A Case report of 9-year-old child with basilar artery occlusion successfully treated with conservative medical management.

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ABSTRACT- Stroke is one of the major causes of childhood morbidity and mortality. Pediatric arterial ischemic stroke (PAIS) has an annual estimated rate as high as 3.3 cases per 100,000 children (with the vertebrobasilar territory involved in up to 36% of cases); however, the incidence of isolated childhood basilar artery occlusion (BAO) and stroke (BAS) is unknown. Adult BAO carries up to a 90% mortality rate, while death or severe neurologic deficits may be seen in 50% of children with BAO/BAS.

Here we are reporting a case of nine years old male child presented with right side hemiparesis who found to have basilar artery occlusion and the patient showed complete clinical recovery with the conservative medical management.

Index Terms-basilar artery occlusion, conservative management, low molecular heparin, aspirin.

INTRODUCTION:

(BAS) results from transient or persistent (BAO) and its branches and frequently presents with brainstem infarction. Occlusive thrombus arises either locally or as an embolus from proximal sources. Most of the brainstem is directly perfused from the basilar artery itself, although portions of the medulla and mid- brain are also supplied by proximal arteries feeding the basilar artery (vertebral arteries) or distal basilar artery branches (posterior cerebral artery). [1]

BAO can present with a variety of neurologic deficits, which may have an abrupt, progressive, or stuttering presentation. [2] Most children present with motor deficits, altered sensorium, and lower cranial nerve palsies. [1] Some children may also present with head- aches. Prodromal TIAs are reported in .50% of adults with BAO. Clinical symptoms of BAO vary with the anatomical level of the vascular obstruction. A proximal or middle segment BAO can result in pontine strokes, which may present with hemiplegia or quadriplegia, dysarthria, dysphagia, cranial nerve palsies, or reduced consciousness. [2] BAO presenting with subtle brainstem signs and uncrossed hemiplegia (ipsilateral lower face, arm, and leg weakness) may eventually progress to the locked-in state (quadriplegia and mutism with preserved sensorium). In such scenarios, the initial hemiparesis represents early stages of stroke-in-evolution and is known as herald hemiparesis. [3] Distal segment BAO may cause bilateral strokes in mesencephalic and thalamic areas (decreased sensorium, quadriparesis, and eye movement abnormalities). [2] Traumatic vertebral artery dissection is one of the most common causes of acute basilar artery occlusion (ABAO) in young patients and must be especially suspected in patients presenting with cervical pain preceding the dramatic neurological deterioration. [4] Delay in the diagnosis of ABAO is frequent due mainly to the misleading symptoms and signs and the rarity of this condition [5]

CASE REPORT

Our patient is 9-year-old boy presented with one day history of right-sided weakness without loss of consciousness. Three days before while riding bicycle on the slope of the road, he fell down and had one episode of vomiting without loss of consciousness level or seizures. No significant family history or past medical history.

On examination, his conscious level was normal but was having difficulty in walking due to right-sided weakness. His right facial nerve was also affected, with upper motor neuron lesion. Power of the right lower limb was 3/5 and upper limb was 3/5. The distal muscles were more severely affected compared to the proximal group of muscles. His speech was also affected, and he was not able to speak fluently. Left side was perfectly well. Rest of the cranial nerves and systemic examinations were normal.

We performed urgently MRI/MRA/MRV (figure 1) which showed acute brain stem infarction mainly on the left side of the pons with evidence of absent flow in the basilar artery. The differential diagnosis included dissection of the basilar artery especially with history of trauma or thrombosed basilar artery. We did CT angiography to differentiate between these two conditions (figure 2), which confirmed basilar artery occlusion and showed complete absent of flow in the main basilar artery suggestive of basilar artery thrombosis. We also requested laboratory investigations looking for the underlying cause of thrombosis. His CBC, Coagulation profile, thrombophilia workup were all within normal limits. Hemoglobin electrophoresis was also normal. Lipid profile was normal. His electrolyte , renal and liver functions were all normal.CBC:WBC: 6.05 x10^9/L , Hb:13.2g/dl , PLT:288 x10^9/Coagulation profile:PT:13.3 sec, INR: 1.2, PTT:32 sec, fibrinogen:221.7 mg/dl Protein S: 57.9% , protein C: 73.9% , Antithrombin III: 104.9% , factor VIII: 267.1 Hemoglobin electrophoresis: Hb A2: 2.7% , Hb A: 97% , HB S:0 , Hb F:0.3 Lipid profile: cholesterol : 3.85 mmol/L , triglyceride :0.92 mmol/L , HDL :1.31 mmol/L , LDL: 3.35 mmol/L Hemoglobin A1C :5.7%

We started LMWH (low molecular weight heparin) and Aspirin as conservative management. Follow up with the patient showed significant gradual improvement.

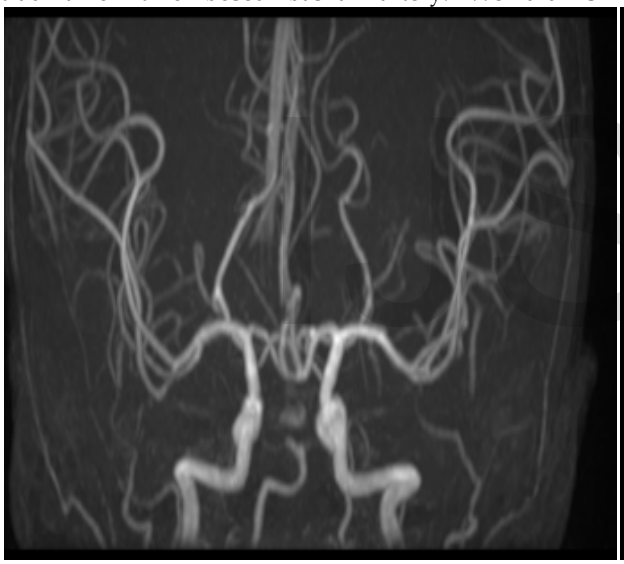

Fig: 1

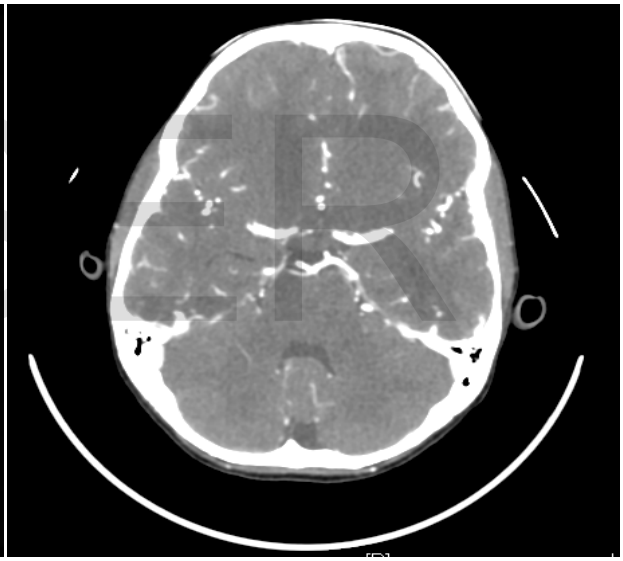

Fig : 2

Discussion:

(ABAO) is an infrequent but potentially fatal cause of stroke, both in adults and children. Locked-in syndrome is a common clinical pattern of this condition and is attributed to the occlusion of the mid portion of the basilar artery (BA) causing ischemic damage to the pontine pyramidal tracts. It is characterized by upper motor neuron quadriplegia, paralysis of lower cranial nerves, bilateral paresis of horizontal gaze and anarthria, with preserved consciousness. [6]

However, our patient was present with right sided weakness, right upper motor neuron lesion facial nerve palsy and speech was also affected as the patient was not able to speak fluently. There were no loss of consciousness. There was a history of trauma so initially we considered the diagnosis of traumatic brain injury.

The aetiology of basilar occlusion is unknown in the majority of cases, but it is associated with prothrombotic disorders including prothrombin gene and factorV Leiden mutation, vasculitis, trauma, spontaneous dissection and neonatal thromboembolism. [7]

Brainstem hemorrhage, space-occupying lesions with transtentorial herniation, metabolic or toxic encephalopathies, and disorders presenting with rapidly progressive cranial nerve dysfunction (Miller Fisher syndrome, botulism, or myasthenic crisis) may all mimic BAO/BAS. [2]

Osmotic demyelination syndromes, demyelinating disorders, and certain encephalitides can also involve the brainstem. Acute subarachnoid hemorrhage or basilar-type migraines should be considered in patients with symptoms of headaches and brainstem dysfunction. BAS-related myoclonic jerks, decerebrate posturing, and unresponsiveness may raise concerns for seizures. In a patient presenting with altered sensorium, cranial nerve dysfunction, and motor tract involvement, the index of suspicion for BAO should be high. [2]

In our patient we requested laboratory investigation to confirm the underlying cause of thrombosis His CBC , Coagulation profile , thrombophilia workup were all within normal limits. Hemoglobin electrophoresis was normal. Lipid profile was normal. His electrolyte, renal and liver functions were all normal.

We performed urgent MRI/MRA/MRV showed acute brain stem infarction mainly the left side of the pons with evidence of absent flow in the basilar artery, because the patient had history of trauma dissection was suspected CT angiography was done which confirmed basilar artery occlusion and showed complete absent of flow in the main basilar artery suggestive of basilar artery thrombosis.

We started our patient on low molecular heparin (LMH) and aspirin. The final decision was to continue on conservative management with LMWH and Aspirin Follow up with the patient showed significant gradual daily improvement. His neurological examination after eight days showed normal gait with normal tone, power and reflexes in all four limbs. His facial palsy improved as well as his speech. Follow up brain MRI showed significant improvement regarding the basilar artery perfusion.

Due to the low incidence of pediatric ischemic stroke, limited data are available, making the development of a standard treatment algorithm difficult. Historically, management of pediatric cases was patterned on adult cases, using intravenous (IV) tissue plasminogen activator (tPA). However, recent guidelines from the American Heart Association (AHA) do not recommend IV tPA in children outside of clinical trials. Recently, the Multicenter Randomized Clinical trial of Endovascular treatment for Acute ischemic stroke in the Netherlands (MR CLEAN) demonstrated that transarterial treatment improved outcomes in adult patients with severe stroke and proximal vessel occlusion within a 6 h window after ictus. Although these findings cannot be directly extrapolated to children, successful endovascular treatment for acute ischemic stroke has been reported with increasing frequency in this age group. [8,9] In a study done by Lagman-Bartolome et al 2013, showed that children with BAS with or without BAO receiving conservative medical treatment have better long term outcomes than adult.[1]

There are several possible explanations for better outcome in childhood BAS compared with adults. First, children with BAS have relatively low rates of comorbid neurological disorders. In contrast, adults with BAO frequently have comorbid atherosclerosis, hypertension, and dementia, which could worsen outcomes. Second, children generally have better collateral circulation because of their low rates of diffuse arterial disease. However, in general, brainstem circulation is poorly collateralized and therefore this is unlikely to be a major factor. Third, enhanced plasticity of the young brain may promote better recovery and outcomes in children after BAS [1]

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