Angiomatous Malformation as a Cause of Cerebral Calculus
Case Report

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The term "cerebral calculus" has been used to describe the radiological appearance of certain abnormal intracranial calcifications, solitary or multiple. The etiology of these calcifications is varied. In most of the reported cases, calcification of an intracerebral hematoma, tuberculosis or brain abscess was considered to be the underlying cause. Massive intracranial calcification and ossification caused by angiomatous malformation has not been reported previously.

Case Report

S.G., a 16-year-old Negro girl, came to the Epilepsy Clinic on January 6, 1965, with a 7-year history of "grand mal seizures." She began to have seizures when 10 years old. The seizures usually began with flashes of light and spots in the right visual field. These unformed visual hallucinations persisted for about 10 minutes, with nausea and occasional vomiting, followed by a generalized seizure with tonic-clonic phases. The patient had been placed on Dilantin and Phenobarbital by her local physician, with poor results. At the age of 9 years, she had sustained a minor head injury in an automobile accident, but did not lose consciousness. Skull x-rays were not made at that time. The past history was otherwise not remarkable. There was no family history of seizures.

Examination. The general physical findings were normal. There were no bruits over the head or neck. The neurological examination showed a normal mental status. The visual acuity was 20/20 without correction. Visual fields, by tangent screen and perimeter, were normal. Ocular fundi were normal with no evidence of angiomatic lesions in the retina. The rest of the cranial nerve examination and detailed motor, sensory and coordination tests were all normal.

The spinal fluid was clear and colorless, the pressure was 180 mm. of water. Spinal fluid protein was 80 mg. /100 ml. There were no cells in the spinal fluid. The white blood cell count was 6,500, the hematocrit 40 per cent, the urine analysis normal, and a fasting blood sugar 88 mg./100 ml. An x-ray of the chest was normal. A brain scan, using radioactive Hg 203 was normal at 20 minutes and at 120 minutes following injection of the isotope. The electroencephalogram was normal; photic driving showed well-developed alpha rhythms.

Skull x-rays revealed a calcified mass, 4 cm. in diameter, in the left occipital region. The mass appeared above the level of the tentorium and did not seem to be attached to the inner wall of the skull (Fig. 1). The calcification appeared diffuse and slightly lobulated, with some mottling.

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Right brachial and left carotid arteriograms were done. The only abnormality demonstrated was medial and inferior displacement of the occipital division of the left posterior cerebral artery (Fig. 2).

Pneumoencephalography was normal except that the occipital horn of the left lateral ventricle did not fill. It could not be determined whether this was due to congenital absence or to obliteration by the mass lesion (Fig. 3).
Cerebral Calculus due to Angiomatous Malformation

FIG. 3. Air study showing calcified mass and non-filling of occipital horn of left ventricle.

Operation. The left occipital pole was exposed and by probing through the cortex, a calcified mass was felt a few millimeters beneath the surface. The surface blood vessels were normal. The cortex was incised and the mass delivered from its bed within the parenchyma. It had obliterated the left occipital horn; there were no abnormal vessels.

Postoperatively, the patient had a right homonymous hemianopsia and developed paranoid delusions for the first time. Within a few days, however, there was complete recovery.

The surgical specimen was a lobulated, firm and friable, brown mass measuring 4 x 8 x 5 cm. in maximum dimensions. On section, there was diffuse calcification in all portions. The surface showed small blood-filled spaces. Histologically, it was composed of compact glial tissue containing foci of calcification and ossification varying from 0.2 mm. to 3.0 mm. in diameter. Most of these showed a concentric lamination. There were large blood vessels of venous and capillary type. Some vessels showed fibrous thickening and many showed calcification in their walls (Fig. 4). The intervening tissue was composed of glial fibers and small fibrillary astrocytes and contained scattered Rosenthal fibers. There was no evidence of recent or old hemorrhage. A portion of choroid plexus was adherent to one edge of the mass. The pathologic diagnosis was capillary-venous hemangioma, with extensive calcification and ossification (Fig. 5).

The patient was discharged on February 1, 1965.

Discussion

Intracranial angiomatous malformations can present a wide variety of clinical syndromes.14,17 In the younger age groups (up to 30 years), subarachnoid hemorrhage produces the most common signs. In the older age groups, focal seizures are a common symptom; various focal neurological signs may occur, depending upon the location of the lesion.17

There were several interesting features in this patient. She had focal seizures at the age of 10 years. During the next 6 years, she continued to have the same type of seizures but did not show

FIG. 4. Large blood vessels of venous type, showing calcification in vessel walls and in surrounding glial tissue. Verhoeff-Van Gieson X144.

evidence of intracranial bleeding at any time. The finding of a calcified mass intracranially, suggested many possibilities in the differential diagnosis. The causes of intracranial calcification fall into several categories,\(^1\) including vascular,\(^2\) inflammatory,\(^3\) neoplastic,\(^4\) traumatic,\(^5\) degenerative, endocrine,\(^6\) the neurocutaneous syndromes,\(^7\) and idiopathic cases.\(^8\) On the basis of the size and location of the calcification, we thought we were dealing with a calcified tumor, such as a meningioma or oligodendroglioma. On the basis of the type of calcification, we felt it was probably a calcified hematoma, the so-called "brain stone" or "cerebral calculus."

Summary

We have reported the case of a 16-year-old girl who for 6 years had had generalized seizures of focal onset. Skull x-rays demonstrated a left occipital "cerebral calculus." Clinical investigation did not clarify the nature of the lesion. The EEG and brain scan were normal. Arteriography and pneumoencephalography showed non-specific abnormalities.

The mass was removed and found to be a capillary-venous hemangioma with extensive calcification and ossification. The patient recovered and is well controlled on Dilantin 110 mg. twice daily.

References