

## Giant Arteriovenous Aneurysm of the Posterior Fossa in a Three-Month-Old Infant

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**Key Words.** Angiography · Arteriovenous aneurysm · Computed tomography · Hydrocephalus · Posterior fossa tumors · Thrombosis

**Abstract.** A 3-month-old female infant presented a 20-day history of drowsiness and maturing deterioration. The neurological exam showed peripheric facial palsy on the right side and a brachioocral hemiparesis on the left. A brain scan revealed a lesion of avascular content in the posterior fossa. The computed tomography scan showed hydrocephalus and a high-density lesion, nonenhanced after introduction of the contrast agent, compatible with a clot at posterior fossa level. Likewise, after contrast new images appeared which had not been seen previously, considered as afferent and efferent vascular elements to the lesion. The cerebral angiography showed an avascular lesion in the cerebellar vermis with important hypertrophy of arterial and venous elements, although no steal phenomena of neighbouring areas was evident. These findings, and the absence of cardiac failure, suggested the diagnosis, confirmed by surgery, of giant clotted arteriovenous malformation. The patient has done well postoperatively.

### Introduction

Vascular malformations are one of the most interesting problems in neurosurgery. The polymorfism of its clinical pattern, the need for careful programming and analysis of the diagnostic procedures and the frequent complexity of its surgical treatment, require the neurosurgeon's maximum judgement and skill. Classically, arteriovenous malformations are divided into angiomatous and fistulous types, which depends on whether or not they disclose a capillary network between the arterial and venous territories. The fistulous malformations which are less frequent than the angiomatous ones [2, 3] can be of traumatic, infectious or congenital origin [12].



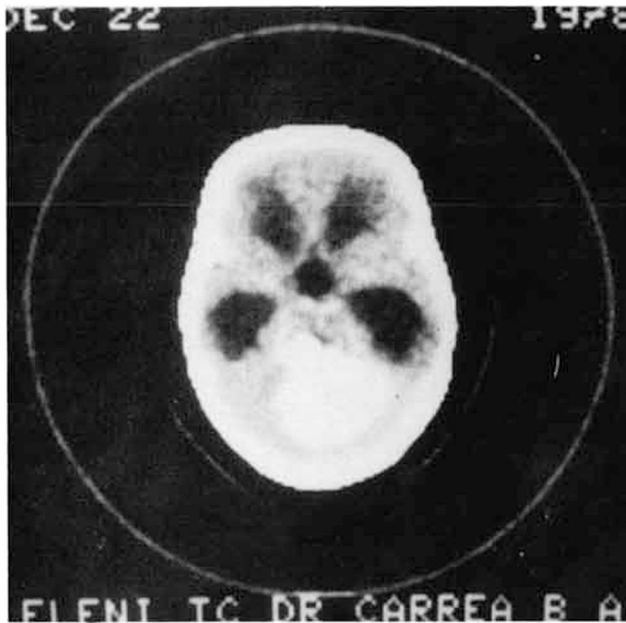
**Fig. 1.** Initial uncontrasted CT scan showing a high-density mass in the vermis and both cerebellar hemispheres.

While carotid-cavernous fistulas are the most common in adults, fistulas of the vertebral or vertebral and carotid arteries with the vein of Galen are the most common in children. Less frequent presentations are internal carotid, vertebral or superficial temporal artery fistulas with their satellite veins at extracranial level or abnormal communications between branches of the internal carotid, external carotid or vertebral artery at intracranial level, with superficial or deep cerebral veins or with dural sinuses.

An unusual case of intracranial hypertension is presented here, produced by a giant arteriovenous aneurysm in the posterior fossa, which was completely clotted. The interesting neurological findings are discussed and comments on the good results obtained by surgery are made.

### **Case Report**

Our patient is a 3-month-old girl who refused feeding and presented drowsiness 20 days before admission. A few days later her parents noticed lagophthalmos on the right and deviation to the left of the bucal commissure. The neurological examination revealed a drowsy



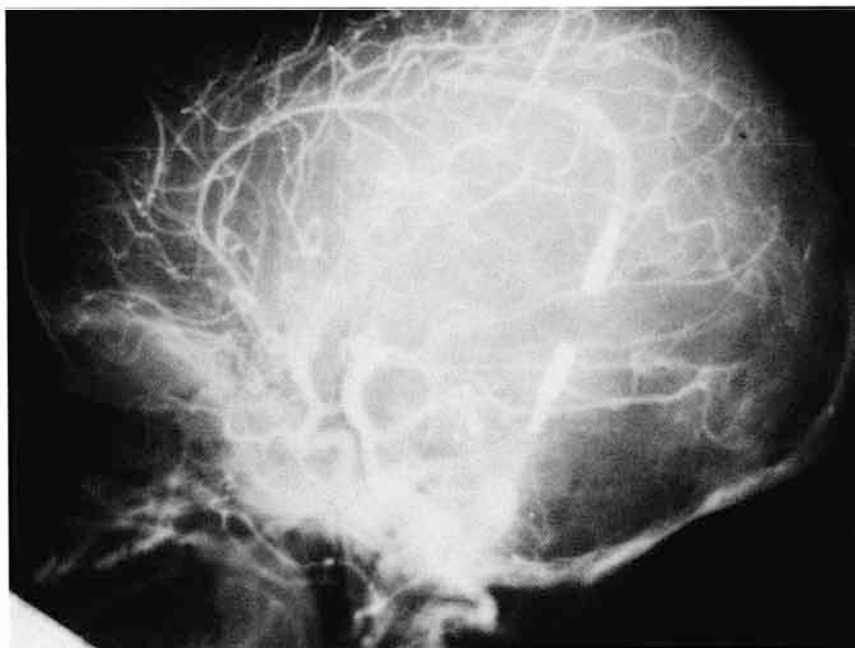
**Fig. 2.** Contrast-enhanced CT scan discloses a vascular area at the anterior part of the mass.

patient with slight retardation, inability to hold her head up or to follow objects with her eyes. Head circumference was 43 cm, which was normal for her age; normotense fontanelle, right peripheric facial palsy and minimal left brachiorural hemiparesis. Discs were normal. Negative cranial auscultation and normal cardiovascular and respiratory exam.

The skull X-ray revealed incipient splitting of sutures. The EEG was normal. A brain scan with technetium-99 showed a tumoral mass in the posterior fossa with high uptake in the periphery showing an annular shaped image. The uncontrasted computed tomography (CT) scan (fig. 1) showed a great tumoral mass in the vermis and both cerebellar hemispheres, more extended towards the right, hyperdense, 21–44 EMI units, with no perilesional edema. The contrast substance (fig. 2) caused the appearance of apparently vascular images which were not visible in the noncontrasted exam, in the front and left part of the tumor, which did not modify its initial densities. An important hydrocephalus secondary to obstruction of the fourth ventricle was also observed.

A right ventricular-peritoneal shunt was inserted at first, to take care of the hydrocephalus, to which the patient's depression was attributed. Despite this shunt procedure, the patient did not improve and some days later developed respiratory and swallowing difficulties attributed to the fact that the low cranial nerves had been affected.

Owing to the unusual tomographic patterns of this lesion a cerebral angiography by brachial route was done (fig. 3, 4) which revealed the ventricular dilation and an expanded avascular lesion in the posterior fossa with important hypertrophy of the vertebrobasilar and right superior cerebellar arteries, which appeared to flow into a vascular lake which is already



**Fig. 3.** Cerebral angiography shows an avascular cerebellar mass with hypertrophy of the vertebrobasilar and right superior cerebellar artery.

observed in the arterial phase and still persists in the venous phase, situated in the foremost and lower part of the avascular area. The vascular drainage was done via a very dilated vein ending in the vein of Galen. Surgery was done with the patient in brow down position and a midline approach of the posterior fossa, which allowed the total removal of a large aneurismatic sac; thick-walled, nonpulsating, measuring approximately  $4 \times 3 \times 2$  cm, and completely thrombosed (fig. 5, 6). As a first step the right superior cerebellar artery, of about 3–4 mm in diameter, which ran to the forward right face of the sac, was clipped, and two efferent thick veins were ligated: one can be observed in the preoperative angiography, and another not revealed in that study which ran horizontally towards the left, to the lateral sinus. The pathology revealed an afferent artery and efferent veins of normal appearance, although they showed hypertrophied walls, whilst the aneurismatic sac presented the replacement of the vascular normal histo-architecture by a connective wall. The intraluminal clot was of the mixed type although there was fibrinoid predominance and no evidence of organization.

The patient evolved well in spite of a difficult postoperative period. The control angiography done selectively by femoral route 3 months later revealed the disappearance of the avascular area, better irrigation of the cerebellar parenchyma compared with the preoperative study, the persistence of the hypertrophied vertebrobasilar system, the partial involution of the afferent artery and the total involution of the efferent veins.

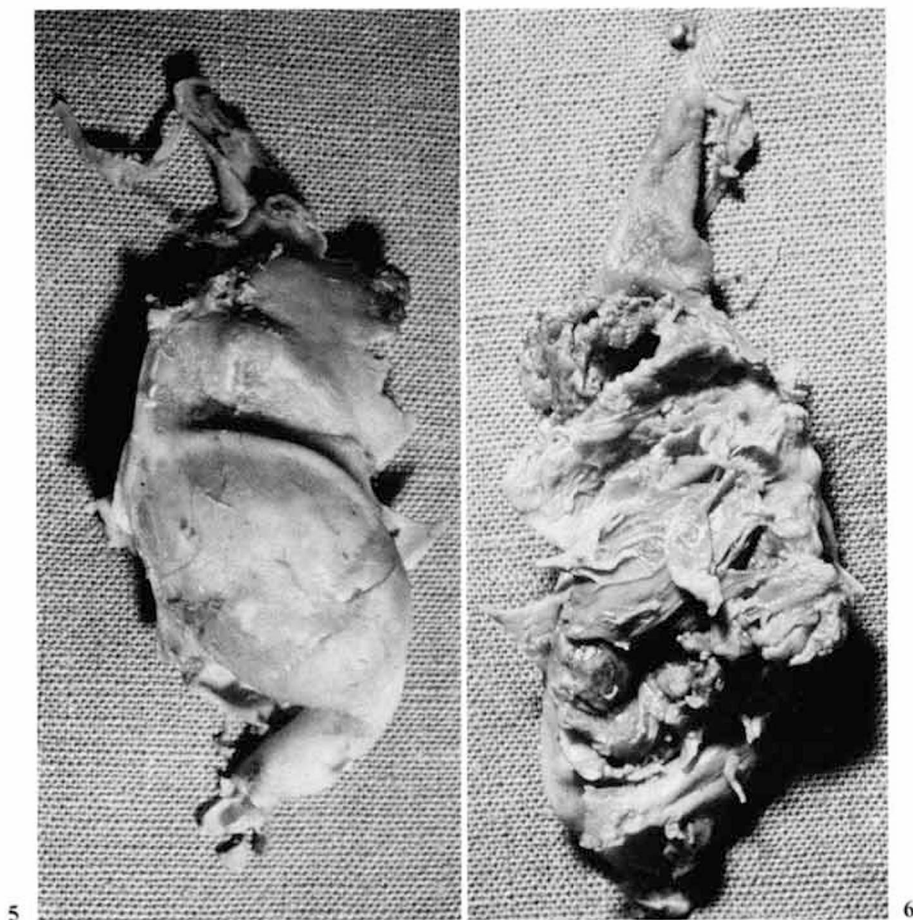


**Fig. 4.** Venous phase of the angiography shows the venous drainage of the lesion.

At present, 3 years after surgery, the patient's progress is good, with slight retardation, convergent strabismus and slight peripheric facial paresis, both on the right side.

### **Discussion**

The cerebral arteriovenous malformations present in different forms. While the angiomatous lesions usually present as a subarachnoid or intra-



**Fig. 5.** External aspect of the partially open aneurysmal sac.

**Fig. 6.** Internal aspect of the sac. Note the nonhomogenous conformation of the clot.

parenchymatous hemorrhage [2, 6, 11, 13], fistulous lesions appear initially by cardiac failure, intracranial hypertension or by the combination of both, associated or not with intracranial bruits [10]. Less frequently, both types of malformation can present as epileptic seizures of Jacksonian type, transient ischemic attacks, focal progressive neurological deficit, etc.

[2]. While the carotid-cavernous fistula is predominant in adults, the most common fistula in infancy is the abnormal communication between branches of the vertebral or vertebral and carotid arteries with the vein of Galen which causes a secondary dilation of the latter. In this type of malformation, four different clinical pictures have been described: (1) decompensated cardiac failure with intracranial bruit in newborn infants; (2) moderate cardiac failure with macrocephaly and intracranial bruit in 3- to 6-month-old infants; (3) macrocephaly and intracranial bruit with no cardiac signs in children between 6 months and 3 years old, and (4) subarachnoid hemorrhage, intracranial hypertension or psychomotor retardation, with or without syncopal episodes or intracranial bruits in older children, adolescents, or adults [1]. *Dean* [4] recommends the consideration of a fifth form of presentation: an intracranial hypertension syndrome with no bruits and without cardiac signs owing to the fact that the aneurysm has clotted. Our patient presented a picture of intracranial hypertension because of hydrocephalus secondary to obstruction of the fourth ventricle by the malformation, as well as signs of compression, of the right side of the pons with right peripheric facial palsy and left brachiorural hemiparesis, but at no moment presented symptoms or signs of cardiac failure with increased cardiac output such as is observed in these types of arteriovenous malformation when they appear at this age [8, 14, 18]. The cause of cardiac failure is the great increase of the venous return to the right atrium by the decrease of the peripheric resistance owing to the abnormal communication between arterial and venous elements through which can circulate up to 90% of the cardiac output.

*Heinz et al.* [7] explain the absence of cardiac symptomatology as the result of the intrasacular thrombosis in pre-natal form or during the birth itself. On the other hand, the cases of *Lazar* [9] and *Six et al.* [16] did not have cardiac signs either, but in both, the clot had a laminated structure with aged areas and others of more recent formation. Our patient did not present symptoms or signs of cardiac failure either, in spite of the fact that the thrombosis of the malformation was relatively recent: owing to the absence of granulation tissue, it can be assumed that the clot was not more than 10 or 15 days old.

The cranial X-ray only showed nonspecific signs of intracranial hypertension. The chronic hypertension at aneurysmatic sac level can account for its calcification, although this is rare before 15 years of age [17]; the incidence of calcification is higher, nevertheless, in clotted cases. The brain scan revealed an increased uptake of the radionucleid in ring form because

of the wall of the aneurysmatic sac, the altered surrounding tissue, or because of both situations simultaneously; sometimes and when it exists, the cause of this high uptake is attributed to the granulation tissue of the clot. The center of the lesion without high uptake on the brain scan made us suppose at that moment that we were dealing with a probable cystic astrocytoma of the cerebellum. The CT scan findings are interesting as they deal with an initially hyperdense lesion, although with uneven densities in its interior and without enhancement after injecting the contrast agent, compatible with a blood clot. This contrast agent revealed new images on CT, not observed before, corresponding to elements with free circulating blood which were afferent and efferent to the lesion. It was considered without free circulating blood inside in view of the absence of enhancement after contrast injection. *Six et al.* [16] say that clotted malformations show a peripheric enhancement after contrast material owing to the granulation tissue which exists at the level of the wall of the sac, which is a phenomenon already observed before by the same author in cerebral arterial aneurysms [15]. Furthermore, he has seen, as in our case, the existence of uneven blood densities within the lesion and the absence of perilesional edema. His case, a clotted arteriovenous aneurysm of the vein of Galen, had a density of 40 EMI units, whilst ours revealed a density of 21–44 EMI units. Usually, clotted blood, with about 90% hematocrit, presents an average density of 40 EMI units and free circulating blood, with about 45% hematocrit, an average density of 18–20 EMI units. The case of *Dean* [4] showed, as the previous one, a hyperdense image with evident ring form enhancement after contrast. *Amacher et al.* [2] reported a similar case to ours (case I of their series), in a 9-year-old patient with intracranial hypertension evidenced by headaches with papilledema and splitting of sutures; the CT revealed a mass lesion in the left cerebellar hemisphere, and the angiography showed hydrocephalus with a cut off vessel at lesion level, which turned out to be a clotted 'venous angioma' [2].

According to *Epstein et al.* [5] we emphasize the indication of angiography when the tomographic image does not coincide, as in our case, with the findings usually observed in tumors of the posterior fossa.

Summarizing, this is an arteriovenous fistula which did not produce cardiac failure, but did produce a secondary aneurysmatic venous dilation, with a high-density image on the uncontrasted CT scan due to a recently formed clot. For this reason, furthermore, it did not fill in the angiography and did not present signs of steal in the vertebral and carotid territories either.



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