Brain-stem hemangioma calcificans

Case report

MICHELE OCCHIOGROSSO, M.D., ARISTIDE CARELLA, M.D., PAOLA D'APRILE, M.D., AND GIACOMO VAILATI, M.D.

Department of Neurosurgery and Neuroradiology, University of Bari, Bari, Italy

A case of brain-stem hemangioma calcificans is described. The few cases reported in the literature prove the rarity of this tumor, which is considered a benign variant of cerebral cavernous hemangioma. Diagnosis and treatment of these tumors are briefly discussed with a review of the literature including 11 previous cases.

KEY WORDS • cerebral vascular lesion • intracranial calcification • hemangioma calcificans

HEMANGIOMA calcificans (HC), a term introduced by Penfield and Ward, 4 is a very rare tumor considered a variant of that category of cerebral vascular lesion known as cavernous hemangioma. 2 From a clinical and radiological standpoint, HC resembles very closely other rare conditions, such as endarteritis calcificans cerebri, 3 calcified hamartoma characterized by the presence of seizures, 1 and intracranial calcifications.

To date, only 11 cases of HC have been reported in the literature. 2'4-6'8 The present paper reports an additional case of HC localized to the posterior portion of the brain stem.

Case Report

This 35-year-old man was admitted to the neurosurgical department of the University of Bari on January 9, 1980. He complained of a 6-month history of headache, vomiting, petit mal seizures, diplopia, and blurring of vision. Six years previously he began to have attacks of true vertigo with transient diplopia, without loss of consciousness, which lasted only a few seconds; these attacks increased in frequency during the month prior to admission.

Examination. The general physical examination was normal; the neurological examination showed a left sixth nerve weakness, bilateral papilledema, and a mild left-sided dysmetria. Electroencephalography demonstrated a slow irregular activity over the left temporal region; routine laboratory tests were normal.

Skull x-ray films revealed a grossly oval calcified mass, 4 cm in diameter, in the occipital area; in the lateral view the mass appeared approximately at the level of the anterior portion of the tentorium, without attachment to the inner wall of the skull. Computerized tomography (CT) disclosed a hyperdense mass located in the posterior portion of the brain stem. The mass was lobulated and slightly mottled; no edema of the neighboring structures was evident. The aqueduct and posterior part of the third ventricle were compressed by the mass, resulting in secondary obstructive hydrocephalus (Fig. 1). There was no enhancement of the lesion after contrast injection. On left brachial arteriography the posterior cerebral arteries appeared stretched and displaced slightly upward in their middle portion; the superior cerebellar arteries were pushed downward. No feeding vessels, puddling of contrast material, or draining veins were seen.

Operation. On January 15, a ventriculoatrial shunt was inserted for the management of the hydrocephalus and, on January 28, a posterior fossa craniotomy was performed with the patient in the sitting position. A generous bilateral suboccipital craniectomy was carried out from the foramen magnum to 2.5 cm above the superior nuchal line. A Y-shaped dural incision was made, and two midline bridging veins from the cerebellum to the tentorium were divided. The vermis was gently displaced downward, and the tentorium retracted upward until the arachnoid of the quadrigeminal cistern was exposed. The arachnoid, which appeared thickened and white, was incised at the upper margin of the culmen and a solid, raspberry-sized grayish mass occu-
Brain-stem hemangioma calcificans

FIG. 1. Plain computerized tomography scans showing a hyperdense mass at the level of posterior surface of the brain stem. The mass, impinging on the aqueduct and on the posterior part of the third ventricle, causes secondary obstructive hydrocephalus.

pying the cistern of the quadrigeminal plate came into view. Dense xanthochromic adhesions between the arachnoid and the mass were divided. By careful microdissection, the tumor was completely exposed; it appeared grossly oval, 3 cm in diameter, occupying the whole quadrigeminal cistern. The mass extended from the pineal gland, anterior and inferior to the vein of Galen, to the anterior vermis which was pushed downward. The vein of Galen, basal veins of Rosenthal, and internal cerebral veins were all identified and dissected free from the superior portion of the tumor. Successively, adhesions between the inferior part of the mass and the anterior vermis were divided, and the mass was isolated from beneath. The tumor surface was then incised. Numerous calcifications, ranging from 1.5 to 0.5 cm in diameter, were removed from within the tumor together with a yellow semi-fluid poorly vascularized material. The residual cavity appeared grossly oval, measuring approximately 4 cm in depth. The irregular border of the mass, made up of the lateral and posterior wall of the tumor cavity, was finally excised with microscissors. The anterior wall was densely adherent to the quadrigeminal plate and was left in situ.

Pathological Examination. Microscopic examination revealed numerous endothelial blood spaces of variable size, separated by a matrix of dense fibrous tissue with irregular areas of calcifications. No neural elements were present. Diagnosis of hemangioma calcificans was made (Fig. 2).

Postoperative Course. The postoperative course was uneventful. A postoperative CT scan showed only a few residual areas of calcification (Fig. 3). The patient was discharged on February 18, 1980; at follow-up examinations 6 months and 1 year after the operation, he showed only a mild left-sided dysmetria.

Discussion

Cavernous hemangioma is a vascular hamartoma consisting of blood vessels (veins and capillaries) lined by single layers of endothelial cells separated by a matrix of fibrous, collagenous tissue with no evidence of neural structures. Hemangioma calcificans is a very rare benign variant of cavernous hemangioma which is characterized by prominent calcification.

Previous reports show that HC is generally found in patients between the 2nd and 5th decade of life. Fig. 2. Microphotograph of the tumor. Numerous endothelial blood spaces of variable size, separated by a matrix of fibrous tissue with irregular areas of calcifications, are evident. H & E, × 110.
M. Occhiogrosso, *et al.*

**FIG. 3.** Postoperative computerized tomography scans showing evidence of few residual calcifications adhering to the brain stem. The ventricular catheter of the shunt is evident in the right frontal horn.

life. The incidence is similar for males and females. The location is always supratentorial (usually temporal), near the Sylvian fissure, frontotemporal or occipital, completely within the cerebral parenchyma a few millimeters beneath the cerebral surface. This tumor is generally solitary; only one case has been reported with multiple and bilateral lesions.2

The medical history of patients affected by HC is characterized by a long-standing focal, psychomotor, or generalized epilepsy. Neurological examination fails to show positive findings or shows only slight focal signs. Skull x-ray films demonstrate the presence of calcifications varying in shape and size. The calcifications can be in the form of scattered granules or radiating lines, or they can be compact (a few centimeters in diameter), rounded with dense nodules, or mottled.

The CT scan shows a compact or nodular hyperdense lesion. Only one case4 is reported with a slight enhancement after infusion of contrast medium; this case is the only one in which the plain skull films failed to show evidence of intracranial calcifications. Edema of surrounding structures has never been noted on CT scan. Cerebral angiography demonstrates the findings of an avascular mass lesion, with no evidence of feeding vessels, tumor stain, or draining veins. In one case4 a vague puddling of contrast material in the midst of the calcifications was seen in the venous phase.

The diagnosis of HC is made on the basis of the radiological and clinical findings. The benign nature of the lesion brings up the problem of indications for surgery. A wait-and-see attitude is justified only when anticonvulsant therapy is effective, the lesion is not accessible, the neurological examination is negative, and serial CT scans demonstrate a non-enlarging mass. Hemorrhage in the lesion has not been described.

In our patient, who represents the first reported case of HC of the brain stem, there was a picture of increased intracranial pressure, due to obstructive hydrocephalus.

**References**


Manuscript received October 12, 1982. Accepted in final form January 27, 1983.

This paper was presented in part at the 10th Congress of the European Society of Neuroradiology, Milan, Italy, September 25–26, 1981.

Address reprint requests to: Michele Occhiogrosso, M.D., Cattedra di Neurochirurgia, Università di Bari, Policlinico, 70100 Bari, Italy.