Cystic cerebral cavernous angioma with dense calcification

Case report

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A case of cavernous angioma in the right temporoparieto-occipital area is reported. The unusually large area of calcification and cystic formation presented problems in preoperative diagnosis. The tumor was treated successfully by subtotal excision.

KEY WORDS • cystic cavernous angioma • cerebral calcification • intracerebral hemorrhage

Cavernous angioma is a relatively rare vascular malformation in the central nervous system that is curable by surgical removal. It produces a wide range of clinical symptoms, and there are no neuroradiological findings typical of this entity. On angiography it most often appears as an avascular mass, making the diagnosis difficult. Epileptic seizures and episodes of headache, probably due to recurrent subarachnoid or intracerebral hemorrhage, are the initial symptoms.4,15,19

We report a cavernous angioma with an unusually large area of calcification and cystic formation in the right temporoparieto-occipital region. The initial diagnosis was an oligodendroglialoma.

Case Report

This 45-year-old woman experienced a grand mal seizure at the age of 27 years, during her first pregnancy. She remained asymptomatic until 3 years before admission to our clinic, during which time she complained of episodic headaches, with nausea and vomiting, and impairment of memory. No other seizures were reported. Six months prior to admission she experienced bilateral visual difficulty and occasional episodes of weakness in the left arm. Her family remarked on an alteration in her behavior, in that she lacked concentration and was indifferent to her surroundings. Before her admission, skull films revealed an extensive area of intracranial calcification in the right temporoparieto-occipital region and in the basal ganglia. She was referred to us for further diagnosis with the suspicion of an oligodendroglialoma.

Examination. The patient was alert and well oriented, but her concentration was impaired, and she was restless and often indifferent. Visual acuity was bilaterally impaired. Her restless condition prevented us from making an accurate examination of the visual fields. Both optic discs were pale, and she had strabismus (medial rectus palsy) on the left, left central facial paresis, mild left hemiparesis, trunk ataxia, and incoordination of the left arm and leg.

Irregular slow activity over the right hemisphere, chiefly in the right tempo-occipital region was noted on electroencephalography. Computerized tomography (CT) scan revealed a tumor, 8 cm in diameter, in the right temporoparieto-occipital region. It showed many amorphous calcium deposits (with maximum density of 700 Hounsfield units) extending from the base of the tumor up to the brain stem. A very large cyst formation with homogeneous content was observed in the parietal part of the tumor. Fine calcified structures were noted in the cyst walls (Fig. 1).

The skull was bulging and appeared thin in the right temporo-occipital region (Fig. 1). A small node at the occipital border of the tumor appeared to enhance following intravenous infusion of contrast material. Compression of the right lateral ventricle was observed, as well as massive displacement of the brain stem and hydrocephalic enlargement of the left lateral
ventricle. Right carotid and left vertebral angiography revealed a large avascular, probably cystic, mass in the right temporoparieto-occipital region (Fig. 2).

Operation. On January 19, 1979, a right temporoparieto-occipital craniotomy was performed. The dura was thin, and the cortex showed a yellowish discoloration. A huge subcortical cyst, with an estimated dimension of $8 \times 5 \times 4$ cm, was found that contained a yellowish-brown liquid. The cyst extended into the temporal region to the midline. There were calcified masses in the lateral and medial cyst walls. Evacuation of the cyst and subtotal removal of the tumor (that is, of the lateral portion) was performed. The mass was adherent to the surrounding tissue; it had no capsule, but could be easily removed with only mild bleeding.

Postoperative Course. The patient remained alert and oriented, but with an exacerbation of her nervous state, which gradually improved. She was discharged on February 2. In the follow-up examination on March 9, she was doing well and her memory had improved.

Pathological Examination. Examination of the specimen revealed marked gliosis as a reaction to hemorrhage. The origin of the hemorrhage was found to be a network of vascular channels with atypical walls and fibroblastic proliferation (Fig. 3). Pearl-like calcifications were found in the surrounding tissue.

Discussion

In 1948, Penfield and Ward\textsuperscript{18} reported 12 calcified epileptogenic lesions of the temporal lobe. Five of these lesions were calcified vascular tumors that did not fit into the classification of Cushing and Bailey, reported in 1928.\textsuperscript{3} In two cases, the calcifications appeared very similar to a condition called “endarteritis calcificans cerebri” by Geyelin and Penfield.\textsuperscript{5} These vascular tumors were classified by Penfield and Ward as hemangioma calcificans.

According to Russell and Rubinstein,\textsuperscript{18} cavernous angioma is classified as a form of blood vessel hamartoma that consists of dilated contiguous vascular spaces, separated by thin layers of fibrous tissue. The absence of intervascular neural tissue distinguishes it from other vascular hamartomas. The surrounding tissue can be scarred and often contains calcifications.\textsuperscript{22} The histological differentiation between capillary teleangiectasia and cavernomas may be difficult.\textsuperscript{22}

Cavernous angiomas are relatively rare. In their
Cystic cerebral cavernous angioma

Fig. 2. Preoperative right carotid angiogram, anteroposterior (left) and lateral (right) views, showing the large mass with calcifications.

review of the literature, Voigt and Yaşargil found 164 cases and added one more. Giombini and Morello reported a further 14 cases in 1978. These entities appear most frequently in the cerebral hemispheres, but have been described in the basal ganglia, pons, sellar region, ventricular system, cerebellum, spinal cord, internal auditory canal, cauda equina, orbit, and pineal region. They can be multiple.

Our case is presented because of the size of the cyst and calcification, which presented problems in the preoperative diagnosis. There is no pattern of calcification of cavernomas, which are usually small; they vary in frequency from 11% to 40% of cases. In most cases, angiography shows an avascular lesion. A wide vein or veins draining a poorly vascularized mass has been demonstrated in some cases. A vascular blush in the capillary phase was related as suggestive of cavernomas by Krayenbühl, et al. Our patient had neither pathological vessels nor early draining veins.

We have found very few reports of CT scanning of cavernomas; a mild contrast enhancement with no significant mass effect has been described. A definitive decision about contrast enhancement in parts of the tumor was not possible in our patient because of the poor quality of the scans (caused by the very high density of the calcification). Usually the diagnosis cannot be established before operation, and the lesions are falsely diagnosed as meningioma or oligodendroglioma. In our case, the CT scan was not indicative of oligodendroglioma because of the amorphous shape and position of the calcium deposits in relation to the cyst.

Cavernous angiomas can usually be treated safely by surgery, which is indicated in order to avoid the risk of an intracranial hemorrhage.

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Fig. 3. Histological appearance of the network of vessels showing irregular walls with fibroblastic proliferation. H & E, X 50.
References


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