Congenital Arteriovenous Malformation Associated with Progressive Hydrocephalus in a Newborn

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Intracranial arteriovenous malformation is rarely presented in newborns or infants. We describe an unusual case of congenital arteriovenous malformation of the brain with multiple sequestered grape-like venous sacs presented with congenital hydrocephalus. This 4-month-old girl born with a large head, presented with progressive hydrocephalus over a period of 4 months. The brain CT showed multiloculated cysts with a high-density mural nodule and thin cerebral mantle. The right lateral ventricle was collapsed by the cystic lesion, and the contralateral ventricle was markedly dilated, which was thought to be due to aqueductal obstruction by the conglomerated nidus of the arteriovenous malformation. Surgical removal of both hemispheric masses, including the overlying thin mantle. The lesion was pathologically confirmed as the arteriovenous malformation which was composed of markedly dilated veins and multiple sequestered aneurysmal sacs. The overlying cerebral tissue was dysplastic and partly infarcted. This case shows that intracerebral arteriovenous malformation is indeed a congenital anomaly and suggests that intrauterine vascular compromise can result in focal or wide maldevelopment of the brain.

Key Words: Arteriovenous malformation, Congenital, Hydrocephalus, Newborn

INTRODUCTION

Intracranial arteriovenous malformation (ICAVM) in children is a rare congenital lesion that can be symptomatic even in the newborn period (Cho, 1978; Gerosa et al., 1981; Jellinger, 1986).

Since the essential feature of AVM is the formation of fistula or shunt, thus permiting arterial blood to enter the venous system without passing through an arteriole-capillary bed, they have usually attained considerable size, and the draining veins may become huge by distended venous channels (Scott et al., 1954). If the development and growth of ICAVM occur during the early embryologic period, the damage or maldevelopment of the surrounding brain parenchyma may be considerable.

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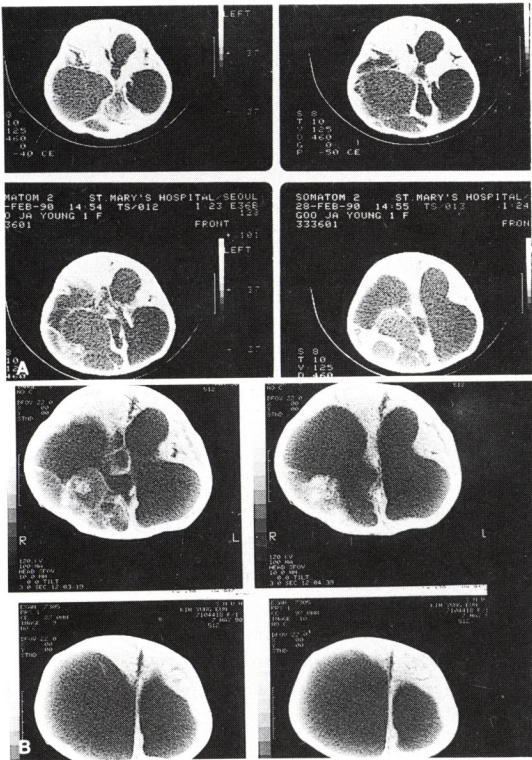
Clinically, ICAVM usually presents with hemorrhage or epilepsy (Wilkins, 1985). In infants, however, it can cause both hydrocephalus and congestive heart failure (Holden et al., 1972; Lavoie et al., 1972). Hydrocephalus associated with AVM is usually of the communicating type, however, its pathogenesis is still a matter of conjecture.

We experienced an unusual case of congenital arteriovenous malformation of the brain in a 4-month-old girl presented with hydrocephalus and associated with multiple grape-like vascular malformation and changes of the cerebral mantle.

CASE REPORT

Clinical manifestations

This 4-month-old-girl was born with a normal full term delivery by Cesarian-section because of cephalopelvic disproportion. At birth, she already had an enlarged head measuring 40.3cm in circumference (normal: 31 ± 1.5cm). Brain CT revealed a multiloculated cystic

Fig. 1. Brain CT reveals a multiloculated cystic lesion with mural nodules (arrows) and paper-thin cerebral mantle. A: brain CT at 1 month of age, B: Brain CT at 4 months of age

lesion with a mural nodule, along with extreme hydrocephalus with a paper thin cerebral mantle of both hemisphere (Fig. 1). At 2 months of age, the head enlarged further to reach 51.4cm in circumference, and it enlarged to 61cm at the 4 months. The scalp veins showed bluish engorgements. The patient was clinically unmeasurable except for mild irritability. Her sucking power was good. Motor and sensory functions were relatively intact. The signs or symptoms of congestive heart failure was not present. No angiographic study was done. At the age of 4 months, she underwent a craniotomy for a total removal of the mass with the clinical impression of primitive neuroectodermal tumor.

Brain CT Scan and MRI

The first brain CT Scan (12/22/'89) showed extreme ventriculomegaly and fluid-blood layering in a deep central cyst located at the interhemispheric as well as supracerebellar cisterns, suggesting bleeding from the lesion. An ill defined isodense solid part adjacent to the central cyst was seen in the huge cyst occupying the entire right temporoparieto-occipital region.

CT Scan (2/28/'90) revealed changes in configuration of the solid part and its location. Main solid part, a cluster of multiple rings, 6 × 4cm in size, located at the superficial surface of the huge cyst. The rim of rings showed moderate enhancement in some portions by the contrast agent.

CT scan (4/11/'90) showed further enlargement of the cysts and ventricles. Plain CT scan revealed 9×5cm sized mixed density nodule which increased in size. No contrast enhancement of the mural nodule was seen.

T1-weighted (TR/TE = 600/30) image (4/16/90) showed 5 × 2.5cm sized right temporoparietal lesion which consisted of multiple beads of high signal intensity in conglomerated nodular fashion or in a band of beads around a cyst. On T2-weighted (TR/TE = 2500/80) image the mural nodule revealed low signal intensity solid mass containing several scattered small (less than 1 cm in diameter) high signal dots.

Numerous cysts of variable size occupied the entire right temporo-parieto-occipital region and ipsilateral frontal horn was small. Contralateral lateral ventricle were extremely dilated as in CT scan findings.

Operative Findings

Right posterior parietal and parts of temporo-occipital lobes were exposed. Exposed cortical surface, 6.7cm, was deformed and disorganized It was impossible to recognize normal gyral and sulcal pattern. The large and small blood vessels were running haphazardly on the deformed flat yellow cortical surface.

Many subcortical small multiloculated cysts abutted on the surface contained yellow fluid and looked like a cluster of grapes as a whole. On deeper dissection several huge cysts occupying the entire temporoparieto-occipital area contained clear watery fluid and separated each other only by cobweb-like thin membraneous septa. Solid part was en bloc removed, and huge cysts were made into a single chamber by fenestration procedure.

Pathology

The removed specimen was an irregular cystic mass showing a grayish white brain-like structure that was mixed with dark brownish solid hemorrhagic areas with several small cystic nodules (Fig. 2). Microscopically, the main lesion including the mural nodule was composed of abnormal arteries and aneurysmally dilated veins containing thrombi and maldeveloped cerebrum (Fig. 3). The isolated grape-like cystic nodules (0.3cm to 0.9cm in diameter) were aneurysmally dilated veins
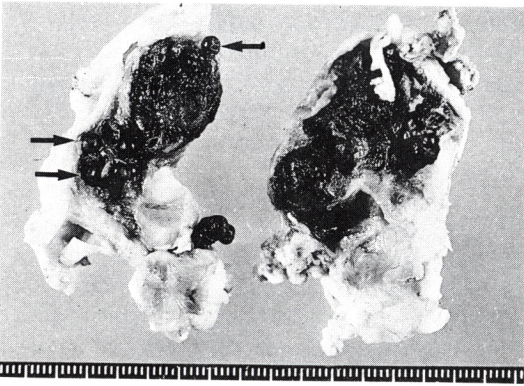
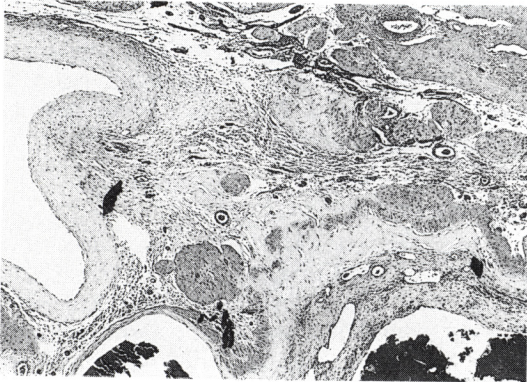**Fig. 2.** Gross specimen shows darkbrownish hemorrhagic area with several grape-like nodules (arrows).

Fig. 3. The main lesion is composed of abnormal arteries, aneurysmally dilated veins and maldeveloped cerebrum. (H&E)
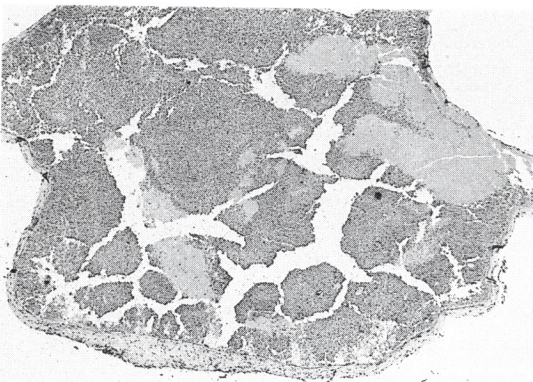

Fig. 4. Each grape-like structure represents a dilated vein containing blood. The wall shows irregular thickening and hyalinization. (H&E)

filled with old blood (Fig. 4). These veins showed mural fibrosis and irrgular thickening. The wall of abnormally distributed arteries showed minimal change. Microscopically, identical dilated veins were also embedded in the cerebral mantle (Fig. 5). Intervening neural tissue showed ischemic change, organizing infarct, focal
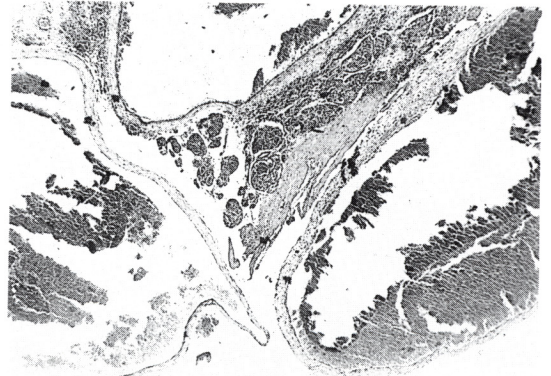

Fig. 5. The dilated veins, which are microscopically identical to the isolated grape-like lesion, are also embedded in the cerebral parenchyma. (H&E)

hemorrhage, hemosiderin deposit, and gliosis. Maldeveloped cortical nests consisting of dysplastic nests of neuronal cells were also scattered, and the remaining cortex showed poor layering.

DISCUSSION

Our case is characterized by (1) presence at birth, (2) megalocephalus and hydrocephalus, (3) grape-like sequestered venous sacs, and (4) maldevelopment of the cerebral mantle along with an old infarct.

The most common congenital intracranial lesions are arteriovenous malformations and arterial aneurysms (Lagos and Riley, 1971). Since experiences of vascular malformation in children are rather limited, definite modes of diagnosis and therapy are still unavailable. Although postnatal enlargement of the head can develop fairly commonly in infants born with ICAVM seen after the neonatal period, congenital hydrocephalus associated with AVM is rare (Askenasy et al., 1953). Exceptionally, aneurysm of vein of Galen, one type of AVM, is not uncommonly reported in infants and is usually accompanied by hydrocephalus and congestive heart failure (Takashima and Becker, 1980; Foroughi, 1960).

Askenasy et al. (1953) and Cronqvist et al. (1972) have discussed 3 types of hydrocephalic mechanisms

by ICAVM, namely, obstructive hydrocephalus, communicating hydrocephalus, and hydrocephalus ex vacuo due to local cerebral atrophy. If the malformation occurs on the great vein of Galen or is large enough to cause obstruction of the aqueduct, the hydrocephalus will be of an obstructive nature. If a large ICAVM is located in other sites, it might raise the venous pressure and could interfer with cerebrospinal fluid absorption and causing the communicating hydrocephalus.

The explanation of the hydrocephalic mechanism in our case could be ascribed for all of the above mechanisms in some way, because the AVM was large enough to cause the obstruction of the aqueduct or increase the venous pressure to interfere with the CSF absorption. Besides, the paper-thin cerebral mantle with infarct could be an element causing hydrocephalus ex vacuo.

The coexistence of AVM and multiple cerebral infarcts is another important finding in this case. AVM is an abnormal direct communication between arterial and venous channels without the interposition of the capillary system. The low-resistance high-outflow fistula predisposes the enlargement of any feeding arteries or draining veins and possibly the enlargement of the shunting vessels as well (Parkinson and Bakers, 1980). Paterson and McKissock (1956) suggest that the apparent growth of AVM's is due to a repeated, often silent, rupture of the vessels, leading to small hemorrhagic infarcts or pressure infarcts of adjacent brain tissue. Therefore, clinically, ICAVM usually present either with epilepsy due to parenchymal change or with actual hemorrhage (Mori et al., 1980). Gillingham (1953) pointed out that the hemorrhages associated with ICAVM often are minor and intraventricular rather than subarachnoid.

The most enigmatic feature in our case was the isolated aneurysmal sacs which were lined by endothelium and a hyalinized, irregularly-thickened muscular wall without elastic lamina. We wondered whether we were dealing with a mixed form of congenital vascular lesion. The coexistence of two types of ICAVM and berry aneurysm (Foroughi, 1960) was reported. However, in our case the grape-like structures were composed of dilated veins instead of arteries, and the identical vascular channels were embedded in the adjacent areas. It is still unknown, however, what create the sequestration of venous sac from the main tributary.

Vascular malformations are the result of incomplete differentiation of the primordial cerebral plexus into arteries, veins, and capillaries, and of a failure of separation of the dural and pial circulation. The mal-

formations are most certainly formed early in the development of the cerebral mantle (Hamby, 1958). Therefore, the cerebral dysplasia seen in our case suggests that the development and growth of AVM took place in early uterine life before the formation of the cerebral mantle and resulted in progressive destruction on the adjacent neural tissue. Such an evolution is evidently due to a progressive dilatation of the affected blood vessels, perhaps related to episodes of raised blood pressure with subsequent implication of other vessels in the immediate neighborhood (Hasse et al., 1986).

The treatment of ICAVM is surgical extirpation whenever feasible. The criteria for surgical intervention are based on a comprehensive angiographic visualization of the vascular malformation, on its location within the brain, and on a careful evaluation of the neurological damage.

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