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Pediatric Cerebellar Infarction Caused by Atlantoaxial Subluxation

—Case Report—

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Abstract

An 11-year-old girl developed cerebellar infarction presenting as a posterior fossa mass lesion after stretching and flexing her neck. Cerebral angiography demonstrated irregular narrowing of the right vertebral artery at the C2 level and x-rays of the upper cervical spine showed atlantoaxial subluxation with os odontoideum. She underwent surgical decompression with removal of infarcted tissue. The cerebellar infarction probably resulted from occlusion of the vertebral artery, followed by brain swelling due to recanalization.

Key words: cerebellar infarction, posterior fossa, atlantoaxial subluxation, vertebral artery, childhood

Introduction

Ischemic stroke in the vertebrobasilar system is rare in children, and pediatric cerebellar infarction presenting as a posterior fossa mass lesion is extremely rare.[4,14,37] Atlantoaxial subluxation was the cause of a few cases of pediatric cerebellar infarction.[21,41] Here, we report an 11-year-old girl with cerebellar infarction and acute hydrocephalus due to atlantoaxial subluxation.

Case Report

An 11-year-old girl was admitted to our hospital on June 15, 1990 with complaints of headache and slowly progressive gait disturbance. Seven days before admission, she first experienced morning headache and nuchal pain. Her mother reported that she had been fatigued for several days since she acted as a model in her art class, holding poses with flexion or extension of her neck. At first, we did not consider this statement was related to her condition.

On admission, she was alert, well oriented, but appeared to be tired. Neurological examination revealed ataxia of the right upper extremity. She complained of unsteadiness but her gait was not ataxic. Her blood pressure was 120/70 mmHg, and her pulse rate was 66/min and regular. Her body temperature was 37.2°C and respiration was normal. Laboratory tests were normal; prothrombin time and partial thromboplastin time were within normal limits. Computed tomography (CT) revealed a hypodense lesion

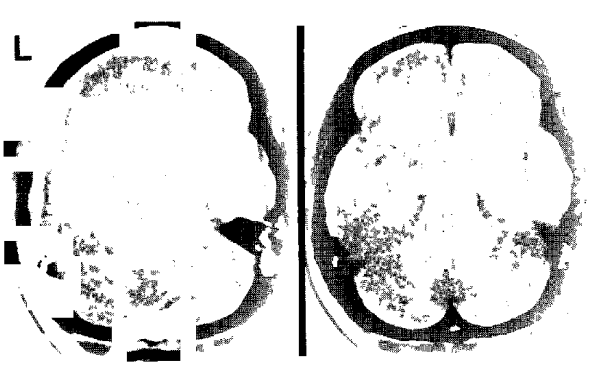

Fig. 1 CT scans on admission, showing a hypodense lesion in the right cerebellar hemisphere (*left*), not enhanced postcontrast (*right*).

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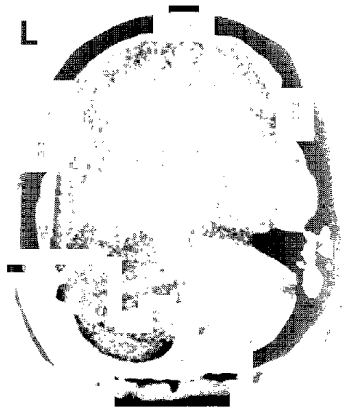

Fig. 2 CT scan 1 day after admission, showing a large hypodense mass lesion in the right cerebellar hemisphere with displacement of the fourth ventricle.

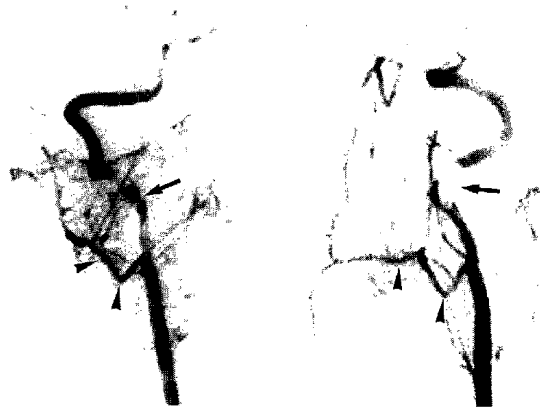

Fig. 3 Right vertebral angiograms, anteroposterior (*left*) and lateral views (*right*), showing irregular narrowing of the right vertebral artery at the C2 level (*arrow*) with a muscular collateral artery (*arrowheads*).

in the right cerebellar hemisphere with no enhancement postcontrast (Fig. 1). A lumbar puncture was performed, and the cerebrospinal fluid analysis was within normal limits. Cerebellar infarction was diagnosed and cerebral angiography planned for 2 days later. However, on the next day her level of consciousness deteriorated rapidly and she became semicomatose. Immediate CT revealed a large hypodense mass lesion in the right cerebellar hemisphere with displacement of the fourth ventricle (Fig. 2).

Suboccipital craniectomy was performed following ventricular drainage. The surface of the right cerebellar hemisphere appeared to be swollen and pale. Petechial hemorrhage was also seen on the upper part of the cerebellum. Softened cerebellar tissue, which was considered to be an infarction, was removed. On the second postoperative day, she became alert and oriented.

Further examinations were performed to identify the cause of the infarction. The electrocardiogram was normal and the echocardiogram revealed no embolic source. Cerebral angiography demonstrated a normal left vertebral artery but the right vertebral artery irregularly narrowed at the C2 level with delayed flow (Fig. 3). This stenotic lesion like an arteriosclerosis was considered to be the cause of the infarction, but the reason for this lesion in a pediatric patient was still unknown. X-rays of the upper cervical spine showed atlantoaxial subluxation with os odontoideum (Fig. 4). The canal diameters at the C1 level on flexion and extension of the neck were 10 and 15 mm, respectively. The instability index was 33%. CT also demonstrated the atlantoaxial

Fig. 4 X-ray film of the upper cervical spine, showing atlantoaxial subluxation with os odontoideum (*arrow*).

Fig. 5 CT scan at the C1 level, showing separation of the odontoid.

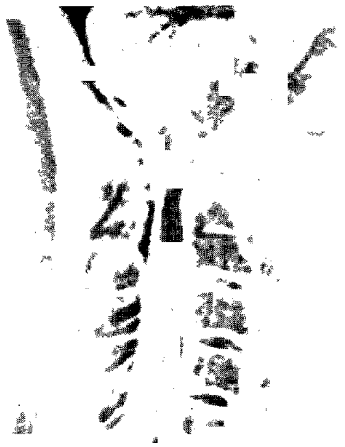

Fig. 6 Sagittal MR image, showing atrophy of the cord and disappearance of subarachnoid space at the C1-2 levels.

subluxation with os odontoideum (Fig. 5). A sagittal cervical magnetic resonance (MR) image showed narrowing of the spinal canal and disappearance of subarachnoid space at the C1-2 levels (Fig. 6).

Surgical treatment for the atlantoaxial subluxation was recommended but she and her parents refused surgery. Her neurological symptoms resolved and she returned to school with her neck immobilized in a soft collar.

Discussion

Only 49 cases of pediatric ischemic stroke in the vertebrobasilar system have been reported[2-43] (Table 1). The patients were aged from 4 months to 19 years old, without a particular age distribution. There were 42 males, six females, and one unknown, showing a strong predominance of males. The cause of the vertebrobasilar occlusion was unknown in 22 of the 49 cases. Blunt head trauma was thought to be the cause in five patients,[22,24,27,42] infection such as sepsis and bacterial endocarditis in four,[12,19,35,40] and congenital vascular anomalies in four.[7,34,36,38] Other suspected causes included congenital heart disease,[13] arteritis,[25] migraine,[9,14] fibromuscular dysplasia,[37] Duchenne muscular dystrophy,[26] chiroplastic manipulation,[43] spontaneous neck rotation,[4] upper extremity injury,[32] and surgery.[5] Atlantoaxial subluxation causing infarction occurred in only three patients, including ours.[21,41] The site of the occlusion/stenosis was the basilar artery in 22 cases, vertebral artery in 12, and both vertebral and basilar arteries in nine. Occlusion/stenosis of the vertebral artery frequently occurred at the C1-2 levels, suggesting that the vertebral arteries at the C1-2 levels tend to be injured or occluded by flexion, extension, or rotation of the neck associated with head trauma or chiropractic manipulation.[4,21,22] A congenital vascular anomaly or cervical spine anomaly would make the vertebral arteries at that site more vulnerable.

Table 1 Summary of 49 reported cases of vertebrobasilar artery occlusion in childhood

Age	4 mos-19 yrs
Sex	
male	42 cases
female	6 (+ present case)
unknown	1
Cause of occlusion	
blunt head trauma	5 cases
infection	4
congenital vascular anomaly	4
atlantoaxial subluxation	2 (+ present case)
migraine	2
congenital heart disease	1
spontaneous neck rotation	1
fibromuscular dysplasia	1
Duchenne muscular dystrophy	1
chiropractic manipulation	1
arteritis	1
upper extremity injury	1
surgery	1
unknown	22
not stated	2
Site of stenosis/occlusion	
basilar artery	22 cases
vertebral artery*	12 (+ present case)
basilar and vertebral arteries*	9
unknown	6

*Occlusion/stenosis of the vertebral artery frequently occurred at the C1-2 levels.

Cervical spine anomalies can cause cerebellar infarction in children. These anomalies are sometimes asymptomatic, and occasionally become symptomatic after trauma, chiropractic manipulation, and others. In our patient the onset occurred several days after she had bent or stretched her neck at school, indicating a cervical spine anomaly as a cause of the cerebellar infarction. X-rays of the upper cervical spine showed atlantoaxial subluxation. Cerebral angiography demonstrated irregular narrowing of the right vertebral artery at the C2 level with delayed flow. The initial CT scan revealed a hypodense lesion in the cerebellar hemisphere, not enhanced postcontrast, while a second CT scan, performed when her level of consciousness deterio-

rated, revealed a large hypodense mass lesion in the same side of the cerebellum. This evidence suggests that the cerebellar infarction resulted from the intimal injury and the occlusion associated with stretching or compression of the vertebral artery, and subsequent recanalization of the occluded vertebral artery caused the cerebellar swelling.

Fifteen of the 49 cases of pediatric ischemic stroke in the vertebrobasilar system were diagnosed as cerebellar infarction. The cerebellar infarction presented as a posterior fossa mass lesion in seven cases,[4-6,11,14,29,37] all of which were treated surgically. Internal decompression was performed in three cases,[6,11,29] ventricular drainage in two,[5,14] shunt operation with external decompression in one,[37] and shunt operation only in one.[4] Surgical treatment for cerebellar infarction presenting as a posterior fossa mass lesion is still controversial. Only ventricular drainage may be an adequate treatment in some patients,[4,5,14] but we routinely perform posterior fossa decompression with removal of the infarcted tissue to relieve brainstem compression.

The atlantoaxial subluxation in our patient was definitely considered to require surgical treatment because it had caused cerebellar infarction and the instability index was over 20%,[1] but the patient and her parents refused surgery. Fortunately, 3 years after the onset, no recurrent stroke or neurological symptoms have developed.

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