Hindbrain Stroke in Children Caused by Extracranial Vertebral Artery Trauma

BY RICHARD A. R. FRASER, M.D., F.R.C.S.(C),* AND SEYMOUR M. ZIMBLER, M.D., F.A.C.S.t

Abstract:
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• Hindbrain transient ischemic attacks (TIAs) culminating in posterior circulation stroke are described in five children. Atlanto-axial subluxation and angiographical documentation of Cl to C2 level arterial pathology are documented in one patient. Four additional patients with nearly identical clinical presentations, posterior fossa TIAs, stroke and basilar angiographical pathology are reviewed. A mechanical traumatic etiology is suggested.

Unexplained transient repeated brain stem and/or cerebellar symptomatology may be due to extracranial vertebral artery stenosis or occlusion by atlanto-axial instability. After appropriate documentation, stabilization may prevent further TIAs or strokes.

Additional Key Words
vertebrobasilar occlusion transient ischemic attack atlanto-axial subluxation extracranial cerebrovascular disease cervical fusion

Case Report
A six-year-old right-handed boy was first admitted to the Boston Floating Hospital because of gait ataxia and clumsiness of the right upper extremity. During the previous month, he had experienced intermittent nausea, headache, frequent vomiting, dysarthric speech, and clumsiness of the right upper extremity. These symptoms would appear for a day, improve spontaneously, and reappear two to three days later. Because of these symptoms, he had been admitted to another hospital where neurological examination, EEG, brain scan, skull x-rays, and CSF formula were reported as normal.

At the time of admission to the Boston Floating Hospital, examination revealed a mild deficit with minor clumsiness of the right hand, bradykinesia, and slow speech. The diagnoses suggested included cerebellar ataxia of infectious or postinfectious etiology. A brain scan (technetium 99) was normal. Skull x-rays on this admission were reported as normal, but closer examination revealed nonfusion of the odontoid and anterior displacement of Cl on C2. While this finding was noted and confirmed on cervical spine tomograms (fig. 1) and flexion and extension films (figs. 2 and 3), its significance and relationship to the current illness were not appreciated and the patient was discharged without cervical stabilization.

One day following discharge, the patient awoke with a mild right hemiparesis which, over the ensuing six hours, evolved into a flaccid hemiplegia and prompted his readmission to the Boston Floating Hospital. His parents stated that he had fallen from bed the night before. Examination on the second admission confirmed severely dysarthric speech, inability to gaze to the left of the midline, nystagmus on right lateral gaze, lower motor neuron weakness of the left face, and a virtually complete right hemiplegia. Deep tendon reflexes were absent in the right extremities and a right-sided extensor plantar response was elicited. Dysmetria was apparent in the left upper extremity. Laboratory studies, including hemogram, white blood count, serum electrolytes, and urinalysis, were normal.

For many years, neurosurgeons and neurologists, when evaluating patients with cervical spine trauma or congenital anomalies of the cervical spine, have emphasized the direct compressive effects of such lesions on the cervical spinal cord. Occasional patients with cervical spine trauma and injury to the vertebral arteries with subsequent brain stem and cerebellar dysfunction have been reported. All of these patients have been adults. Additional patients have had posterior fossa stroke symptoms following chiropractic cervical spine manipulation.

Occlusive cerebrovascular disease in children is uncommon. The overwhelming majority of such patients display lesions in the carotid circulation. Only seven children have been reported previously with disease in the posterior cerebral circulation. Septic emboli occurred in two, while no clearly defined etiology could be identified in the remaining five (table 1).

We wish to describe an additional patient, a six-year-old boy with a history of vertebrobasilar TIAs, which culminated in a major posterior circulation stroke. Cervical spine x-rays and angiographical data lead us to suggest that the cerebrovascular accident in this patient was the result of atlanto-axial subluxation, traumatic deformation of the vertebral arteries and distal embolization to the basilar artery. Clinical and angiographical similarities in other patients suggest a common syndrome.

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HOSPITAL COURSE

The following day, four-vessel angiography was performed, prompted by a diagnostic impression of "arteritis." The presence of atlanto-axial subluxation was not communicated to the neuroradiologist so that the study was performed in the usual hyperflexed position, causing maximum anterior displacement of the atlas. The arteriogram revealed occlusion of the right vertebral artery at the level of C2. Collateral vessels were seen at this level bypassing the occluded portion of the vertebral artery and rejoining the vessel at the rostral border of C2. The right vertebral artery terminated in the posterior interior cerebellar artery without opacification of the basilar artery (fig. 4). The left vertebral artery demonstrated aneurysmal dilatation at C2. This vessel also terminated in the posterior interior cerebellar artery without filling of the basilar artery (fig. 5). Carotid injections opacified the rostral tip of the basilar artery and both superior cerebellar arteries and posterior cerebral arteries.

Neurosurgical consultation was again requested. It was our impression that the arterial pathology revealed by angiography was the result of mechanical trauma to each vertebral artery as it crossed the transverse foramen of C2. The occlusion of the right vertebral artery and the aneurysmal dilatation of the left was thought to result from chronic intermittent mechanical trauma and stretching of each vessel by the anterior displacement of C1 upon C2. A presumptive diagnosis of a posterior circulation stroke was suggested.

It was decided to stabilize the patient externally in halotraction and await maturation of his neurological deficit. One week after admission, he was able to flex and extend the right leg against gravity and demonstrated a flicker of movement in the right toes. Functional improvement occurred in conjunction with active physiotherapy.

Neurological improvement continued for another four weeks, at which time the patient was ambulatory with a right leg brace and demonstrated proximal movement against gravity in the right upper extremity. The right hand, however, remained plegic. Eye movements were normal. Four-vessel angiography was repeated at this time and revealed no change in the vertebral artery anomalies. These films did demonstrate, however, that the basilar artery had recanalized with filling of the superior cerebellar and posterior cerebral arteries from the posterior circulation. Slight narrowing of the distal basilar artery just proximal to the origin of the superior cerebellar arteries remained (figs. 6 and 7).

At this time, a cervical spine fusion of C1 to C3 using wire and iliac bone was performed. The patient was discharged on the tenth postoperative day after an uneventful convalescence. Follow-up at three months revealed little improvement with persistent severe right hemiparesis, most marked in the right upper extremity.

Discussion

Only seven children have been previously reported with occlusive or stenotic pathology of the vertebrobasilar system. Review of the case material (table 1) reveals that in five of these patients
no apparent cause for the vascular pathology could be assigned and all were designated as either a congenital or idiopathic disorder. The remaining two patients had septicemia and obvious septic emboli to many vessels, including the intracranial circulation. Closer attention to this table reveals that four of the remaining five presented with syndromes strikingly similar to the present case. All were male. Intermittent attacks of vertigo, ataxia, and sensory motor disturbances, presumably posterior circulation TIAs, were followed after weeks or months by a sudden stroke-like syndrome of hemiparesis and ataxia. In case 4, a severe fall and a documented occipital fracture preceded the permanent stroke-like syndrome by a month. There was no antecedent trauma in case 3 or 5, but the patient described in case 7 experienced the onset of sudden hemiparesis while walking home from baseball practice where cervical trauma might have occurred. In each of these patients, arteriography revealed stenosis or occlusion of the vertebral arteries of one or both sides at the level of the second cervical vertebra. In addition, incomplete filling of the basilar artery was
Right vertebral angiogram four weeks later — lateral view. Vertebral artery unchanged. Basilar artery now opacified, but narrow. Marked stenosis of basilar artery just proximal to origin of superior cerebellar artery.

It seems apparent that our case is a clear example of a posterior circulation stroke. Serial angiograms revealed an occluded basilar artery which later recanalized concomitant with minor improvement in the patient's neurological deficit. (Anticoagulants were not administered at any time.) We believe that the most reasonable explanation for this vertebrobasilar arterial lesion is traumatic and that repeated anterior subluxation of Cl on C2 caused mechanical deformation and occlusion of both vertebral arteries. Thrombus formation in the deformed vertebral arteries with distal embolization into the basilar artery presumably caused the stroke in our patient. Though no mention was made of cervical spine anomalies in four previously reported patients with almost identical findings, we wonder whether a similar mechanism was playing an etiological role.

It is possible, even in the absence of bony anomalies, that mechanical distortion of the vertebral arteries may result from subluxation secondary to laxity of the ligaments connecting the atlas and axis. In this instance, no radiographical changes would be apparent except on flexion and extension films.

Atlanto-axial subluxation with non-union of the odontoid is a frequent congenital anomaly of the spinal column. Previous series of patients with atlanto-axial anomalies do not contain a single example of angiographically documented vertebrobasilar pathology. Ford has described a 17-year-old boy similar to the case reported here. Recurrent posterior circulation TIAs were eventually diagnosed after atlanto-axial instability was discovered and treated by cervical fusion. Unfortunately, this case lacks angiographical documentation. The patient's symptoms were recurrent, transient, and closely resembling those reviewed above; they seem a convincing, if not proved, example of this syndrome.

Several patients presenting with brain stem and/or cerebellar deficits due to stroke have been reported following cervical spine trauma and an even larger number following chiropractic neck manipulation. In two patients of the latter group, angiography confirmed vertebral artery lesions at the level of the axis, similar to those described above.

Schneider and Crosby have suggested that the vertebral arteries are especially vulnerable to compression at three sites: (1) at any fracture dislocation above C6, (2) at the atlanto-axial level with Cl to C2 compression, and (3) at Cl because of atlanto-axial...
dislocation resulting from the occipital condyles sliding forward over the articular facets of Cl. Arteriograms in the present case, in two post-neck manipulation cases,\textsuperscript{12} and in four children without antecedent trauma\textsuperscript{8,19,21} revealed similar Cl to C2 arterial pathology, and confirm the potential vulnerability of the vertebral artery at this level.

Our patient was treated by fusion of the first three cervical vertebrae when his neurological condition stabilized. This procedure seemed warranted to prevent future additional mechanical trauma to the vertebral arteries and further posterior circulation strokes. Potential cervical cord compression was also prevented.

A recent report attempts to define angiographically prognostic indicators in patients with basilar artery occlusion.\textsuperscript{30} These authors point out that such patients present in two ways: (1) with abrupt onset of coma, often leading to death, or (2) with transient symptoms of brain stem ischemia. The latter patients are a diagnostic problem. These authors also suggest, not surprisingly, that small or absent collaterals from the anterior circulation carry a poor prognosis. Basilar artery occlusion has been regarded in the past as nearly always fatal.\textsuperscript{31} More recent reports,\textsuperscript{30,32} however, suggest that survival is the rule.

Increased awareness of the syndrome described in this report might lead to documentation of similar pathology in patients with otherwise unexplained repeated transient brain stem and cerebellar symptomatology. Appropriate x-ray studies, including flexion and extension cervical spine films and vertebral arteriography, are urged in such cases. These patients represent an additional syndrome of extracranial occlusive cerebrovascular disease, one in which surgical stabilization provides appropriate therapy and removes further risk of stroke.

Summary

A patient is described with bilateral vertebral artery anomalies at Cl to C2, with occlusion of the basilar artery occurring in conjunction with a posterior circulation stroke. The anomalies are believed to be the result of chronic and repeated atlanto-axial subluxation.

Review of the literature provides four previous patients with strikingly similar clinical presentations. All were male and all presented with intermittent symptoms suggestive of brain stem and/or cerebellar dysfunction. All were retrospectively diagnosed as experiencing posterior cerebral circulation transient ischemic attacks, which culminated in a stroke with varying degrees of static neurological deficit. Patients with severe atlanto-axial instability are at risk for posterior circulation injury and possible brain stem stroke. Vertebral artery angiography should be considered in such patients.

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