CASE REPORT

IMAGING FEATURES OF ARTERIAL ISCHEMIC STROKE IN A 13 YEAR OLD BOY - AN INTERESTING CASE REPORT

Karthik Krishna Ramakrishnan, Debashis Dakshit

Department of Radio Diagnosis, Medical College, Kolkata, W.B, India

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ABSTRACT

Childhood arterial ischemic stroke is a rare disease with an estimated incidence of 2-13/100000 per year.[1] Variability in the clinical presentation, the inability of children to effectively communicate their symptoms, lack of early clinical suspicion and overlapping of symptoms with many other diseases gives rise to delay in the diagnosis and delayed treatment. This leads to multiple long term co morbidity, disability and socioeconomic dependency with affected quality of life. The causes and risk factors of childhood ischemic stroke aremany, differing greatly from that of adult ischemic stroke[2,3,4,5] and needs a comprehensive diagnostic work up including imaging techniques like MRA, DSA.[7,8,9] The etiology should be accurately deduced since future recurrence rates depend on the etiology and so are the specific treatments with medical management like steroids for vasculitis or surgery or minimally invasive endovascular options[10] for stenotic lesions. The aim of this case report is to highlight the usefulness of DSA in diagnosing the etiology of childhood Arterial Ischemic Stroke (AIS) when the MRA remained inconclusive.

Key words: Stroke, childhood, vasculitis, ais, mca stenosis, MRA, DSA, endovascular therapy.

ABBREVIATIONS: AIS - Arterial Ischemic Stroke, CT - Computed Tomography, NCCT - Non contrast CT, CECT - Contrast Enhanced CTCTA - CT Angiography, MRI - Magnetic Resonance Imaging, MRA - Magnetic Resonance Angiography, DSA- Digital Substraction Angiography, DWI - Diffusion Weighted Imaging, FLAIR - Fluid Attenuated Inversion Recovery, ACA - Anterior Cerebral Artery, MCA - Middle Cerebral Artery, PCA - Posterior Cerebral Artery, VA - Vertebral Artery

Introduction

Stroke is a clinical syndrome which refers to the sudden neurological deficit due to a cerebrovascular event. It is can be hemorrhagic stroke (15%) or Ischemic stroke (80%). Ischemic stroke occurs when a blood vessel carrying blood to the brain is blocked by a blood clot. This causes loss of blood supply to certain part of the brain and resulting in brain damage. When artery is blocked, the term arterial ischemic stroke is used. If a vein is blocked causing stroke, the term cerebral venous thrombosis is used. Arterial ischemic stroke can occur in two ways- thrombotic or embolic.

Stroke in childhood is rare but is associated with relative high morbidity, mortality, recurrence rates and is responsible for neuro psychological disability. It is frequently undiagnosed or misdiagnosed due to a low level of clinical suspicion and patients presenting with subtle symptoms that mimic other diseases and thus leading to a delay in the diagnosis of stroke.

Given that therapeutic decisions depend on stroke etiology, accurate diagnosis is crucial for proper management of these patients.

If a stroke is suspected in a child, the diagnostic work-up should include systemic blood tests (haemato-

Correspondence : Dr. Karthik Krishna Ramakrishnan
Department of Radio Diagnosis,
Medical College, Kolkata,
W.B, India
Email: kkdrkr@gmail.com

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logical disorders, infections, genetic and cardiac examinations.

The neuroradiological diagnosis comprehends CT scan, CT angiography, MR imaging with MRA and DSA.

MR imaging with MRA better delineates the brain tissue status, the presence and nature of the lesion, and sometime the timing of possible structural lesions, the probable cause of Arterial Ischemic Stroke (AIS).

In this case MRA, as an initial screening modality of choice could not detect the cause of AIS.

DSA could identify the cause of AIS in this case. DSA is done only in selected cases because of its inherent complications. It is the gold standard for diagnosis of vascular pathologies.

In the appropriate setting, besides medical treatment, minimally invasive procedures with endovascular approach to be considered.

Case Report

The case under study is a 14 year old boy asymptomatic previously, suddenly presented with a brief spell of dizziness and fell down while playing, injuring his scalp posteriorly.

The boy did not have any loss of consciousness or altered awareness, vomiting, seizures, headache, loss of vision or difficulty in speaking.

During his 10 day hospital stay, initially his mother noted deviation of angle of mouth to the right, followed by gradual onset left upper limb and lower limb weakness slowly progressing over two days to reach a static point on the third day.

The weakness was non progressive from there on, but left upper limb was involved more than left lower limb. After the third day the symptoms started improving with good recovery of the left upper limb and on the day of DSA the patient was having only mild walking difficulty. DSA was done on the eighth day after admission. By then he had undergone an overall general medical examination in which the CVS, RS, Abdominal examination were within normal limits.

Detailed neurological examination was done that revealed only mild motor weakness of left upper and lower limb with power of 3/5 and gait abnormality on left side due to weakness.

The higher functions, sensory, speech and autonomic functions, cerebellar function tests and reflexes were normal except for extensor plantar on left foot.

All the basic blood investigations like complete hemogram, renal function tests, liver function tests, coagulation profile and special tests like Anti-Nuclear antibody. Antiglyoprotein B2 antibody testing and anti cardiolipin antibody testing which were all normal. The patient's baseline ECG and echocardiography, chest radiograph were normal.

Imaging Findings

The initial NCCT brain revealed normal study except mildscalp swelling in the high parietal regionposteriorly. MRI brain with MRA revealed a focal area of acute infarct in right internal capsule in the posterior limb with diffusion restriction on DWI.

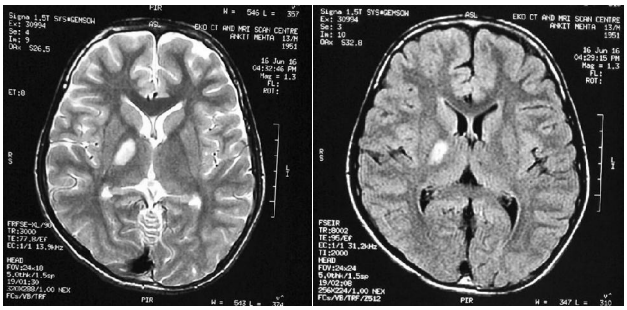

Figure 1: T2 WI and T2 FLAIR shows a focal area of hyperintensity (S/O acute infarct) in the posterior limb of right internal capsule

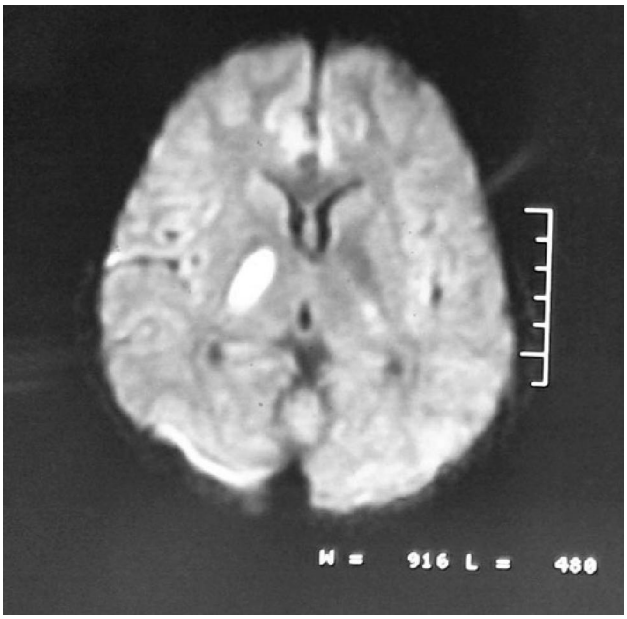

Figure 2: DWI reveals diffusion restriction confirming acute infarct

Figure 3: MRA reveals a normal study besides a hypoplastic right ACA

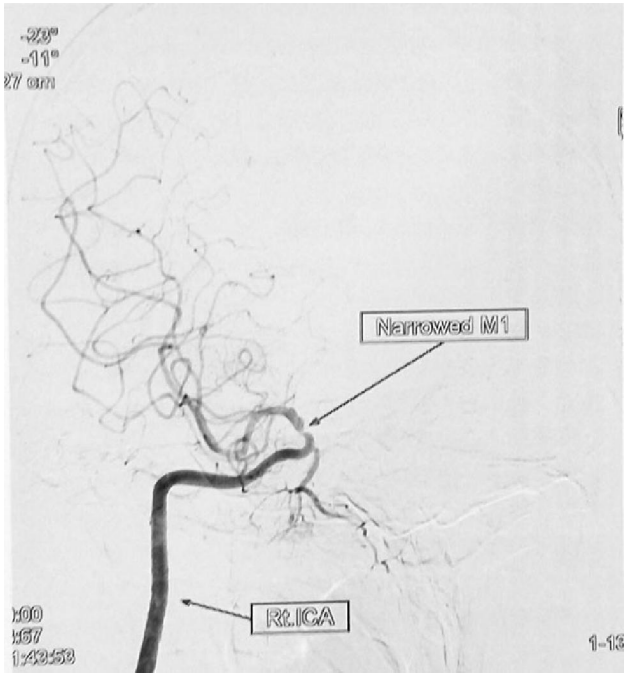

Figure 4: DSA reveals significant stenosis of >90% narrowing in the proximal M1 segment of rt. MCA

MRA revealed a normal study besides a hypoplastic right ACA (Anterior cerebral artery).

The four vesselcerebral DSA revealed significant stenosis of >90% narrowing in the proximal M1 segment of Rt. MCA (Middle cerebral artery) with preserved flow distal to the stenosis. Reduced flow was noted in the lateral lenticulostriate branches of Rt. MCA. Development of collaterals was not found which would have saved the brain from ischemia. The study also confirmed the hypoplastic Rt. ACA, a normal variant.

Treatment Given

This patient's neurological condition improved over 10 days and by the time of DSA his gait had also improved considerably. He was given supportive treatment along with oral steroids. He was later referred to Institute of Neurosciences for further workup and treatment.

Discussion

Arterial blood supply to the brain is through circle of Willis which is formed by two major systems, the anterior circulation by bilateral internal carotid arteries (ICA) and its branches anterior cerebral artery (ACA) and middle cerebral artery (MCA) and the posterior circulation by the vertebral artery (VA) and posterior cerebral artery (PCA). The majority of brain parenchyma is supplied by MCA, the largest artery which is commonly involved in AIS in children.

AIS is the most common type of stroke in children in which the artery supplying the brain parenchyma gets blocked leading on to ischemic brain damage.

Unlike adults, AIS in children is rare event with incidence around 2 to 13 cases per 100,000 children[1] under 18 years of age. High incidences of stroke in young individuals have been reported in developing countries also.Stroke is more common in boys than girls especially during adolescence. Black racial preponderance is also seen.

For children who have an initial stroke, the risk for recurrent strokes is between 15% and 18%.

Of children surviving stroke, about 60% will have permanent neurological deficits, most commonly hemiparesis or hemiplegia.

As therapeutic decision depends on stroke etiology, accurate diagnosis is crucial for proper management. AIS in children is frequently undiagnosed or misdiagnosed due to a low level of suspicion by the clinician and patients presenting with subtle symptoms that mimic other diseases and hence the delay in the diagnosis.

AIS most often presents as a focal neurologic deficit commonly hemiplegia, hemiparesis, aphasia and speech difficulties, visual deficits and headache.

Risk Factors and Causes:

• Arteriopathies,[2,3,4] – These are the leading cause of AIS and is associated with poor outcome.E.g.:

o Transient cerebral arteriopathy (TCA)

o Focal cerebral arteriopathy of childhood (FCA)

o Post-varicella angiopathy

o Craniocervical arterial dissection

o Moyamoya disease, fibromuscular dysplasia, systemic vasculitis, collagen disorders, vasculitis secondary to CNS infections, leukemia, radiation therapy.

o Neurocutaneous syndromes like neurofibromatosis 1 (NF1) arterial dysplasia.

• Cardiac disease – cyanotic, rheumatic, valvular and congenital like patent foramen ovale (PFO)[5]

• Sickle cell disease (SCD)

• Iron deficiency anemia.

Diagnosis:

Early recognition and treatment during the first hours and days after a stroke is critical in optimizing long-term functional outcomes and minimizing recurrence risk. Early treatment focuses on protecting the brain and keeping the blood vessels open to prevent further brain damage and to prevent more strokes.

Laboratory assessment may include a variety of nonspecific blood tests and more specific laboratory tests such as work up for coagulopathies, hematological disorders &vasculitis.

Imaging and Testing

Non-contrast head computed tomography (CT)of brain remain often the initial test, readily available and done within minutes. It is done mainly to rule out hemorrhagic stroke.[6]

On CT Infarcts appear as low-density lesions within vascular territories, but can be missed if too small or if the CT is done very early in the course of the disease.

CT angiography (CTA) with perfusion- can help us to assess vascular anatomy and relative cerebral blood flow.[6]

MRI: T2 and DWI with ADC map – can easily pickup areas of acute infarct within few hours of onset of occlusion.[7,8]

MRA - can give us an idea about the vessel involved in occlusive process. It is a useful noninvasive screening tool.[8]

DSA

• gives the most precise detail of vascular anatomy of all imaging modalities[9]

• is superior to MRA and computed tomography angiography (CTA) for visualization of tertiary branches and small cerebral arteries[9]

• can be performed in conjunction with endovascular procedures like balloon angioplasty or stenting.

• even though, DSA is an invasive procedure it should be strongly considered in cases with equivocal or negative findings on MR vascular imaging or where no other explanation for stroke is identified.

Differential diagnosis: Certain neurological conditions that mimic AIS like complicated hemiplegic migraine, postictal Todd's paresis, intracranial neoplasms & infections, metabolic abnormalities like hypoglycemia & MELAS.

Management:

Early treatment aims at early initiation of reperfusion to injured brain as soon as possible, preferably within 2 hrs, as the chances of brain tissue salvage is maximum during that period. Treatment can be broadly categorized as:

Neuro-protection

Supportive measures include:

Control of fever, normalization of serum glucose, maintenance of normal oxygenation and blood pressure, treatment of ^ICP, correcting dehydration, and anemia, early recognition and treatment of seizures.

Emergency Recanalization

t- PA, has a limited role in childhood stroke.

Anticoagulants especially LMWH can be used, its dose titrated by coagulation tests.

Antiplatelets like aspirin are proven to be very safe in children.

Additional measures:

• Infectious meningitis control by antibiotics and

antivirals.

- Supportive cardiac care for cardiac dysfunction in cardiogenic strokes.
- Early initiation of corticosteroids in vasculitis.
- Blood transfusion in SCD.

Surgical Management:

Surgical management for stenotic lesions consists of various bypass channels most commonly - EC-IC bypass - now being increasingly replaced by endovascular techniques.

Endovascular Options[10]

In childhood arterial ischemic stroke balloon angioplasty is the preferred first line endovascular surgical option and can be combined with any of the following.

(1) intra-arterial thrombolysis/ mechanical thrombectomy.[10]
(2) revascularization procedures with stenting.

Stenting

Stents have reduced the complications often associated with balloon angioplasty.

Advantages

- exclusion of the plaque and regions of dissection from the vessel lumen
- prevention of vessel recoil and rupture
- barrier to platelet aggregation of plaque
- provides additional wall support
- prevents acute closure by intimal flap
- reduces the incidence of restenosis

Conclusion

Etiology of childhood stroke is completely different from adult stroke. Arteriopathies, vasculitis, cardiogenic embolic stroke, sickle cell disease, and coagulopathies are common causes of AIS in children where as diabetes mellitus, hypertension are major causes in adult. So, proper and complete work up is required to find out the etiology and correction of primary cause to prevent future recurrence.

MRI with MRA is ideal modality in investigation of AIS. MRI with DWI with ADC map picks up the infarct very early within few hours of the onset of event. MRA is the initial screening procedure for vascular pathologies of brain.

DSA, in spite of its inherent complications is the gold standard in diagnosing vascular pathology. In order to reduce complications in children, DSA should be done in selected cases and done by experienced personal only in super-specialty setup. Minimally invasive endovascular procedures can also be combined with DSA.

Balloon angioplasty is the ideal endovascular intervention in this case of childhood AIS due to short segment stenosis of Rt. MCA and the patient has been accordingly counseled and referred to higher centre.

This case report is being presented for it's rarity of the etiological occurrence as AIS in children and to stress on the specificity of DSA over MRA as diagnostic tool in arriving at the etiology.

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