



A Rare Pediatric Case of Top of the Basilar Syndrome

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Received: July 15, 2024

Revised: August 1, 2024

Accepted: August 6, 2024

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Arterial ischemic strokes, particularly those resulting from basilar artery occlusion, are exceedingly rare in children. Posterior circulation strokes account for approximately 15% to 20% of all ischemic strokes, and basilar artery occlusive strokes comprise only 1% to 4% of these cases [1]. Nonetheless, basilar arterial strokes are associated with significantly higher mortality rates than other types of stroke in pediatric patients. They require a prompt diagnosis and appropriate management. Top of the basilar syndrome, also known as distal basilar artery infarction, presents with a broad range of neurological symptoms. These can vary from mild manifestations to severe impairments affecting consciousness, motor and sensory functions, and cranial neuropathies, including visual, pupillary, and oculomotor deficits [2].

Here, we present a rare case of top of the basilar syndrome in a pediatric patient from South Korea. This case was reviewed and approved by the Institutional Review Board (IRB) of Keimyung University Dongsan Hospital (IRB No. 2024-05-023). The requirement for informed consent was waived by the IRB.

A 5-year-old boy visited the emergency department with dysarthria and right-sided weakness that had lasted for 1 hour. He also complained of disequilibrium 17 hours before admission. He had experienced headaches three to four times a week for the past 2 months, without a clearly identified pattern. He was born at 38 weeks

through a normal spontaneous vaginal delivery with a birth weight of 4.0 kg. His medical history was unremarkable, with no growth or developmental disorders and no familial history of epilepsy or cerebrovascular disease. Upon arrival at the emergency department, his height, weight, body mass index, and head circumference were 113 cm, 21.3 kg, 16.7 kg/m², and 52 cm, respectively. His vital signs were as follows: blood pressure, 120/80 mm Hg; pulse, 132 beats/min; respiratory rate, 33 breaths/min; and body temperature, 36.8°C. He was alert. His initial Glasgow Coma Scale was 15. A neurological examination revealed decreased motor strength in the right arm and leg (2–3 out of a scale 5), flattening of the right nasolabial fold, and a positive Romberg sign. Brain magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) demonstrated suspicious findings for distal basilar artery occlusion and fetal configurations of both posterior cerebral arteries (PCA) with a hypoplastic right PCA. These findings were associated with acute and chronic infarctions in both thalami, the pons, and the left cerebellum, indicative of top of the basilar syndrome (Fig. 1). Tests for vasculitis, coagulopathy, and heart disease were conducted to determine the cause of the stroke. Laboratory results for coagulopathy and autoimmune diseases were negative. A two-dimensional echocardiogram showed a very small atrial septal defect with a left-to-right shunt flow, but no other specific

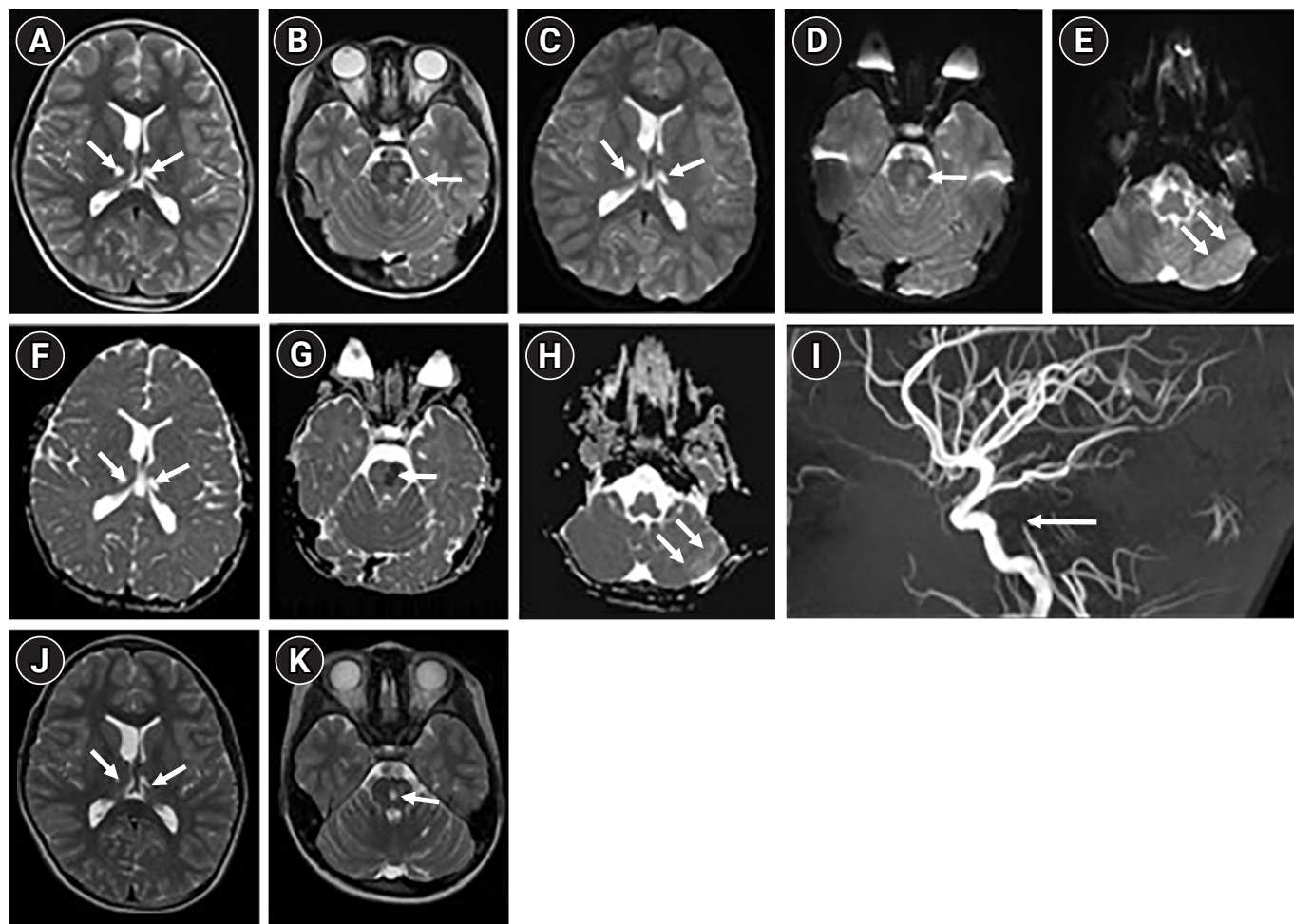


Fig. 1. Top of the basilar syndrome in a 4-year-old boy. Initial T2-weighted (A, B) images, diffusion-weighted images (C, D, E) and the apparent diffusion coefficient value (F, G, H) reveal bilateral symmetrical lesions in the posteromedial aspect of both thalami (white arrows), the pons (white arrow), and left cerebellar cortex (white arrows). Three-dimensional magnetic resonance angiography with the time-of-flight technique (I) shows no demonstrable flow signal intensity in the distal basilar artery (long white arrow) and both superior cerebellar arteries (white arrows). Follow-up T2-weighted images (J, K) reveal decreased sizes of the encephalomalacic lesions in both thalami and pons (white arrows).

findings. An electroencephalogram displayed normal waking and sleep tracings for his age. Treatment with aspirin (1 mg/kg/day) for anticoagulation commenced on the first day of hospitalization. Over time, there was improvement in his right hemiplegia, facial asymmetry, and dysarthria. On hospital day (HD) 11, follow-up brain MRI showed improvement in the left pontine infarction and no change in the encephalomalacic cavities in both thalami. Follow-up brain MRA showed no demonstrable flow signal intensity in the distal basilar artery, both superior cerebellar arteries, and the fetal configurations of both PCAs with a hypoplastic right PCA. He was discharged on aspirin and reported only a mild headache at the time of discharge. Three months post-discharge, follow-up brain MRI and MRA were performed. MRI showed encephalomalacic changes in the pons (9 mm) and both thalami (right <5

mm; left 10 mm). MRA indicated improved flow signal intensity with residual luminal narrowing in the distal basilar artery and improved flow signal intensity in both PCAs and the inferior cerebellar arteries. A multi-gene panel test for hereditary stroke-related mutations was negative. He continues on aspirin and is currently symptom-free, under follow-up at an outpatient clinic.

Although basilar artery stroke in children is rare, it is a serious condition with an unclear incidence. Research on this topic has been limited due to its rarity. A study conducted in Switzerland reported an incidence of approximately 0.037 per 100,000 children per year [3]. Specifically, occlusion at the top of the basilar artery can lead to a condition known as top of the basilar syndrome. This syndrome causes widespread lesions that affect the thalamus, temporal and occipital lobes, midbrain, pons, and cerebellum, due to

its anatomical characteristics. MRI is crucial for diagnosing top of the basilar syndrome and identifying these lesions due to its superior sensitivity to ischemic lesions to computed tomography [4].

Arterial strokes in children can result from several causes, including arteriopathies, cardiac disease, prothrombotic conditions, cerebrovascular anomalies, vasculitis, infection, and others. The 2011 International Pediatric Stroke Study identified arteriopathies and cardiac diseases as the predominant risk factors for pediatric stroke [5]. Unlike in adults, arteriopathies in pediatric patients are typically non-atherosclerotic [2,6]. Recently, genetic testing has also been used to identify the causes of pediatric stroke. In our case, after a comprehensive examination, the exact cause remained undetermined.

The symptoms of pediatric acute stroke are similar to those in adults and commonly include hemiparesis, hemifacial weakness, language disturbances, vision disturbances, and ataxia. However, pediatric strokes also frequently present with non-localizing symptoms such as headaches and altered mental status, and they have a relatively higher incidence of seizures. Posterior circulation stroke in children is particularly challenging to diagnose due to its non-specific and variable symptoms, which can often appear simultaneously. These symptoms tend to be mild at the onset of the stroke, contributing to delays in diagnosis [7].

Therapeutic options for stroke include intravascular thrombolysis, mechanical thrombectomy, anticoagulants, and aspirin, depending on the timing and etiology of the event. However, there is

still a lack of unified treatment guidelines for pediatric stroke, particularly regarding intravascular thrombolysis and endovascular thrombectomy. According to the latest pediatric stroke guidelines, administering recombinant tissue plasminogen activator to children aged 2 years and older is considered feasible if they present with persistent disabling neurological deficits and cerebral large artery occlusion confirmed by radiographic imaging within 4.5 hours of symptom onset [2,6]. Endovascular thrombectomy may also be considered within 6 hours of symptom onset [6]. However, previous studies have found that children have better outcomes and prognoses than adults [3]. Therefore, aggressive intervention remains a matter of debate, and it is important to select a treatment based on the overall condition of the individual patient.

In this case, the patient was admitted to the hospital 17 hours after experiencing disequilibrium and 1 hour following the onset of hemiplegia. The most prominent symptoms were hemiplegia and dysarthria, whereas altered mental status and visual or oculomotor impairments were less pronounced. An MRI confirmed a diagnosis of top of the basilar syndrome (Fig. 1). Given that 17 hours had elapsed since the onset of the stroke, thrombolysis was not considered an appropriate treatment option. Instead, aspirin therapy was initiated. By HD 3, there was a noticeable improvement in the patient's right-sided weakness, and the dysarthria began to improve by HD 5.

We observed that the patient's clinical improvement outpaced the changes seen on MRI, likely due to the greater plasticity of a

Table 1. Summary of the three other cases of basilar artery stroke.

Variable	Patient 1	Patient 2	Patient 3
Sex	Male	Male	Male
Age at diagnosis	11 years	15 years	5 years
Clinical manifestation	Sudden onset of neck pain, double vision, articulation disorder, fluctuating consciousness	Headache, vomiting, altered mental status	Frontal headache before fall down. Altered mental status with slurred speech after fall down
Neurologic examination	Gaze-changing vertical nystagmus, dysarthria, positive left Babinski sign	Right-sided weakness, dysarthria	Global aphasia, right-sided weakness
Brain MRI	Acute ischemic infarcts in the pons and the right cerebellar hemisphere	Acute infarction involving the upper third of pons and both cerebellar hemispheres	Restricted diffusion in the left pons and cerebellum, and right occipital lobe
Other imaging findings	CTA: Thrombus at the top of the basilar artery CA: Occluded basilar artery	CTA: Basilar artery occlusion at the level of anterior inferior cerebellar arteries CA: Detached intimal flap which closes the basilar artery completely or partially	CTA: Paucity of flow in the basilar artery CA: Irregular left vertebral lumen and two areas of stenosis due to dissection, and significantly diminished blood flow distal to the top of the basilar artery
Possible cause of stroke	Unknown	Spontaneous basilar artery dissection	Vertebral artery dissection
Management	Intravenous thrombolysis and endovascular thrombectomy	Endovascular mechanical thrombectomy and stent deployment	Intra-arterial thrombolysis and vertebral artery coiling
Outcome	Complete recovery after 3 months, except for slight dysarthria during physical exhaustion	Fully recovered after 11 months	Near complete recovery with deficit of mild incoordination and weakness in his right hand and forearm

MRI, magnetic resonance imaging; CTA, computed tomography angiography; CA, conventional angiography.

pediatric brain, which facilitates faster recovery. Fortunately, our patient experienced a favorable outcome without significant neurological sequelae thus far. However, given the high likelihood of recurrence for posterior circulation strokes [2], continued monitoring is necessary. Considering the rarity of this condition in pediatric patients, future multicenter studies are needed to collect similar cases. Among previously reported cases, some were suspected to involve top of the basilar syndrome [8-10]. The characteristics of these cases are summarized in Table 1. To the best of our knowledge, this is the first pediatric case of top of the basilar syndrome reported in South Korea.

Conflicts of interest

No potential conflict of interest relevant to this article was reported.

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Conceptualization: JYL and JCB. Data curation: JYL, HJL, and JCB. Methodology: HJL and JCB. Visualization: HJL. Writing -original draft: JYL. Writing - review & editing: JCB.

References

1. Demel SL, Broderick JP. Basilar occlusion syndromes: an update. *Neurohospitalist* 2015;5:142-50.
2. Ferriero DM, Fullerton HJ, Bernard TJ, Billingham L, Daniels SR, DeBaun MR, et al. Management of stroke in neonates and children: a scientific statement from the American Heart Association/American Stroke Association. *Stroke* 2019;50:e51-96.
3. Goeggel Simonetti B, Ritter B, Gautschi M, Wehrli E, Boltshauser E, Schmitt-Mechelke T, et al. Basilar artery stroke in childhood. *Dev Med Child Neurol* 2013;55:65-70.
4. Barkhof F, Valk J. "Top of the basilar" syndrome: a comparison of clinical and MR findings. *Neuroradiology* 1988;30:293-8.
5. Mackay MT, Wiznitzer M, Benedict SL, Lee KJ, Deveber GA, Ganesan V, et al. Arterial ischemic stroke risk factors: the International Pediatric Stroke Study. *Ann Neurol* 2011;69:130-40.
6. Mastrangelo M, Giordano L, Ricciardi G, De Michele M, Toni D, Leuzzi V. Acute ischemic stroke in childhood: a comprehensive review. *Eur J Pediatr* 2022;181:45-58.
7. Fink M, Slavova N, Grunt S, Perret E, Regenyi M, Steinlin M, et al. Posterior arterial ischemic stroke in childhood. *Stroke* 2019;50:2329-35.
8. Fink J, Sonnenborg L, Larsen LL, Born AP, Holtmannspotter M, Kondziella D. Basilar artery thrombosis in a child treated with intravenous tissue plasminogen activator and endovascular mechanical thrombectomy. *J Child Neurol* 2013;28:1521-6.
9. Borota L, Libard S, Fahlstrom M, Latini F, Lundstrom E. Complete functional recovery in a child after endovascular treatment of basilar artery occlusion caused by spontaneous dissection: a case report. *Childs Nerv Syst* 2022;38:1605-12.
10. Condie J, Shaibani A, Wainwright MS. Successful treatment of recurrent basilar artery occlusion with intra-arterial thrombolysis and vertebral artery coiling in a child. *Neurocrit Care* 2012;16:158-62.

1. Demel SL, Broderick JP. Basilar occlusion syndromes: an up-