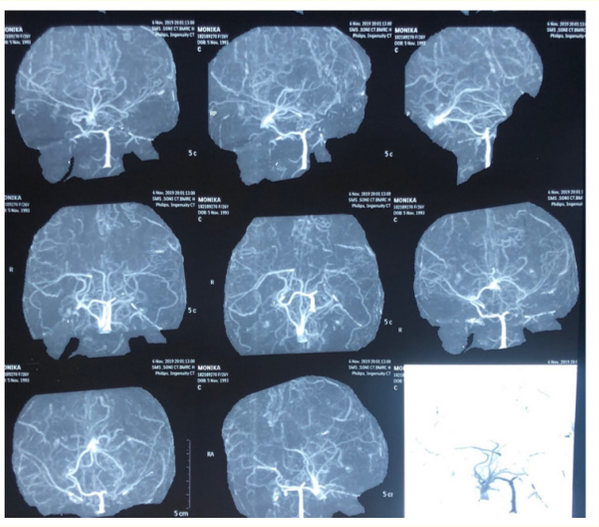


Figure 2

This was a very unusual finding as there was no thickening of vessel wall of ICA in cross sectional views, as seen with arteritis, and was more likely collapse of the vessel along with the vessel wall. Besides, arteritis/vasculitis is unlikely to spare all other vessels and specifically involve bilateral ICA only. Furthermore, primary Moya-Moya disease is mostly known to be associated only with distal ICA occlusion sparing the proximal extracranial part [1,2].

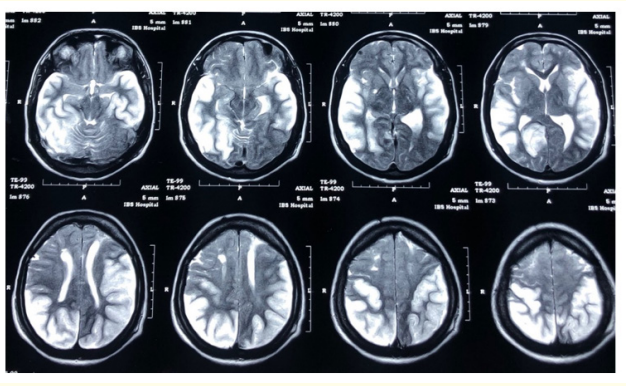


Figure 3

Literature [3-5] was reviewed for such lesion, and we concluded that this was the "Champagne bottle neck" sign seen with advanced Moya-Moya disease, wherein following a distal ICA steno-occlusive lesion, the proximal extracranial ICA collapses except for the carotid bulb. Thus, diagnosis of "primary Moya-Moya disease with bilateral large parieto-occipital-temporal infarctions associated with bilateral long segment ICA collapse" was made.

Conclusion

Traditionally, focal narrowing of extracranial carotids is considered atherosclerotic, while diffuse and uniform ones are considered vasculitic in origin. However, we hereby emphasise that a collapse of extracranial ICA till carotid bulb can be a rare manifestation of distal ICA stenosis like that in Moya-Moya disease, leading to peculiar appearance of a champagne bottleneck, and hence diagnostically helpful.

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