Giant cystic cavernoma in a child

Case report

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A totally cystic giant cavernous hemangioma is described in a 3-year-old girl. The clinical presentation and
computerized tomography findings were both unique. The patient was successfully treated by surgery.

KEY WORDS • cavernous hemangioma • cystic tumor • child •
computerized tomography

CRYPTIC arteriovenous malformations (AVM's)
have been described by Crawford and Russell.3
Cavernous hemangiomas form a distinct group
of these AVM's. According to Giombini and Morello,8
cavernous hemangiomas "are made up of large vascular
sinusoidal spaces, separated by walls of varying thick-
ness common to several luminae. The cavities, occupied
by thrombi in different stages of organisation, are in-
vested with endothelium but have no smooth muscle
or elastic covering." The walls of cavernous heman-
giomas are wholly composed of collagen and are lined
by a single layer of endothelium,19 and the surrounding
tissue has been known to contain calcium.8

We are reporting the case of a child with a giant
cystic cavernoma. She presented with progressive en-
largement of the head, which has not been described
before in association with proven cavernous heman-
giomas. Only two previous reports of cystic cavernous
hemangioma visualized on computerized tomography
(CT) scanning are available.18,23 Our patient under-
went total surgical extirpation of the mass, with subse-
quent cure.

Case Report

This 3-year-old girl was admitted on February 22,
1983. She was a third child, delivered by Caesarean
section. Her early development was normal. Symptoms
were first noted in October, 1982, with low-grade fever
lasting for about a week. She was treated by a general
practitioner and the fever subsided. A few days later,
she again had fever with cough and a swelling in the
neck, which disappeared with medical treatment, and
the child recovered. In the 1st week of November, 1982,
she began to have difficulty in walking, with repeated
falls, and this unsteadiness of gait slowly progressed. A
week later she began to have "tremors" of both upper
limbs and was unable to hold objects. This was followed
by difficulty in sitting up and progressive increase in
the size of her head. In January, 1983, her speech
became slurred and unclear. Clinical deterioration pro-
gressed to the extent that the child was unable to sit,
stand, or walk on her own. There was no history suggest-
tive of subarachnoid hemorrhage, raised intracranial
pressure, or cerebral seizures.

Examination. Examination revealed a conscious
child with preserved higher mental functions. There
was bilateral papilledema. Muscle tone was increased
in all limbs and tendon jerks were brisk, with bilateral
extensor plantar responses. She had right hemiparesis
with gross truncal and gait ataxia, and was unable to sit
or walk, even with support. Cerebellar signs were posi-
tive bilaterally, Head circumference was 54 cm. Mac-
ewen's sign was positive and the anterior fontanel was
wide open. There were no meningeal signs. Skull, spine,
and systemic examination was unremarkable.

Laboratory investigations revealed a hemoglobin
count of 12.5 gm%. Electrolytes, urea, and chest radi-
ogram were all normal. An x-ray film of the skull
showed sutural diastasis, but no calcification. A com-
puterized tomography (CT) scan (Fig. 1) revealed a
large irregular cystic lesion with well defined margins occupying a major part of the left cerebral hemisphere. There was a midline shift to the right. The cyst contents had a mean density of 18.1 Hounsfield units (cerebrospinal fluid (CSF) density in the same patient was 9 Hounsfield units). No enhancement was seen after administration of contrast medium. The cyst was separated from the ventricular system, ventricular dilatation was present, with no evidence of calcification or a high-density lesion. Lumbar puncture yielded globulin-positive CSF containing 97 mg% glucose and 2.4 gm% protein.

Operation. On March 2, 1983, a left frontal craniotomy revealed a large cystic cavity containing about 250 ml of decomposing liquid blood. After drainage of the cyst, a network of blood vessels was disclosed lying on the medial surface of the frontal lobe. This was excised in toto. There were no large feeding or draining vessels.

Postoperative Course. The patient’s postoperative course was smooth. At follow-up examination 4 months later, she showed remarkable improvement and was functioning totally independently. Her head circumference had decreased to 52 cm.

Pathological Examination. The fixed specimen was a dark 3 × 2 × 1-cm mass with tiny cystic spaces visible on the cut surface. One surface was composed of rusty brown cortical brain tissue. Microscopically, the specimen consisted of thin-walled closely packed tissue, with large vascular spaces, most of which were about 250 μm in diameter (range 50 μm to 1 mm). These spaces were lined by a single layer of endothelium and in some there was focal hyaline thickening of the wall. Neural tissue was absent between these vessels. The brain tissue adjoining the lesion showed marked gliosis and an excessive number of capillaries which were heavily impregnated with hemosiderin. Several hemosiderin-containing macrophages were also identified (Fig. 2). A diagnosis of cavernous hemangioma was made.

Discussion
Cavernous angiomas are rare lesions.4,8,17 They may be intra-axial or extra-axial;14 90% are supratentorial and 5% are infratentorial, according to the relative brain volume.8 In 1976, Voigt and Yaşargil25 analyzed 164 cases, but in a later review, Giombini and Morello8 found histological proof and complete data in only 37 of these patients. Reporting the cases of 14 patients of their own, these authors analyzed a total of 51 cases, with only 33 surgical cures. The youngest patient in that series was aged 2½ years. Familial congenital occurrence and multiple site involvement have been described.2,21,25 The Rolandic area is the commonest site for these lesions.19,25 Seizures are the presenting symptom in 38% of cases, headache in 28%, and intracranial hemorrhage in 23%.8 Focal deficits occur in only 12% of cases.8 A few cavernomas are reported in children;1,8,17,19 however, there are no previous reports of patients with proven cavernous angiomas presenting with an enlarging head. The clinical features in our 3-year-old patient and the CT appearance form a unique presentation.

Calcification is seen in 11% to 40% of cases, and even less in children.8,25 Angiographic demonstration of these lesions is poor and not diagnostic.14 It may show an avascular mass with or without a draining vein,4 or a blush.1,8,10 Non-opacification of the lesion could be caused by vasospasm, thrombosis, pressure by hematoma, blood vessels too small in caliber to be visualized, a sluggish circulation, or complete obliteration by hemorrhage. Angiography may localize the lesion, but cannot suggest the pathology.1,4,7,10,12,13,16,30

The CT appearance in this condition has been de-
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scribed only recently.\textsuperscript{1,3,14,17,18,22,24} Pozzati, et al.\textsuperscript{17} suggested certain criteria for CT diagnosis, and Bartlett and Kishore\textsuperscript{1} have described similar findings but with definite evidence of enhancement of the lesion. Mizutani, et al.\textsuperscript{14} found a nonhomogeneous diencephalic mass with marked enhancement which proved to be a cavernous hemangioma. Bitoh, et al.\textsuperscript{3} described three cases of cavernoma and believed that there were no definite diagnostic features; various lesions are known to mimic cavernous angiomas on CT scanning.\textsuperscript{1,14,18} However, none of these authors described cystic cavernous hemangiomas. Only two previous reports of cystic cavernomas examined by CT scanning are available.\textsuperscript{1,18} Only one lesion was totally cystic and that cystic cavernomas examined by CT scanning are avail-

table.\textsuperscript{18,23} Our patient's scans showed no enhancement. Increased density in cavernomas on CT scanning is attributed to calcium, local hemmorhages, or thrombosis,\textsuperscript{1,12,11} and thus can be mod-
ifed by the degree of calcification or hemorrhage. Val-

tues below calcium levels might be due to hemosiderin or smaller hemorrhages.\textsuperscript{14}

Our patient had a large cyst without calcification or hemorrhage but with mass effect, which is at variance with most of the above reports. We have failed to find any similar report of CT scanning in cases of cavernoma. We believe that the cavernous hemangioma underwent a “suicidal” bleed, was compressed by the cystic mass, and hence could not be demonstrated on the CT scan. We recommend that patients with a large cyst of the kind described should be subjected to a craniotomy and a careful search should be made for a vascular anomaly leading to complete excision and cure. With any other mode of treatment, this curable condition would probably be missed.

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