Sir,

Childhood stroke usually occurs as a result of arterial ischemic strokes affecting around 1.6 per 1,00,000 children every year[1] and about 15%–22% of them involve posterior circulation. Causes of childhood ischemic stroke especially involving the posterior circulation are of different etiology compared to those in adults and include search for craniovertebral junction (CVJ) anomalies and vertebral artery dissection. CVJ anomalies usually present in the second or third decades of life[2] with a classical neurological involvement and there have been many case reports of CVJ anomaly leading to posterior circulation stroke[3] but the presentation of CVJ anomaly with a recurrent posterior circulation stroke is rare. We present here a case of 15-year-old boy with recurrent posterior circulation strokes secondary to atlantoaxial dislocation (AAD).

A 15-year-old boy presented with recurrent vertigo and vomiting in 2017 which lasted for a week. He was initially seen in an outpatient clinic and was prescribed antihistaminics considering the possibility of peripheral vertigo. His symptoms improved almost completely with only supportive treatment. Following this, he had recurrent intermittent episodes of vertigo. However, 20 months later he had an episode of vertigo, vomiting, and imbalance and eventually developed complete loss of consciousness. He improved with conservative management. During his second hospital admission, he developed unconsciousness with weakness of all four limbs (0/5 power). Given his recurrent neurological symptoms, a possibility of demyelination was considered and was treated with steroids. Though his sensorium improved over a period of next 10 days, he continued to have quadriplegic with only minimum gain in strength. He was referred to our center (third hospital admission) for a diagnostic review. He was admitted in our center, conscious, and well oriented. His motor system examination showed spasticity in all four limbs with brisk reflexes. Rt upper extremity power was 1/5, right lower extremity 3/5, left upper and lower extremity 4/5 with extensor plantar responses. On examination, he had a short neck with neck-height ratio of 1:10.5. Rest of the cranial nerve examination was normal.

Based on history and examination, we suspected posterior circulation stroke. Work up for vasculitis, sickle cell disease, and cardiac abnormalities was negative. His MRI brain (2017) Figure 2 showed acute infarcts in right thalamus, cerebellar hemispheres, and pons. His MRI brain showed bilateral pontine and thalamic infarcts with nonvisualization of both vertebral arteries (VA). His CT Brain [Figure 1] revealed persistent occiput terminale with midline integration defect of the basal odontoid.

Posterior circulation stroke (PCS) comprises infarction occurring in brain regions perfused by vertebral arteries which includes cerebellum, thalamus, brainstem, and occipital lobes. In children, PCS may have only brief or minor brainstem symptoms such as ataxia, vomiting, vertigo or may even lead to impaired consciousness which is what happened in our case. The risk factors, treatment options as well as underlying mechanisms of pediatric stroke are remarkably different from those in adults. Most common cause of posterior circulation strokes in children is vertebral arterial dissection which accounts for about half of all PCSs.[4]

Atlantoaxial dislocation, which is the most common anomaly especially in Indian population, may give rise to PCS.[5-8] Other CVJ anomalies causing PCS include odontoid aplasia, basilar invagination, occipitalization of the atlas, Klippel-Feil anomaly, and anomalous osseous process of the occipital bone projecting to the posterior arch of the atlas.[9] AAD refers to a loss of stable articulation between first and the second cervical vertebra. This instability predisposes VA to repeated undue compression and stretching leading to intimal damage or dissection that in turn giving rise to thrombosis.
In our patient, longstanding undiagnosed atlantoaxial dislocation lead probably to thrombosis and embolism causing recurrent ischemic strokes in the territory perfused by vertebral artery. Imaging in the index case is remarkable for a midline defect in the basal odontoid with a resultant bifid morphology, an extremely rare anomaly. Embryologically, it points to a faulty midline integration of the basal dental segment early in embryogenesis. The probable timepoints of affliction may be the mesenchymal prevertebral stage or the phase of chondrification. As with this case, a dislocated and persistent ossiculum terminale has been documented as an accompanying bony aberration.\[10,11\] Atlantoaxial instability co-exists as the central pivot is hypoplastic when bifid; this is further exaggerated by dislocated ossiculum terminale. Occasionally dynamic instability with flexion-extension may occur in hypermobile “hemi-os.” It is to be differentiated from bicornuate dens, wherein the abnormality is confined to the upper/terminal aspect.

The rate of recurrence for posterior circulation stroke generally varies from 20%–50% and is highest in children with arteriopathies and heart diseases. In a study done by Uohara et al.\[12\] on 107 patients of arterial ischemic stroke, out of the 46 patients with posterior circulation arterial ischemic stroke, 19% had a recurrence after the first episode of stroke. Males were found to be more likely to have recurrent strokes, especially secondary to VA dissection. Most of the patients with recurrent PCS also had a history of forceful or repeated neck movements or minor trauma to neck.

In our case, the boy initially diagnosed as peripheral vertigo and treated symptomatically. This delay in diagnosis is a common problem in children with stroke because of the nonspecific nature of the symptoms along with the difficulty of these young children in expressing their symptoms. Gurley and Edlow\[13\] have recently provided strategies such as consideration of posterior circulation stroke in patients with altered sensorium, attributing abrupt symptom onset to stroke and use of clear approach in diagnosing vertigo. Kulkarni et al.\[14\] in their review of 7 patients with posterior circulation stroke reiterate the importance of a careful physical examination in children with stroke. Delay in diagnosis and treatment especially in children with posterior circulation stroke would not only increase the morbidity but also increases the risk of recurrence. In conclusion, CVJ abnormalities should be ruled out in these patients since it is a potentially treatable risk factor. A good

Figure 1: Sagittal reformat (a) of noncontrast cervical CT shows a cleft in the basal odontoid (blue arrow) suggestive of a midline integration defect with persistent ossiculum terminale (white arrow). Note hypertrrophied anterior arch of atlas (yellow arrow). Coronal reformat (b, c) of the same shows persistent ossiculum terminale (white arrow in b and c) with midline integration defect of the basal odontoid (black arrow in b). Lateral cervical radiograph in flexion (d) shows anterior displacement of the anterior arch of atlas and ossiculum terminale (arrow) relative to the position in extension (arrow in e) and neutral position (arrow in f).

Figure 2: Sagittal T2 weighted image (a) shows persistent ossiculum terminale (white arrowhead) with midline integration defect of the basal odontoid (yellow arrow). Loss of flow void is seen in the basilar artery (red arrow) with a chronic infarct in the pons (black arrow). Axial T2 weighted images (b, c) show a chronic pontine infarct (black arrow in b) with loss of flow void in V4 segment of both vertebral arteries (arrowheads in c). TOF-MRA (d, e) show nonvisualization of both vertebral arteries and the basilar artery with bilateral fetal posterior cerebral arteries (arrows in d). Post-contrast axial T1-SPACE (f) shows subtle eccentric hyperintensity along the V4 segment of both vertebral arteries (white arrowheads)
physical examination, suspecting the possibility of PCS in a child with impairment of consciousness can yield important clues and can in turn prevent delay and misdiagnosis.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

N. Devavrat, K. Karthik¹, J. Shumyla¹, K. Neeraja, M. Netravathi

Departments of Neurology, *Neuroimaging and Interventional Neuroradiology (NIIR), National Institute of Mental Health and Neurosciences (NIMHANS), Bengaluru, Karnataka, India

**Address for correspondence:** Dr. M. Netravathi, Department of Neurology, National Institute of Mental Health and Neurosciences (NIMHANS), Hosur Road, Bangalore - 560029, Karnataka, India.

E-mail: sundernetra@yahoo.co.in

**References**

1. Mallick AA, Ganesan V, Kirkham FJ, Fallon P, Hodderly T, McShane T, \textit{et al.} Childhood arterial ischaemic stroke incidence, presenting features, and risk factors: A prospective population-based study. Lancet Neurol 2014;13:35-43.
2. Bharucha EP, Dastur HM. Craniovertebral Anomalies (A Report on 40 Cases). Brain J Neurol 1964;87:469-80.
3. Verma R, Sahu R, Ojha BK, Junewar V. Thalamic syndrome as the heralding manifestation of atlantoaxial dislocation. BMJ Case Rep 2013;2013:bcr2012007712.
4. Ganesan V, Chong WK, Cox TC, Chawda SJ, Prengler M, Kirkham FJ. Posterior circulation stroke in childhood: Risk factors and recurrence. Neurology 2002;59:1552-6.
5. Shim SC, Yoo DH, Lee JK, Koh HK, Lee SR, Oh SH, \textit{et al.} Multiple cerebellar infarction due to vertebral artery obstruction and bulbar symptoms associated with vertical subluxation and atlanto-occipital subluxation in ankylosing spondylitis. J Rheumatol 1998;25:2464-8.
6. Panda S, Ravishankar S, Nagaraja D. Bilateral vertebral artery dissection caused by AAD. J Assoc Physicians India 2009;58:187-9.
7. Kulkarni GB, Veerendrakumar M, Nupur P, Hima P, Shailesh M, Advait K. Profile of patients with CV junction anomalies with posterior circulation strokes. J Stroke Cerebrovasc Dis 2014;23:2819-26.
8. Sawlani V, Behari S, Salunke P, Jain VK, Phadke RV. “Stretched loop sign” of the vertebral artery: A predictor of vertebrobasilar insufficiency in AAD. Surg Neurol 2006;66:298-304.
9. Uohara MY, Beslow LA, Billinghurst L, Jones BM, Kessler SK, Licht DJ, \textit{et al.} Incidence of recurrence in posterior circulation childhood arterial ischemic stroke. JAMA Neurol 2017;74:316-23.
10. Gurley KL, Edlow JA. Avoiding misdiagnosis in patients with posterior circulation ischemia. Acad Emerg Med 2019;26:1273-84.
11. Kulkarni GB, Mustare V, Pruthi N, Pendharkar H, Modi S, Kulkarni A. Profile of patients with craniovertebral junction anomalies with posterior circulation strokes. J Stroke Cerebrovasc Dis 2014;23:2819-26.

DOI: 10.4103/aiian.AIAN_744_20

Annals of Indian Academy of Neurology | Volume 24 | Issue 6 | November-December 2021