

Case Report

Extracerebral Cavernous Hemangioma of the Cavernous Sinus: Diagnosis with MR Imaging and Labeled Red Cell Blood Pool Scintigraphy

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Summary: We present the case of a 64-year-old man with a presumed diagnosis of extracerebral cavernous hemangioma involving the cavernous sinus. The diagnosis was made on the basis of labeled red cell blood pool scintigraphy findings in conjunction with those of MR imaging. This lesion was not altered in appearance at 6-year follow-up MR imaging. We also present the labeled red cell blood pool scintigraphy findings obtained in three other patients with similar-appearing cavernous sinus lesions at MR imaging who underwent subsequent biopsy; histologic findings confirmed chondrosarcoma, chordoma, and meningioma, respectively.

Cavernous hemangiomas are common vascular malformations found in many organs including the central nervous system (1). Intracranial cavernous hemangiomas have been reported to comprise 5–13% of all intracranial vascular malformations (2) and are usually intraparenchymal and occult at angiography (3). Extracerebral cavernous hemangiomas are very rare lesions encountered in the cavernous sinus region or at the cerebellopontine angle with extension into the internal acoustic canals (3, 4).

Intracranial extracerebral cavernous hemangiomas involving the cavernous sinus comprise less than 1% of all parasellar masses (1) and have different clinical and imaging features compared with the more common intraaxial cavernous hemangiomas (4). Accurate preoperative diagnosis of extracerebral cavernous sinus cavernous hemangiomas is generally difficult, with preoperative misdiagnosis of meningioma often made (2, 5, 6). This has serious surgical implications, because piecemeal removal of cavernous sinus cavernous hemangiomas is often difficult because of the hemorrhagic nature of the tumor and the complex relationship to nearby neurovascular structures (7, 8).

Case Reports

A 64-year-old man presented to his local doctor following an episode of numbness affecting the right side of his body that

lasted approximately 5 minutes. On suspicion of a transient ischemic attack, CT was performed, which demonstrated a lesion in the left cavernous sinus. He was referred to a neurosurgeon, and examination at the time of referral revealed no focal neurologic deficit related to this episode. He was then referred for MR imaging.

T1-weighted MR images demonstrated a hypointense mass centered on the left cavernous sinus (Fig 1A). This lesion was markedly hyperintense on both proton density- and T2-weighted images (Fig 1B). There was encasement of the cavernous segment of the left internal carotid artery without narrowing. Following administration of gadolinium-DTPA (Magnevist, Schering AG, Germany), there was heterogeneous enhancement of the left cavernous sinus mass with centripetal “filling in” of the lesion between the axial and coronal fat-saturated T1-weighted images (Fig 1C and D).

A labeled red cell blood pool scintigram was performed by using the modified in vitro method of labeling red cells with ^{99m}Tc-pertechnetate. Dynamic planar flow images, sequential blood pool planar images at 60-second intervals for 4 minutes and single photon emission tomography (SPECT) images at 30 minutes and 3 hours were obtained. Flow images did not demonstrate hypervascularity. Sequential blood pool images showed progressive slow accumulation of labeled red cells in the region of the left cavernous sinus. Delayed SPECT images at 30 minutes and 3 hours also showed persistent tracer accumulation in the region of the left cavernous sinus (Fig 2A and B).

On the basis of the MR and nuclear medicine findings, a presumptive diagnosis of left cavernous sinus cavernous hemangioma was made, and the patient was treated conservatively with annual follow-up MR imaging. The appearance of the left cavernous sinus lesion has remained unchanged over a 6-year period with no evidence of interval lesion growth or progressive narrowing of the cavernous segment of the left internal carotid artery.

Subsequent to the investigations in our subject, three other patients with similar-appearing cavernous sinus lesions at MR imaging (ie, T2 hyperintense and T1 iso- or hypointense with contrast enhancement) were evaluated with labeled red cell blood pool scintigraphy:

1. A 51-year-old woman with a mildly T2 hyperintense left cavernous sinus mass demonstrated photopenia of the left cavernous sinus region at labeled red cell blood pool scintigraphy. The lesion was resected and histologic analysis confirmed the presence of a meningioma (Fig 3A–D).

2. A 50-year-old woman with a markedly T2 hyperintense left cavernous sinus mass that demonstrated photopenia of the left cavernous sinus region on labeled red cell blood pool scintigraphy. This mass was resected, and histologic analysis confirmed the presence of a chordoma (Fig 4A and B).

3. A 22-year-old man with a markedly T2 hyperintense left cavernous sinus mass that demonstrated photopenia of the left cavernous sinus region on labeled red cell blood pool scintigraphy. This mass was resected, and histologic analysis confirmed the presence of a low-grade chondrosarcoma (Fig 5A and B).

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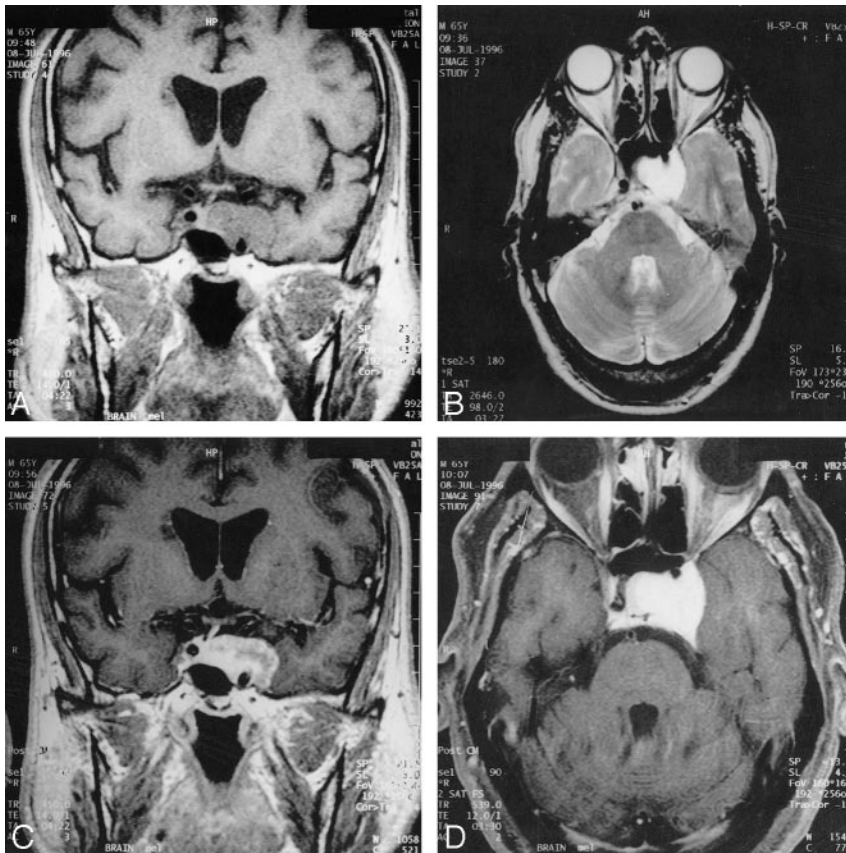


FIG 1. Cavernous hemangioma.

A, Coronal T1-weighted image demonstrates a hypointense left parasellar extraaxial lesion.

B, Axial T2 image demonstrates a homogeneous markedly hyperintense lesion.

C, Initial coronal gadolinium-enhanced T1-weighted image demonstrates patchy peripheral lesion enhancement.

D, Subsequent axial gadolinium-enhanced T1-weighted image now demonstrates homogeneous lesion enhancement with centripetal "filling in."

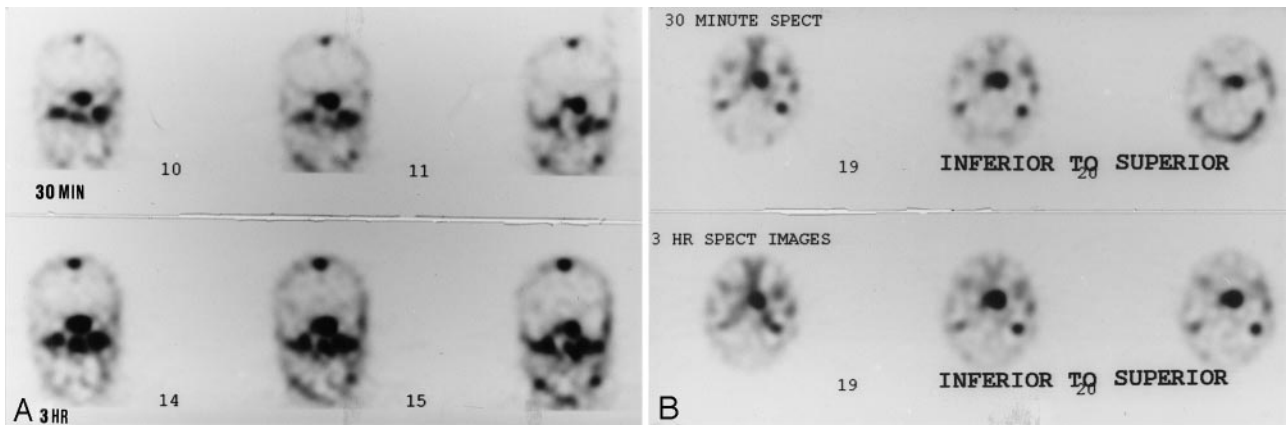


FIG 2. Cavernous hemangioma.

Coronal (A) and axial (B) labeled red cell blood pool SPECT images demonstrates progressive increase in tracer accumulation in the left parasellar region at 30 minutes (top) and at 3 hours (bottom).

Discussion

Cavernous hemangiomas are common vascular malformations found in many organs, including the central nervous system (1). Intracranial cavernous hemangiomas have been reported to comprise 5–13% of all intracranial vascular malformations (2). These lesions are usually intraparenchymal and occult at angiography (3). Cavernous hemangiomas are rarely found in an extracerebral location but are usually found in either the middle cranial fossa involving the cavernous sinus or the cerebellopontine angles with extension into the internal acoustic meatus (3, 4, 7, 9–11).

Extracerebral cavernous hemangiomas have a different natural history from their intracerebral (intraparenchymal) counterparts (12). The clinical presentation, imaging findings, and surgical outlook are also different (4, 9). Extracerebral cavernous sinus cavernous hemangiomas have a strong female predominance of as much as 7:1 (12–14) and are most frequently encountered in the middle aged, especially in those in the 4th decade of life (8, 15). An exacerbation of symptoms related to pregnancy has been described (7).

Extracerebral cavernous hemangiomas may origi-

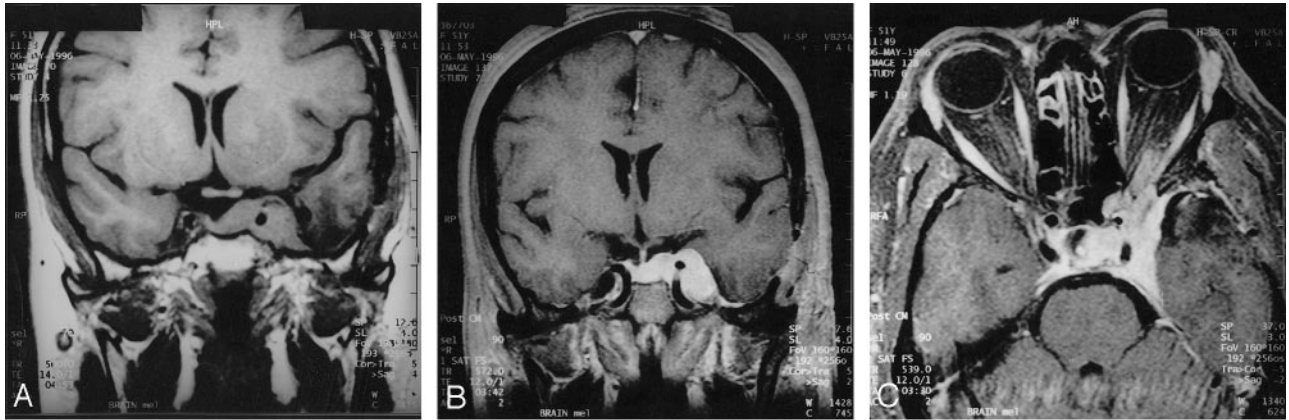


FIG 3. Meningioma.

A, Axial T1-weighted image demonstrates a hypointense left extraaxial parasellar lesion.

B, Initial coronal gadolinium-enhanced T1-weighted image demonstrates intense uniform enhancement of the lesion.

C, Subsequent axial gadolinium-enhanced T1-weighted image demonstrates similar uniform enhancement characteristics in contrast to the heterogeneous centripetally "filling-in" enhancement pattern exhibited by the cavernous hemangioma.

D, Three-hour axial-labeled red cell blood pool SPECT images demonstrate a photopenic left cavernous sinus region.

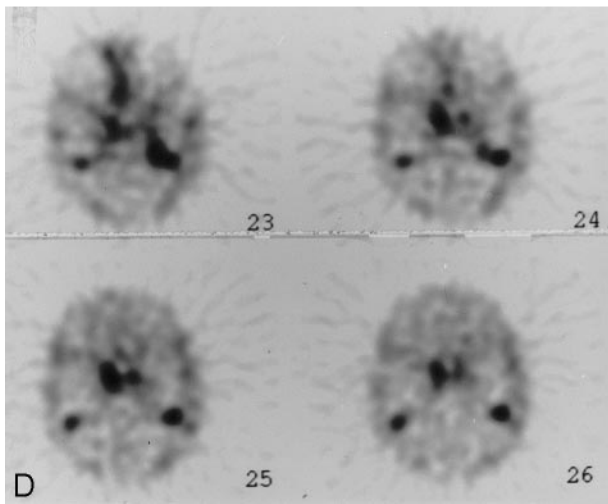
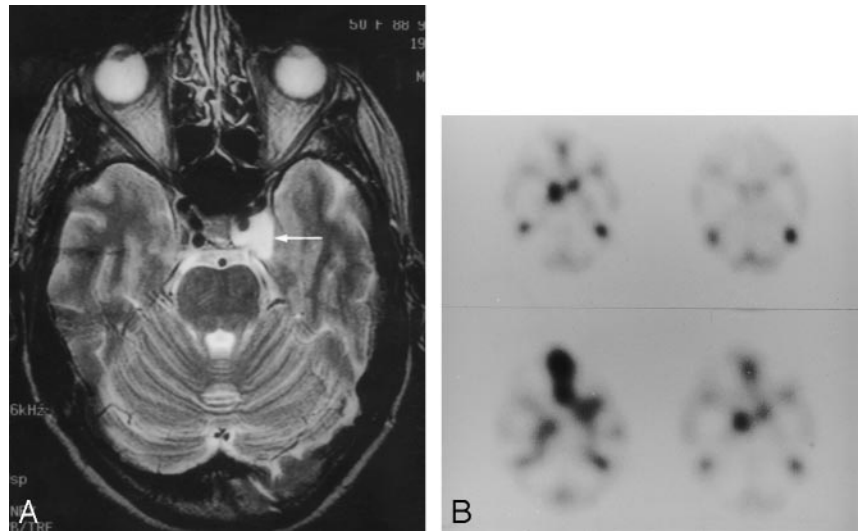


FIG 4. Chordoma.

A, Axial T2-weighted image demonstrates a hyperintense left extraaxial parasellar lesion (white arrow).

B, Three-hour axial-labeled red cell blood pool SPECT images demonstrate a photopenic left cavernous sinus region, appearance similar to Figure 3D.



nate from components of the cavernous sinus itself (intracavernous) or from the surrounding tissues (extracavernous) (3, 11). These non-neoplastic lesions may grow slowly, with progressive enlargement of thin-walled vascular channels, producing compression on adjacent retro-orbital neural structures and resulting in the insidious onset of slowly progressive symptoms (3, 11). Presenting complaints of headaches, retro-orbital pain, and multiple cranial nerve palsies

with ophthalmoplegia have been described (3, 6, 8, 10). Other symptoms include anisocoria, proptosis, obesity, amenorrhoea, and sensory changes in the face (2, 9, 13, 16).

Both extracerebral and intraparenchymal cavernous hemangiomas have similar histopathologic appearances (17); these are homogeneous, well-defined masses composed of numerous vascular channels of variable size and lined by a single layer of endothelial

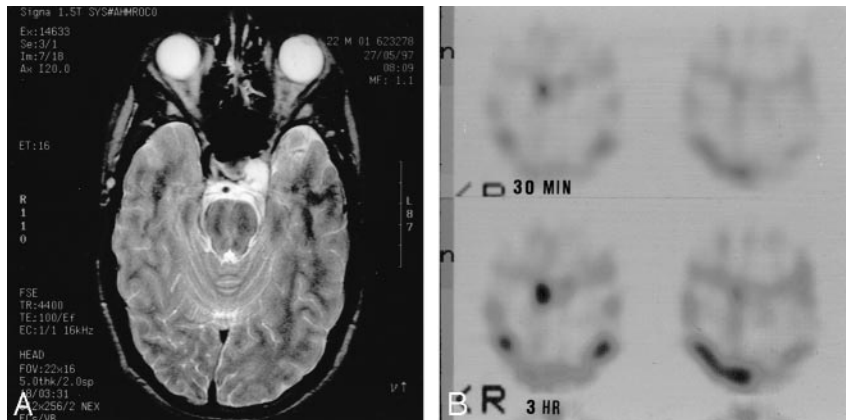


FIG 5. Chondrosarcoma.

A, Axial T2-weighted image demonstrates a hyperintense left extraaxial parasellar lesion.

B, Axial labeled red cell blood pool SPECT images at 30 minutes (top) and 3 hours (bottom) demonstrate a photopenic left cavernous sinus region, appearances similar to Figures 3D and 4B.

cells with no elastic membrane or smooth muscle cells present. The walls are closely contiguous, composed of collagen and endothelial cells, and have no cellular stroma beyond the limiting walls of the vascular spaces. Unlike capillary telangiectasia, there is no intervening neural tissue in these lesions. Hyaline degeneration, thrombosis, cholesterol clefts, calcification, and hemorrhage may be seen in extracerebral cavernous sinus hemangiomas (1, 2, 4, 10–12, 18). These lesions usually do not have well-formed supplying or draining vessels, although there may be nearby tortuous feeding arteries, draining veins, or superficial vascularization (2).

Extracerebral cavernous sinus hemangiomas are rare, comprising less than 1% of all parasellar masses (1). More common cavernous sinus masses that may also have sphenoid bone involvement include meningioma, chordoma, chondroma, chondrosarcoma, metastasis, and lymphoma. Further cavernous sinus masses that have been reported include epidermoid, neurofibroma, schwannoma, sarcoid granuloma, aspergilloma, Wegener granulomatosis, chemodectoma, Tolosa Hunt syndrome, and thrombosis of the cavernous sinus or an aneurysm. In practice, extracerebral cavernous sinus hemangiomas are most frequently misdiagnosed as meningiomas on the basis of imaging findings (4, 7, 11, 18).

The need for accurate preoperative diagnosis of extracerebral cavernous sinus hemangiomas is important, because these lesions are notoriously difficult to surgically excise completely because of their location, propensity for profuse bleeding, and relationship to complex neurovascular structures (2, 6–9, 14, 15). Surgical mortality rates of as much as 25% and rates of complete resection as low as 16% have been reported (6). There are risks of operative damage to the internal carotid artery and the cranial nerves within the cavernous sinus during resection (8).

Preoperative radiation therapy to the involved cavernous sinus has been used in an attempt to reduce the extent of intraoperative hemorrhage during resection with doses as high as 30 Gy. In one study, some lesions not presenting with severe symptoms were treated solely with radiation therapy (15).

Skull radiographs are of limited utility in the diagnosis of extracerebral cavernous sinus cavernous hem-

angiomas; findings include enlargement or erosion of the sella turcica, greater wing of sphenoid, petrous apex, or clinoid processes (2, 5, 6, 13–15, 17, 18).

Noncontrast CT may demonstrate a well-defined isoattenuated to hyperattenuated mass in the parasellar region with marked homogeneous enhancement following contrast material administration (2–6, 8–12, 15, 17, 18). Lesion calcifications are unusual (11). The cavernous hemangioma may extend from the cavernous sinus into the sella turcica and demonstrate erosion of adjacent bony structures. The CT pattern of enhancement between extracerebral cavernous sinus cavernous hemangiomas and meningiomas has been described as indistinguishable (7).

Digital subtraction angiography (DSA) findings may be normal (3, 8, 10, 18), may manifest as an avascular mass producing obstruction of the cavernous sinus (10–12, 17, 18), or may demonstrate tumor staining with delineation of feeder arteries arising from the cavernous portion of the internal carotid artery, which may be displaced by the lesion (5–7, 9, 13, 14, 17). The venous phase may demonstrate a faint oval shadow of the lesion (15). At DSA, Simard et al (2) demonstrated four extracerebral cavernous sinus cavernous hemangiomas as avascular masses, whereas seven lesions demonstrated tumor staining.

MR imaging is the imaging investigation of choice owing to its elegant demonstration of the lesion and its relationship to surrounding structures (3). At MR imaging, extracerebral cavernous sinus cavernous hemangiomas are well-defined masses with low to isointense signal on T1-weighted images and marked hyperintensity on proton density- and T2-weighted images (3, 4, 6, 8, 9, 12, 13, 16, 17). In contrast to intraparenchymal cavernous hemangiomas, extracerebral cavernous sinus cavernous hemangiomas do not demonstrate a T2 hypointense hemosiderin rim (6). The extradural location of the mass involving the cavernous sinus is well shown by MR imaging with demonstration of the lateral border of the cavernous sinus (6, 7, 13). Homogeneous enhancement following administration of gadolinium has been described (4, 7, 12, 17).

Little published information regarding nuclear medicine imaging is available. In an early case report, Kawai et al (14) described an extracerebral cavernous

sinus cavernous hemangioma as a large area of increased activity in the middle cranial fossa on a ^{99m}Tc -pertechnetate brain scan.

In this case report of presumed extracerebral cavernous sinus cavernous hemangioma, the MR imaging findings were a T1 hypointense and marked proton density and T2 hyperintense left parasellar mass. Following administration of gadolinium-DTPA, there was a heterogeneous pattern of enhancement with slow centripetal "filling in" of the lesion in the time period between the initial coronal and subsequently obtained axial images (Fig 1C and D). This finding was not observed in the pathologically proved cases of meningioma and chondrosarcoma, which both demonstrated a rapid uniform enhancement pattern with no significant change in appearance between the initial axial and subsequent coronal images (Fig 3B and C).

The ^{99m}Tc -pertechnetate-labeled red cell blood pool study of the brain did not reveal a hypervascular mass on the dynamic flow images. Sequential planar images taken at 60-second intervals for a period of 4 minutes demonstrated slowly progressive accumulation of red cells in the left cavernous sinus region. Delayed SPECT images at 30 minutes and 3 hours demonstrated persistent increased activity in the region of the left cavernous sinus, more intense than the normal contralateral right cavernous sinus. By comparison, the SPECT images of the red cell blood pool scintigrams in the histologically proved cases of meningioma, chondrosarcoma, and chordoma demonstrated persistent photopenic lesions, consistent with replacement or compression of the involved cavernous sinus by tumor.

The findings of marked T2 hyperintensity, centripetal enhancement pattern on sequential gadolinium enhanced images, and the positive labeled red cell blood pool study of this left cavernous sinus lesion are analogous to the imaging findings encountered in cavernous hemangiomas arising in other organs, such as the liver. The true accuracy of this examination may be determined as further vascular lesions, such as chemodectoma, are assessed and reported in the literature. The specific lesions most likely to be confused with a cavernous hemangioma are sufficiently rare that further progress in determining the accuracy of this test is likely to require the input from multiple institutions.

In view of the above findings, the planned surgery was canceled and the patient was treated conservatively. Over a period of six 6 years, the left parasellar lesion in this case report has not altered in size, signal intensity, or contrast material enhancement pattern. Although there is no histologic proof of the nature of this lesion, its unchanged appearance would mitigate against it being an aggressive meningioma, chordoma, or chondrosarcoma. Furthermore, in view of the significant morbidity and mortality encountered at surgery for extracerebral cavernous sinus cavernous hemangiomas, the prospective diagnosis by using the combination of MR imaging and red cell blood pool

scintigraphy of the brain has been of potentially enormous benefit to the patient.

Conclusion

Extracerebral cavernous sinus cavernous hemangiomas are rare lesions that may be confused with meningiomas at CT and MR imaging. A preoperative diagnosis is imperative so that the use of surgery in these lesions, which is often complicated by incomplete removal, hemorrhage and significant operative morbidity and mortality rates, may be carefully considered.

A centripetal "filling-in" appearance following administration of gadolinium contrast medium in a markedly T2 or proton density hyperintense parasellar lesion should suggest the diagnosis. Increased persistent activity on both early and delayed red cell blood pool scintigraphy of the brain, analogous to the findings seen in hepatic cavernous hemangiomas, is likely to be confirmatory in establishing the nature of the lesion thus potentially avoiding hazardous biopsy or surgery.

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