

Unilateral steno-occlusive endoluminal proliferative cerebral angiopathy of middle cerebral artery causing hydrocephalus ex-vacuo.

Ramesh Sangle¹, Abhijit Mahaveer Patil², Amarjit Singh²

Departments of Neurosurgery¹ and Radiology²,
Dr. D.Y. Patil Medical College & Research Centre, Sant Tukaram Nagar,
Pimpri, Pune, Maharashtra, India – 411018

ABSTRACT A 11-year-old girl presented with hemiparesis and hydrocephalus ex vacuo. Magnetic resonance angiography showed endoluminal steno-occlusive angiopathy of the middle cerebral artery (MCA). It's a rare case wherein angiopathy remained unilateral and without moyamoya (collateral vessel) formation.

Key words: Cerebral angiography, Endoluminal proliferative angiopathy, Hydrocephalus ex vacuo, Middle cerebral artery, Moyamoya Disease

Abbreviations: CSF – Cerebrospinal Fluid, CT - Computed Tomogram; DSA - Digital Subtraction Angiography, EEG – Electroencephalography, ICA – internal carotid artery, MCA – Middle Cerebral Artery, MMD – Moyamoya Disease, MRI – Magnetic Resonance Image

Introduction

Hydrocephalus ex vacuo is condition in which the CSF volume is increased due to encephalic volume loss [1]. In 1957 it was first described in patients with hypoplasia of bilateral internal carotid arteries [2]. Radiologically it appears as enlargement of cerebral ventricles and subarachnoid spaces.

Proliferative cerebral angiopathy is progressive and irreversible disorder that involves intracranial vessels at the skull base. It can result in aneurysmal, dolicho or endoluminal cellular proliferation of cerebral vessels. Endoluminal proliferation leads to luminal stenosis or occlusion. Intra-arterial steno-occlusive process is neither inflammatory nor atherosclerotic in nature. [3] Striatal collaterals that occur in response to luminal occlusion are seen as “cloudy puff of smoke” in angiography and hence this condition is called as moyamoya disease in Japanese language [4].

We report a rare combination of unilateral steno-occlusive

Address for correspondence

Abhijit Mahaveer Patil
Flat 203, La-Chapelle Co.op HSG Society, Baner Road, Near Hotel Mahabaleshwar Pune - 411045.
Mobile phone: 9766006688 ;
Email: abhi_patil6688@yahoo.com

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intraluminal proliferative angiopathy without moyamoya formation presenting with hydrocephalus ex vacuo.

Case report

A 11-year-old girl presented with gradual onset of progressive weakness of the left upper limb. There was no history of headache, vomiting, visual disturbance, trauma or fever. She was neurologically normal until the age of 4 years when she developed two generalized seizures with loss of consciousness. Clinical examination, CSF analysis, EEG and CT scan of brain done at that time did not show any abnormality. (Fig.1) Epilepsy was diagnosed and treated with oral carbamazepine. Her younger sister was healthy.



Fig 1 – Non-contrast CT Brain done at 4 years of age showing normal appearing brain

Parents noticed gradual and progressive deterioration of her neuro-cognitive functions, more noticeable in the last 3-4 years. Intelligence, memory and speech were severely affected. General physical examination did not reveal anything abnormal. Neurologically, she was fully conscious and could speak a few words. Cranial nerves were normal. Optic fundus examination showed no papilloedema. The neuro-cognitive functions were deranged. Upper motor neuron type of hemiparesis with brisk deep tendon reflexes and up-going plantar response was noted on the right side. On the left side hemiparesis of lower motor neuron type was found.

MRI brain showed unilateral hydrocephalus of the right side with cerebral volume loss (Fig. 2). Magnetic resonance angiography showed steno-occlusive angiopathy involving the distal segment (M1) of right MCA only. M2, M3, and M4 were all found to be occluded. Internal carotid artery, anterior cerebral artery as well as vertebro-basilar system were normal on both sides. Left MCA, M2, M3, M4 were normal (Fig. 3). Ventricular shunt operation was recommended for hydrocephalus.

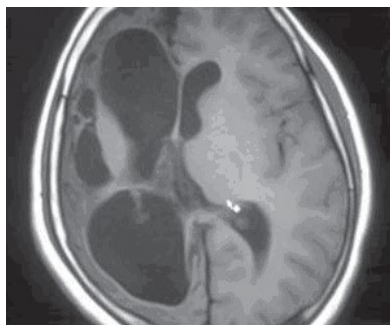


Fig 2 – Magnetic Resonance Imaging of the brain (1.5 Tesla; T1 image) at 11years of age showing unilateral hydrocephalus-ex vacuo on the right side. On diffusion weighted MRI-images cause of brain volume loss was confirmed as ischemic atrophy. The fronto-parietal, temporo-occipital, telencephalic affected areas are supplied by middle cerebral artery.



Fig 3 - MR-angiography: Steno-occlusive angiopathy involving the distal (M1) of right MCA only. M2, M3, and M4 all occluded. ICAs, ACA bilaterally and Vertebro-Basilar system normal. Left MCA, M2, M3, M4 were normal.

Discussion

Pediatric cerebro-vascular disease is extremely rare with

a prevalence of approximately 3 cases per 100,000 children [7]. MMD in pediatric patients has bimodal presentation [5] with a peak distribution at the age of 5-years [9,10]. Unilateral disease is a rare phenomenon.[5] Contralateral disease develops in up to 40% of patients with unilateral disease.[6] Females are twice more affected than males [8].

About 60% of childhood MMD gradually deteriorate in their cognitive functions [6]. The mortality rate of 4.3% is due to repeated stroke [7]. Only 2.8% Asian children present with the hemorrhagic strokes.[10] Different stages of MMD correlate well with angiographic findings. Severity of MMD reflects higher grades of occlusion of ICA and disappearance of collateral moyamoya vessels. “Puff of smoke” appearance in angiography caused by these collateral vessels represents intermediate stage of the disease. Only a few case reports of isolated MCA angiopathy in their initial stages have been reported [11]

Cortical atrophy is known to occur in various arterial and venous disorders. Malformed vein of Galen causes atrophy of the occipital and infra-temporal lobes. The infra-tentorial atrophic areas get compensated with CSF cavity formation called as Dandy Walker cyst [12]. Brain atrophy of cerebro-facial metamerism syndrome (Sturge-Weber) clinically manifest as seizure. DSA shows calcified superficial cortical veins, enlarged deep veins, cortical atrophy and angiogenesis [13]. Occlusive angiopathy is also seen in “PHACE/S” syndrome which includes haemangioma, coarctation of aorta, posterior fossa malformation, vascular abnormality in the eye and stroke. [14] Proliferative diffuse-angiopathy, seen in 20% of vascular diseases in children [15], is a telencephalic arteriovenous malformation with diffuse neo-angiogenesis without nidus-formation. Abnormal feeding arteries and draining veins are scattered amongst normal brain parenchyma.

Endovascular proliferative angiopathy is an important cause of cortical atrophy. It may occur during fetal or neonatal periods, infancy, early childhood or adolescence. Understanding the embryology and phylogenetic evolution of MCA is essential for the understanding the patho-anatomy of MMD. The MCA is formed of the anastomotic channels within the lateral striate arteries and it does not contribute to the formation of the Circle of Willis. MCA originates as a branch from anterior cerebral artery. Radiologically descriptive terms such as M1 (sphenoidal), M2 (insular), M3 (opercular) and M4 (cortical) are used to denote various segments of the MCA. Although M1 and M2 appear as one continuous artery they have different phylogenetic anatomy. M1 segment supplies lenticulostrates and lateral olfactory areas while M2 segment arising from lateral striate artery supplies

telencephalon (frontal parietal, occipital lobes), caudate nucleus and anterior limb of the internal-capsule. Phylogenetically M2 segment evolved several million years later than the M1 segment.

The incidence of accessory MCA which supplies basal ganglia occurs in 0.3% to 4%. [9] It is medial striatal artery and can be seen as hypoplastic or dominant type. The type1 accessory MCA is more proximal and it appears as if arising from the ICA. Type 2 and type 3 accessory MCA are seen arising from the ACA. Type 2 has more striatal and less cortical supply, but arises more proximally than the type 3 accessory MCA.

In our case significant change in radiological features developed within a span of 6 to7 years. It could be postulated that the MCA was well matured and diseased-free until 4-years of age. This is evident from the well developed normal size cerebral-hemispheres and ventricles. Telencephalon had also been well developed and foramen of Monro was intact. At 11 years of age hydrocephalus ex-vacuo and cortical hemiatrophy developed. MR angiography showed underlying atypical unilateral angiopathy of distal M1 segment of MCA. MCA occlusion is not associated with collaterals in lenticulostriate or with transdural angiogenesis. Trigger factors in MMD typically affect supraclinoid ICA along with proximal segment of the ACA. It is usually bilateral. "Trigger-factors" have different pathological effects on M1 and M2 segments. In this case spontaneous M1 occlusion, without non-sprouting angiogenesis resulted in complete ischemic atrophy of striato-telencephalic portion.

Conclusion

Only a few case reports of isolated MCA angiopathy in their initial stages have been reported.

Clinically, deteriorating neuro-cognitive functions of the child at different stages must always be co-related with angiographic studies and followed-up at least once in every two years. Therapeutic window-period and the time lost, play important role in the overall management results.

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