

Case report

Lower brainstem dysfunction in an infant with persistent primitive trigeminal artery

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Abstract

A 6-month-old boy with persistent primitive trigeminal artery (PPTA) presented with stridor, dysphagia, delayed motor development and postural neck and shoulder dystonia. Magnetic resonance imaging / angiography and ultrasonography revealed PPTA, with flow from the dilated basilar artery to the right internal carotid artery, lower brainstem compression by the dilated basilar artery, and cerebellar vermis hypoplasia. Evoked potentials showed lower pons and medulla oblongata functional disruption. These lesions may be related to vascular etiology in the lower brainstem or to congenital malformation syndrome involving infratentorial structures. The relationship of this condition to Möbius syndrome is discussed.

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1. Introduction

Primitive trigeminal artery (PTA) appears in the developing brain at the 3–5 mm embryo stage, and is one of the embryonic arteries that connect the dorsal aorta (future internal carotid arteries) and the longitudinal neural arteries (future basilar artery) [1]. Persistent PTA (PPTA) is the most common anomaly of the carotid-basilar anastomosis, and is observed in 0.1–0.6% of angiograms [2]. Although most individuals with PPTA remain asymptomatic, some manifest with clinical

symptoms related to cranial nerve root compression by the PPTA [2], or to brainstem ischemia, infarction and intracranial hemorrhage that result from accompanying vascular anomalies and aneurysms [3–5]. A rare case of PPTA that was complicated with lower brainstem dysfunction and lower vermian hypoplasia is described.

2. Case report

A Japanese boy (birth weight, 2924 g; head circumference, 33 cm) with no birth asphyxia was delivered at 40 weeks gestation by cesarean section, resulting from premature membrane rupture. In the neonatal period, feeding difficulty and poor weight gain were noted.

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Upon hospital admission at four months of age, weight, height and head circumference were 4620 g (−3.1 SD), 57.7 cm (−2.6 SD), and 40.0 cm (−1.2 SD), respectively. The auricular upper portion was thin, and funnel chest was present; with retraction augmented in a see-saw respiration pattern. Inspiratory and expiratory stridor was noted, which was aggravated during crying and associated with a gurgle sound in the pharynx. Deep

tendon reflex and muscle tonus at rest were normal, although in the prone position there was prominent retraction of both shoulders and dorsiflexion of the neck. Fisted fingers with adducted thumbs limited grasping and toy holding to only a few seconds. Muscle tonus and motion of lower extremities were normal; as was facial expression, for age. Tracking of moving objects with both eyes and showing interest in surroundings

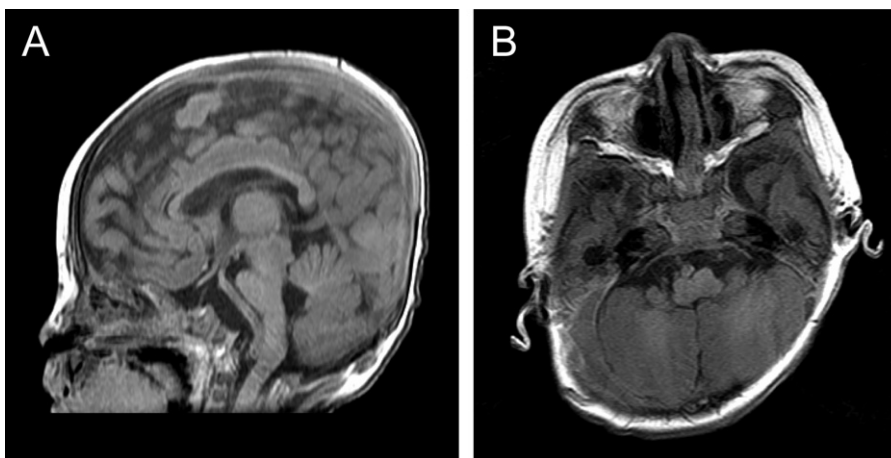


Fig. 1. T1-weighted brain magnetic resonance imaging (A,B). Compression on the lower brainstem by a dilated basilar artery is noted, and the lower half of the cerebellar vermis is absent (A). Medulla oblongata is compressed by the basilar artery, although there is no evidence of vagal nerve root compression (B).

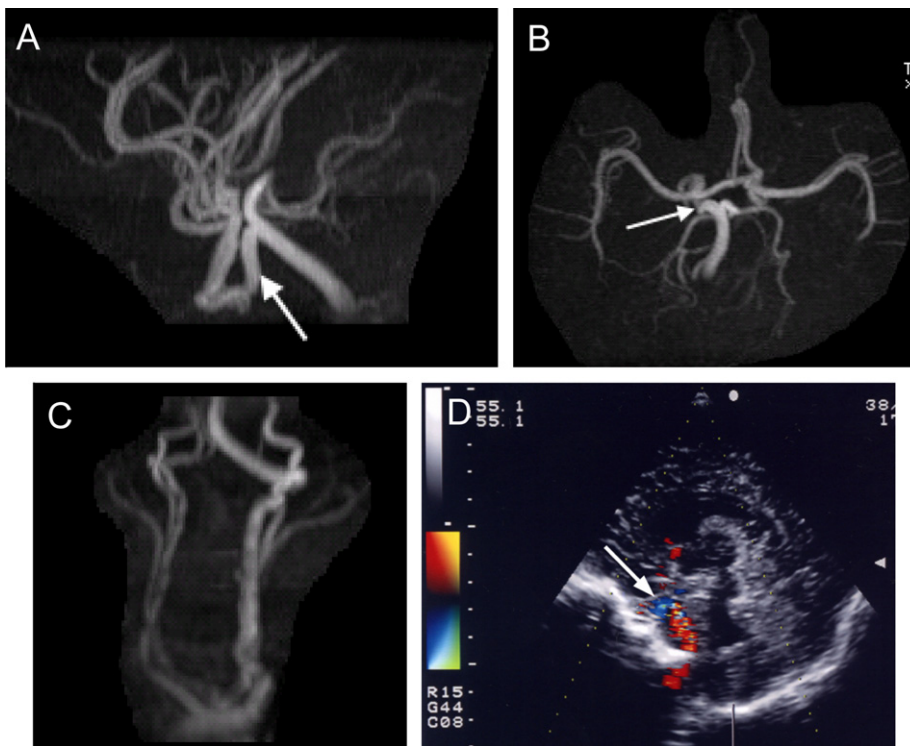


Fig. 2. Magnetic resonance angiography (A–C) and color doppler US (D) of cranial and cervical vessels. The PPTA (A,B,D, arrows) connects the basilar artery and the right internal carotid artery. No aneurysm or arteriovenous malformation was present in the craniocervical vascular system.

was also apparently normal. However, vestibulo-ocular reflex was attenuated and response to auditory stimuli mild to weak. Sternocleidomastoid muscle was not atrophic, and hypertonic due to the cervical dystonia.

Blood gas, lactate, thyroid function, screening tests for metabolic disorders and G-banded chromosomal analysis were normal. Magnetic resonance imaging / angiography revealed a dilated basilar artery (Fig. 1) and the presence of a right PPTA (Figs. 2A and B). Ventral pons and medulla oblongata were compressed and deformed by the dilated basilar artery (Fig. 1A). Both anterior cerebral arteries appeared to originate from the communicating artery in the right internal carotid artery extension (Fig. 2B). Left vertebral artery was dilated (Fig. 2C), although no other abnormalities were observed in the cerebrovascular system.

Lower cerebellar vermis was hypoplastic (Fig. 1B). Cerebral cortex, white matter myelination, basal ganglia and cerebellar hemisphere were normal. Doppler ultrasonography (US) showed a blood flow through the PPTA, from the basilar artery to the right internal carotid artery (Fig. 2D). Electroencephalography was normal for age. Brainstem auditory evoked potentials were attenuated, showing only a trace of wave I and V. Otoacoustic emissions were negative. Median nerve short-latency somatosensory evoked potentials showed a delayed N18 latency with right side stimulation, and an absence of N20 and N30 with left side stimulation. R1 latency was markedly delayed (17.4–18.8 ms; normal 10.1 ± 0.5 ms), and R2 and R2' components were absent on blink reflex. Motor nerve conduction velocity was normal. Chest computed tomography revealed atelectasis in the dorsal lung bilaterally, suggesting the presence of aspiration. Cardiac US showed normal results, and abdominal US showed mildly small right kidney with a 40 mm diameter in the long axis.

After admission, weight gain of 35 g/d was achieved by nasogastric tube feeding. Upper gastrointestinal series showed a normal coordination pattern of oropharyngeal muscles, although the initiation of swallowing reflex appeared diminished. Patient head control improved gradually, but remains incomplete at 6 months of age. He can now eat orally small part of his meal, and the laryngeal stridor has been ameliorated.

3. Discussion

In the present case, PPTA alone cannot account for the clinical symptoms. Basilar artery compression on the ventral brainstem, to a degree that is often encountered in the elderly, can explain the brainstem dysfunction, including the primary involvement of the auditory pathway. Accompanying developmental defects in the central nervous system may be assumed to fully explain the clinical picture of this patient.

Möbius syndrome is characterized by congenital paralysis of the 6th and 7th nerves. Since these motor nuclei are located in the lateral tegmentum of the brainstem, which corresponds to the watershed area, vascular etiology may explain the pathophysiology of this syndrome [6,7]. Such an ischemic lesion during the embryonic period, usually not visible with neuroimaging, can involve the 8th to 10th cranial nerve nuclei, which results in feeding and respiratory symptoms, and auditory disturbance [7,8], similar to that of the present patient. Premature regression of carotid-basilar blood supply, or a delayed basilar artery formation, may be causative in some cases of Möbius syndrome [6]. Thus, it is interesting that Klippel–Feil syndrome, another disease of vertebral-basilar artery disruption, is associated with facial nerve paresis, inner ear developmental defects, and rarely, with PPTA [9]. Ischemic insults during the fetal period, possibly through premature absorption of otic or stapedial arteries, may be related to the otolaryngological abnormalities of Klippel–Feil syndrome [7], in a similar manner to those of the present patient. Thus, the present patient may be an atypical Möbius syndrome subject.

PPTA also accompanies non-vascular malformations, including Chiari malformation type 1, cerebellar cyst, and polycystic kidney [10,11]. Although these might be accidental associations, in the present patient, the dystonia and cerebellar vermis hypoplasia may represent a congenital malformation, where the vascular anomalies and resultant brainstem ischemic lesions are part of its manifestations. The etiology of PPTA in the present patient may contribute to broaden the spectrum of vertebral-basilar artery disruption to a category of malformation syndromes involving the primordial cranial vascular system and other central nervous system structures.

References

- [1] Padgett DH. The development of the cranial arteries in the human embryo. *Contrib Embryol* 1948;32(212):207–61.
- [2] Ikezaki K, Fujii K, Kishikawa T. Persistent primitive trigeminal artery: a possible cause of trigeminal and abducens nerve palsy. *J Neurol Neurosurg Psychiatry* 1989;52:1449–50.
- [3] Stern J, Correll JW, Bryan N. Persistent hypoglossal artery and persistent trigeminal artery presenting with posterior fossa transient ischemic attacks. Report of two cases. *J Neurosurg* 1978;49:614–9.
- [4] Okada Y, Shima T, Nishida M, Yamada T, Yamae K, Okita S, et al. Bilateral persistent trigeminal arteries presenting with brain stem infarction. *Neuroradiology* 1992;34:283–6.
- [5] Ohtakara K, Kuga Y, Murao K, Kojima T, Taki W, Waga S. Posterior fossa arteriovenous malformation associated with persistent primitive trigeminal artery - case report. *Neurol Med Chir (Tokyo)* 2000;40:169–72.
- [6] Bouwes Bavinck JN, Weaver DD. Subclavian artery supply disruption sequence: hypothesis of a vascular etiology for Poland,

- Klippel-Feil, and Möbius anomalies. *Am J Med Genet* 1986;23:903–18.
- [7] Sarnat HB. Watershed infarcts in the fetal and neonatal brainstem. An aetiology of central hypoventilation, dysphagia, Möbius syndrome and micrognathia. *Eur J Paediatr Neurol* 2004;8:71–87.
- [8] Roig M, Gratacòs M, Vazquez E, del Toro M, Foguet A, Ferrer T, et al. Brainstem dysgenesis: report of five patients with congenital hypotonia, multiple cranial nerve involvement, and ocular motor apraxia. *Dev Med Child Neurol* 2003;45:489–93.
- [9] Paksoy Y, Seker M, Kalkan E. Klippel-Feil syndrome associated with persistent trigeminal artery. *Spine* 2004;29:E193–6.
- [10] Oertel M, Kästner S, Winking M, Böker D-K, Deinsberger W. Chiari malformation type I with concomitant persistent trigeminal artery. *Zentralbl Neurochir* 2006;67:30–5.
- [11] Matsumura M, Wada H, Nojiri K. Persistent primitive trigeminal artery, cavum septi pellucidi, and associated cerebral aneurysm in a patient with polycystic kidney disease: case report. *Neurosurgery* 1985;16:395–7.