Cerebellar Infarction From Fibromuscular Dysplasia and Dissecting Aneurysm of the Vertebral Artery

Report of a Child

A. Perez-Higueras, MD, F. Alvarez-Ruiz, MD, A. Martinez-Bermejo, MD, R. Frutos, MD, O. Villar, MD, and E. Diez-Tejedor, MD, PhD

Infarction in the vertebrobasilar system presenting as a posterior fossa mass lesion is extremely rare in children. Of only 21 cases published in the literature, the cause was unknown in half. The etiologic factors in the others were trauma in four cases, infection in three cases, and dehydration, cervical spine anomaly, and basilar artery migraine in each case each. A congenital vascular anomaly was implicated only once.1

We recently treated a 9-year-old boy with cerebellar infarct produced by Type I fibromuscular dysplasia (FMD). Angiography confirmed this diagnosis and revealed a dissecting aneurysm. To our knowledge this is the first case of this type reported in the literature. (Stroke 1988; 19:521–524)

Case Report

This 9-year-old boy was well until July 15, 1986, when he had a 10-minute episode of loss of consciousness, ocular deviation, and myoclonus in the upper extremities. Similar episodes accompanied by vomiting were repeated 2 and 4 days later. He was admitted to a local hospital on July 21, 1986, when computed tomography (CT scan) was performed and he was transferred to our hospital. His medical history was unremarkable. His 36-year-old mother had suffered from convulsive seizures between the ages of 10 and 20 years.

The findings of a general physical examination were normal. Neurologic examination revealed a somnolent boy who opened his eyes and responded to simple commands when aroused. He had weakness of the right lateral rectus muscle and no papilledema; the other cranial nerves were normal. His motor function showed general hypotonia, hyporeflexia, and flexor plantar responses. His gait was ataxic.

CT scan performed at the local hospital (Figure 1) demonstrated a low-density mass lesion in the right cerebellar hemisphere with displacement of the fourth ventricle and obstructive hydrocephalus.

The patient was operated on, and a medium-pressure ventricular–peritoneal unishunt was placed to alleviate the hydrocephalus.

His level of consciousness improved, and neurologic examination showed an alert patient with improved strength of the lateral rectus muscle. Dysmetria of his right limbs was evident.

Repeat CT scan 2 days after ventricular shunting showed no hydrocephalus, but the right cerebellar mass was still seen. There was no enhancement with injection of contrast material except along the folia (Figure 2).

Because the patient continued with his cerebellar symptoms and because the hypodense lesion remained on CT scan, a suboccipital craniectomy was performed and exploration of the right cerebellar hemisphere was undertaken. Swollen white matter without tumoral tissue was found. Several tissue samples were taken and pathologic study confirmed the existence of a cerebellar infarct. To discover the origin of the infarction, angiography of both carotid and vertebral arteries was performed.

Left vertebral angiography (Figure 3) demonstrated a “string-of-beads” lesion in the segment between C1 and C2, with a dissecting aneurysm just below. No other angiographic alteration was noted in the remainder of the vessels.

The patient did well, and neurologic examination 6 months after his discharge showed a moderate right cerebellar syndrome. He was being treated with physical therapy.

Discussion

Posterior fossa infarction in children is rare. Only 21 cases have been published in the literature1 and of these cases the lesion was limited to a cerebellar hemisphere.

From the Departments of Neuroradiology, Neurology, and Neurosurgery, Hospital La Paz and Faculty of Medicine, Autonomous University, Madrid, Spain.

Address for correspondence: Antonio Perez-Higueras, C) Rosas de Aravaca Numero 16, 28023 Aravaca, Madrid, Spain.

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Vascular obstructive disease in children is less frequent in the vertebrobasilar system than in the anterior circulatory system (carotid territory). The etiologic or predisposing factors causing the obstruction vary little between the two systems. General factors, such as congenital heart disease, metabolic disorders, hypertension, vascular malformation, and/or dehydration, affect the two territories with the same frequency. Other factors are more specific to one or the other territory. Cervical trauma is the most common cause of obstruction in the posterior circulation, and infection, especially nasopharyngeal infection, is the most frequent cause of occlusion in the anterior circulation. Migraine has been also described as a cause of posterior circulation obstruction. FMD affects the carotid arteries much more frequently than the vertebrobasilar system.

The clinical syndrome associated with acute cerebellar infarction gives essentially the same general neurologic picture in almost all cases; that is, a severely ill patient with signs of a posterior fossa mass. The symptoms, in order of frequency, are headache, dizziness, nausea and vomiting, disorientation, and drowsiness. Seizures, with loss of consciousness, are also frequent.

In most instances, the clinical history helps the examiner to distinguish between an infarct and a tumor in only four, with no damage to the brainstem occurring. CT scan in the acute phase of a recent cerebellar infarction discloses a low-density, smooth-bordered lesion that collapses the fourth ventricle and produces obstructive hydrocephalus. Such lesions do not enhance after contrast injection. In this phase there is great difficulty in differentiating a neoplasm from a cerebellar infarct. To distinguish between them, it is necessary to perform serial CT scans. In some cerebellar infarcts increased attenuation after contrast infusion can be observed 1–4 weeks after onset.

In the two cases of Nobuhiko, contrast CT scans 15 and 11 days after onset showed increased attenuation along the folia corresponding to the distribution of the superior cerebellar artery. This picture is pathognostic for a cerebellar infarct but it is also similar to that of an arteriovenous malformation, as well as of leptomeningeal dissemination from a tumor or from meningitis.

In our case sequential CT scans showed a low-density lesion with mass effect at first; 2 days later the lesion was more sharply differentiated, with a lightened mass effect, and did not enhance with contrast infusion, except along the folia.

There is disagreement as to the best neurosurgical management of a cerebellar infarct that presents as a...
posterior fossa mass with acute obstructive hydrocephalus. Some authors advocate immediate decompression of the posterior fossa with resection of the infarcted cerebellum, whereas others have suggested that ventricular drainage may be adequate treatment.

In our case Type I FMD in the left vertebral artery appears to be the cause of the cerebellar infarct. FMD was first described in a renal artery in 1938 by Leadbetter and Burkland, and cranial artery involvement was first reported by Pabulinskas and Ripley in 1964 on the basis of radiologic evidence and later histologically confirmed by Connet and Lansche.

FMD is a segmental nonatheromatous stenotic disease involving small to medium-sized arteries. The most common radiographic finding is the so-called "string of beads" appearance created by a formation of a chain of arterial dilations that are separated from one another by localized strictures. The dilated areas are always wider than the normal lumen, and the strictures usually narrow the normal lumen by <40%.

A second, less common, roentgenographic pattern is unifocal or multifocal tubular stenosis (Type II). The angiographic differential diagnosis includes Takayasu's arteritis, which involves the aortic arch and the proximal segments of its branches. FMD characteristically spares the origins and proximal segments of vessels.

A third angiographic type of FMD has been termed atypical by Houser et al. One wall of the affected segment is spared, and on the opposite wall an elongated, ovoid diverticulum, sharply circumscribed at either end by a noncircumferential narrowing, resembles a localized FMD constriction.

In the review by Mettinger and Ericson the internal carotid arteries, especially the extracranial parts, were the most common site (90%) of cervicocranial FMD. Involvement was bilateral in 60% of those in whom both carotid arteries were examined. In this series including 52 patients with FMD involvement of the cervicocephalic arteries, seven had a vertebral artery lesion. Healton found that the vertebral artery was affected in 12% of the cases. The disease was localized to the portion of the vertebral artery between the posterior arch of the atlas and the third cervical segment.

The occurrence of FMD in children is rare. Andersen reported two cases, one of whom was the first report of FMD of the vertebral arteries. That child had lesions in many vessels, including both the carotid and vertebral arteries as well as the renal and mesenteric arteries.

Our case is probably the first published with FMD affecting only a single artery, a vertebral artery. Spontaneous dissection is a recognized FMD complication. However, only a few cases have been reported of FMD in the internal carotid artery, and most of these without histologic proof. Arterial dissection has also been described associated with vertebrobasilar FMD. Hirsch and Rasmusen reported a child with dissection of the basilar artery.

In our case, angiography of the left vertebral artery showed a dissecting aneurysm on the anterior wall of the artery. The dissection began 1.5 cm below the lesion. The lumen of the artery was stenosed by approximately 50%. The cerebellar infarction in this case was probably due to embolism arising within the FMD lesion or its complicating dissecting aneurysm.

References

KEY WORDS • aneurysm, dissecting • cerebral infarction • child • fibromuscular dysplasia
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