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Anomalous Intracranial Venous Drainage Mimicking Orbital or Cavernous Arteriovenous Fistula

Kurt E. Tech, Cristie J. Becker, Alfredo Lazo, Thomas L. Slovis, and I. M. Rabinowicz

Summary: A 9-year-old child presented with right eye swelling and bluish discoloration over the periorbital region. MR and CT displayed prominent vessels within the orbit and in the facial region, suggesting a carotid-cavernous fistula. Doppler ultrasound indicated the vascular engorgement was not secondary to a proximal arteriovenous shunt. Arteriography with its inherent time sequence image capability prompted the proper diagnosis of bilateral atresia of the sigmoid sinuses.

Index terms: Dural sinuses; Veins, cerebral; Orbits, abnormalities and anomalies; Pediatric neuroradiology

There are numerous possibilities as to the locations of congenital anomalies and variants involving the transverse and sigmoid venous sinuses, the most common of which involve septation of the transverse sinus near the torcular (1). This report presents a case of bilateral sigmoid sinus hypoplasia/aplasia with the majority of the cerebral venous drainage occurring through the right cavernous sinus and diploic emissary veins. There was sufficient collateral flow through the right superior ophthalmic vein to mimic an arteriovenous fistula or malformation of the right orbit.

Case Report

The patient was a 9-year-old boy with increasing headaches in the right frontal and periorbital region with right eye swelling. He also reported an intracranial noise like "a sea shell against the ear." The patient has a history of right-sided glaucoma treated with topical medication. There was no history of seizures. The mother first noticed a bluish appearance to the skin around his right orbit when the boy was 4 ½ years of age. Because of proptosis and decreased vision in the right eye, as well as a bruit (described as a "hum"), a provisional diagnosis of arteriovenous fistula or malformation was given and a cerebral arteriogram was performed at 6 years of age. A diagnosis of right orbital malformation (nonarterial) was given to the patient.

The current physical examination revealed right periorbital and upper facial swelling. Nonpitting edema over this region was elicited. The previously described decreased vision improved and was nearly equal to the left eye. A slight esotropia was apparent along with a small 2-mm right proptosis. A very soft bruit was suggested over the eye. Funduscopic exam on the right revealed venous tortuosity and mild engorgement with a slight increase in the size of the optic nerve cup; no aneurysms or papilledema was observed. Right ocular pressure was somewhat elevated.

A color Doppler ultrasound of the right orbit displayed large nonpulsatile veins in the orbit superior and posterior to the globe (Fig 1). Postcontrast computed tomography (CT) of the orbits demonstrated bilaterally enlarged cavernous sinuses and a large right superior ophthalmic vein, as well as prominent superficial facial veins (Fig 2). Magnetic resonance (MR) revealed prominent veins in the periorbital soft tissue and the right orbital cone (Fig 3).

Selective right and left internal carotid, left vertebral, and right external carotid arteriograms were performed. The internal carotid arteries were normal. The right ophthalmic artery was also normal, with no early filling of veins. There was no opacification of the sigmoid sinuses (Fig 4A). The pattern of drainage on the right was altered so that flow from the superior sagittal sinus entered superficial cortical veins, the sphenoparietal sinus, and the cavernous sinus. The cavernous sinus on the right was dilated along with the right superior ophthalmic vein and numerous collateral periorbital veins (Fig 4B). The periorbital veins drained into a dilated angular vein and via facial veins, into the right external jugular vein (Fig 4C). Additionally, the sagittal sinus drained via diploic emissary veins into scalp veins with later visibility of the right superficial temporal vein. In the arterial phase, there was no evidence of arteriovenous shunt in the cavernous sinus (Fig 4D). The venous pattern on the left carotid injection had a more extensive collateral circulation externally via diploic emissary veins, as well as some

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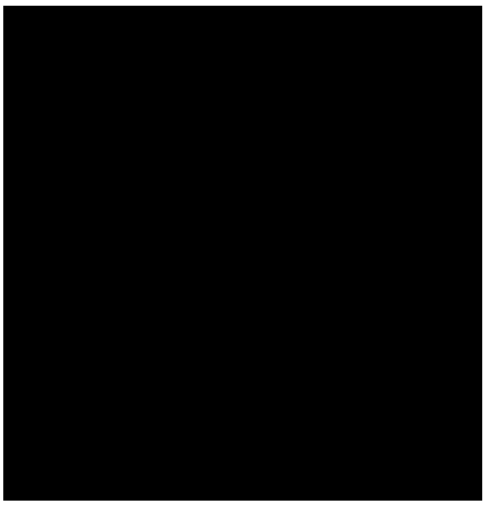

Fig 1. Color flow ultrasound of right superior orbit region. The top of the image is nearest the transducer immediately adjacent to the globe. The nose is in the upper right corner (*asterisk*). The red and blue areas represent engorged vessels (*hooked arrows*), which demonstrated venous type pulsations when examined with Doppler.

retrograde flow seen in the left superior ophthalmic vein; however, this was not to the same degree as on the right. Drainage from the left hemisphere into the right cavernous sinus was also demonstrated.

Discussion

Marked narrowing or atresia of the sigmoid sinus is rare (1). More commonly, incomplete or complete septation of the transverse sinus is observed particularly near the torcular. When there is obstruction at the level of the sigmoid sinus or lateral transverse sinuses, venous drainage occurs via three routes: (*a*) collateral mastoid emissary veins or posterior condyloid emissary veins; (*b*) the opposite lateral sinus if open; and (*c*) anterior drainage into the Galenic system or into the superior sagittal sinus (1).

Fig 3. Sagittal T1-weighted MR image (500/30/4 [repetition time/echo time/excitations]). Signal void is demonstrated in the periorbital soft tissue (*black arrow*) and in the right orbit cone (*curved arrow*).

When considering the differential diagnosis, another consideration would include bilateral thrombosis of the sigmoid sinuses secondary to inflammation or dehydration. However, this possibility is unlikely because the patient had no history of a debilitating illness, mastoiditis, or meningitis and no thrombosis was seen on MR imaging. Additionally, sigmoid sinus thrombosis has been associated with dural arteriovenous malformations, which have been postulated to be an attempt at revascularization (2–5); in these patients neurologic deficits, increased intracranial pressure, hydrocephalus, seizures, and hemorrhage often develop (2, 5–9). Our patient had none of these symptoms or physical signs described previously.

Fig 2. Contrast-enhanced CT showing enlarged right cavernous sinuses (*dark arrows*) and superior ophthalmic vein (*curved white arrow*). Also noted are engorged veins (*small straight arrows*).

Fig 4. *A*, Left internal carotid artery injection, venous phase; both transverse sinuses are apparent (*small black arrows*) with no sigmoid sinuses demonstrated. The cavernous sinus is seen (*small black arrow*). Collateral diploic emissary veins are demonstrated (*straight white arrows*) along with engorged supraorbital facial veins (*small straight arrow*).

B, Right internal carotid artery injection, submentovertex and lateral views of venous phase; cavernous sinus (*black arrows*) and right ophthalmic vein (*curved white arrow*) engorgement is apparent. Collateral diploic emissary veins are again demonstrated (*straight white arrows*).

C, Left internal carotid artery injections, late venous phase; diploic emissary veins (*small arrows*) filling the superficial temporal vein (*large arrows*).

D, Right internal carotid artery injection, arterial phase; no carotid-cavernous fistula demonstrated.

At approximately 2 years after our radiologic evaluation, the patient has had no progression of symptoms. The patient's family elected conservative treatment and, therefore, no surgery was performed.

This case is unusual in that the sigmoid sinus atresia is bilateral. Because the findings on MR and CT mimic those of an arteriovenous fistula or malformation, the necessity of angiography for accurate diagnosis is reinforced. As MR angiography becomes more advanced, it may be possible to arrive at the definitive diagnosis of obstructed sigmoid sinuses as opposed to a malformation. However, it is important to note that this diagnosis was confirmed with "time sequence" evaluation of the conventional arteriogram images. Specifically, the appearance of an enlarged right cavernous sinus and superior ophthalmic vein was apparent late in the angiography run, excluding the possibility of a carotid-cavernous fistula. This is the current advantage of conventional angiography over MR angiography. Ultrasound was a useful, but not definitive, adjunct in this case.

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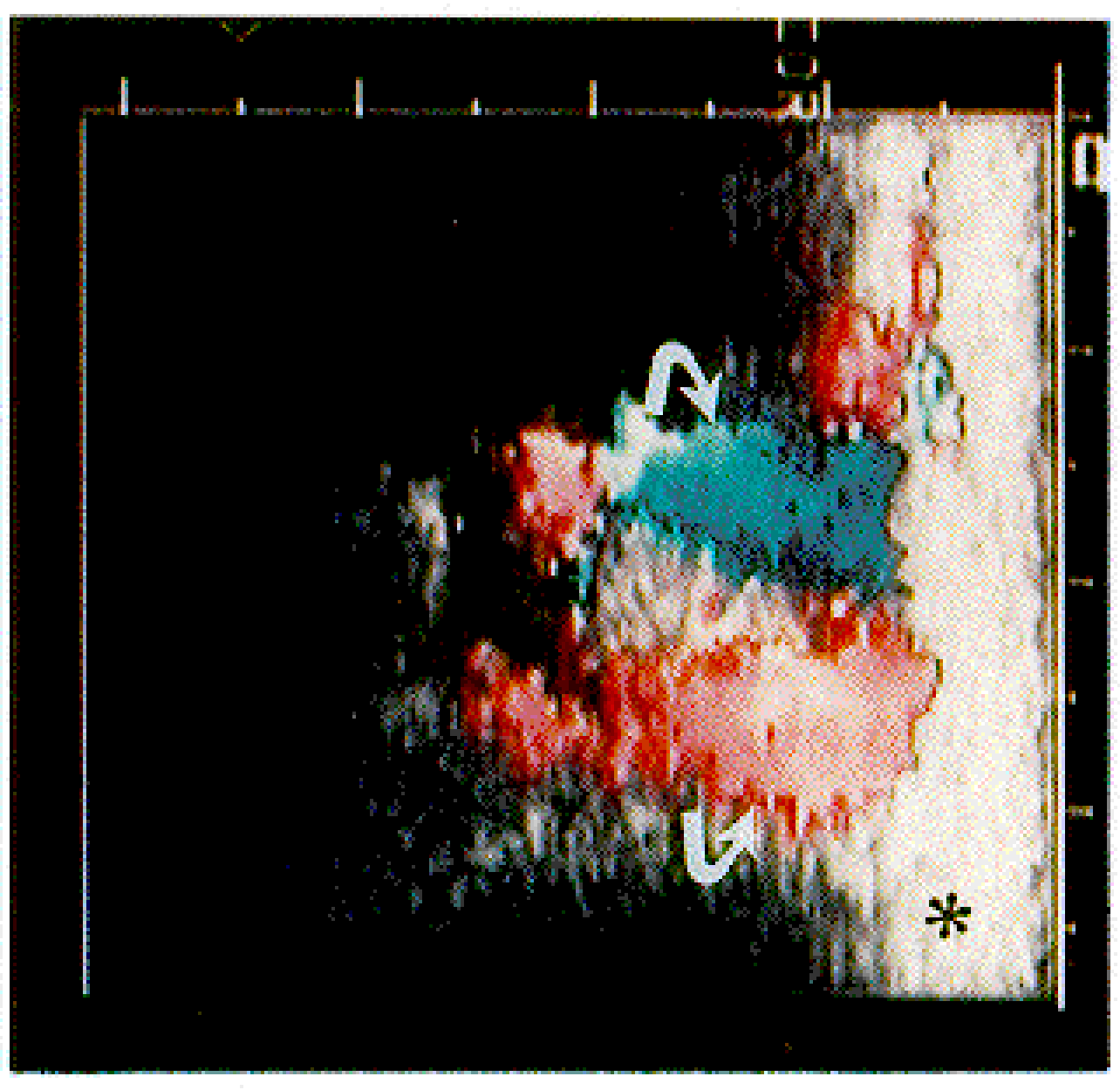

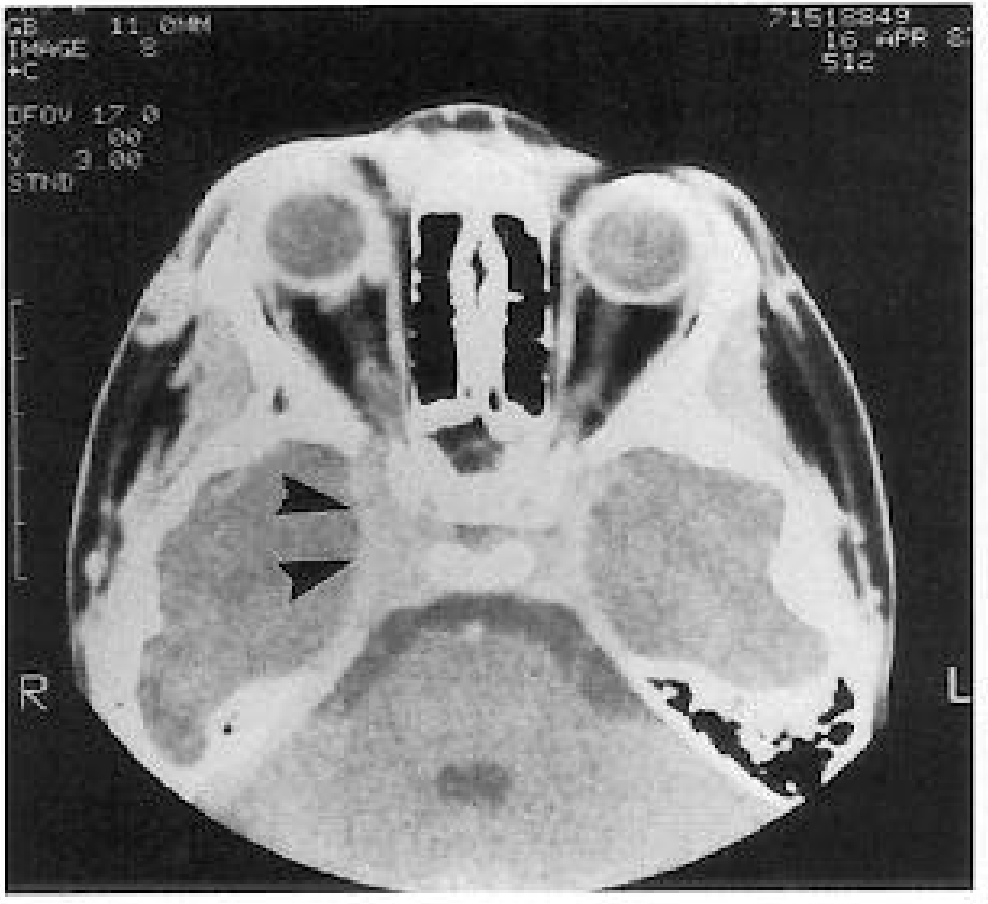
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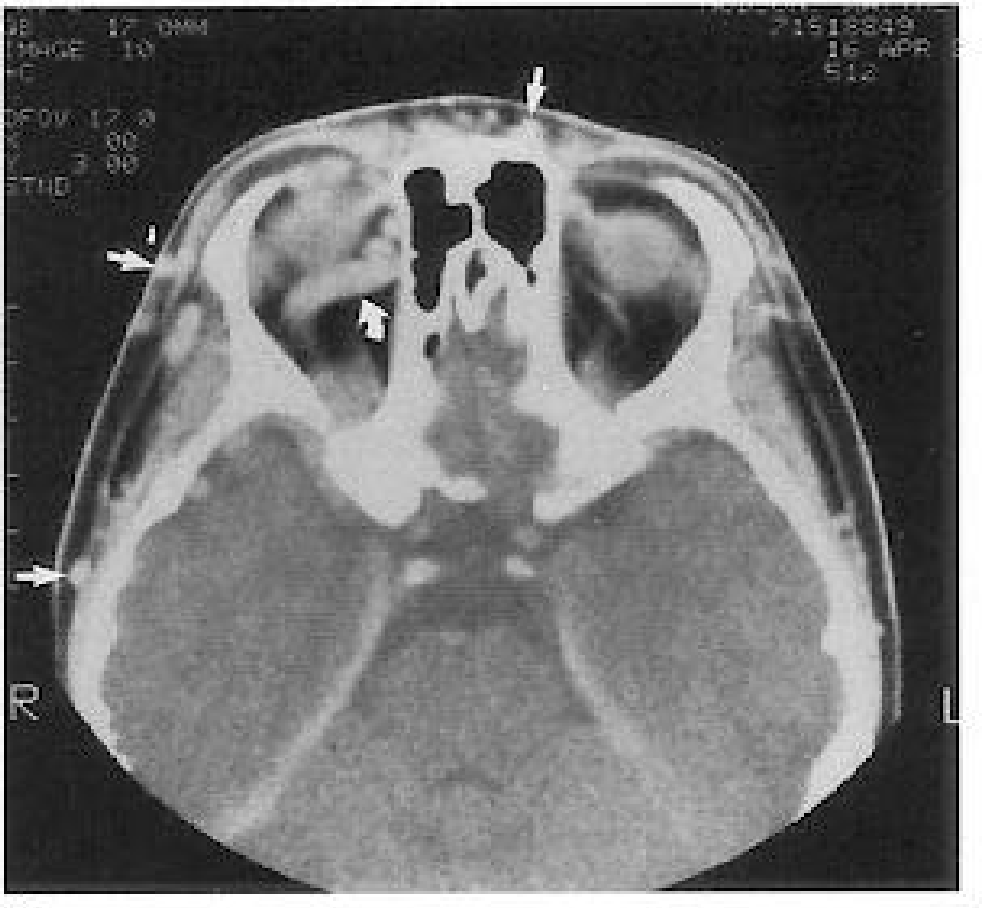
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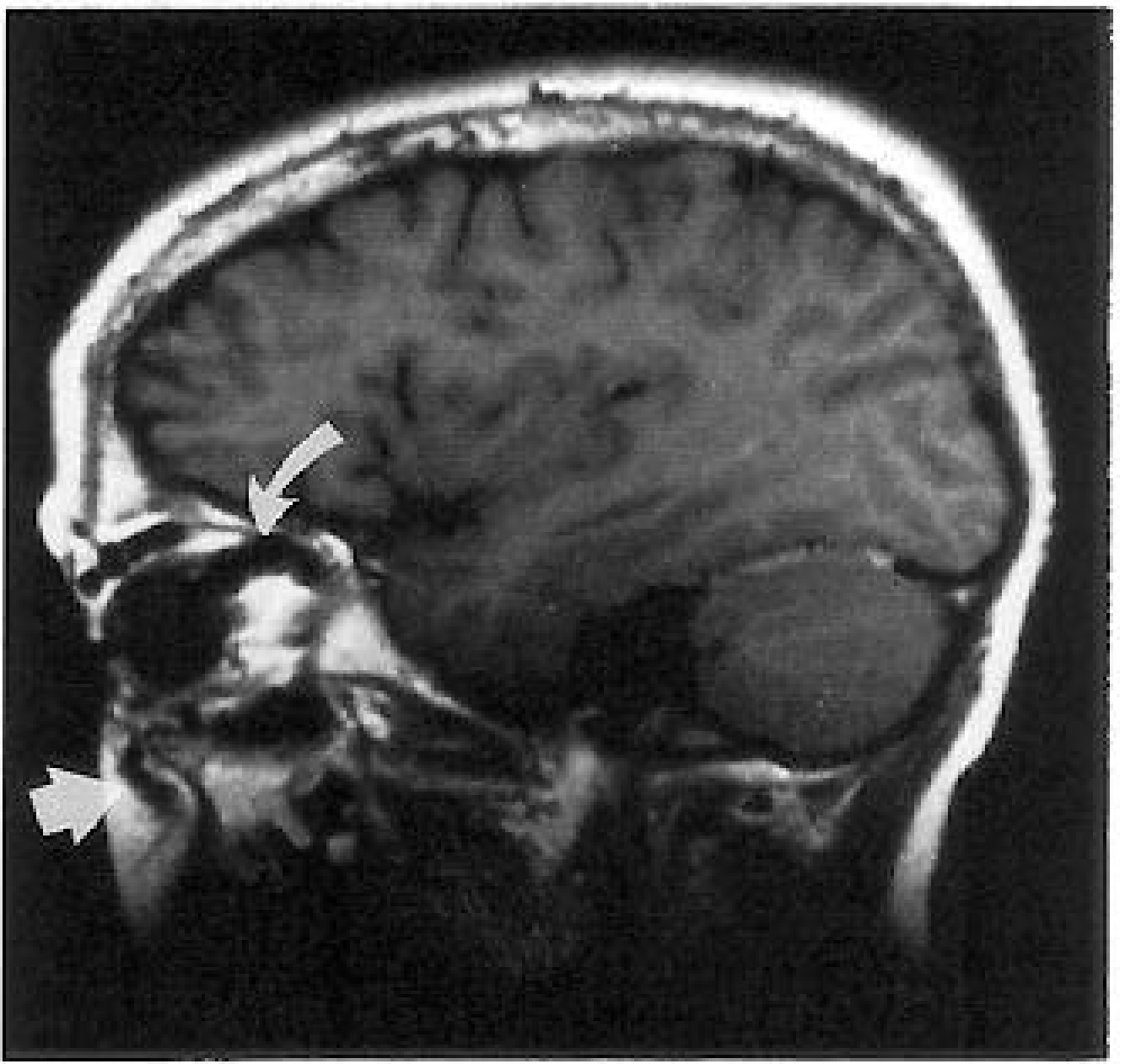

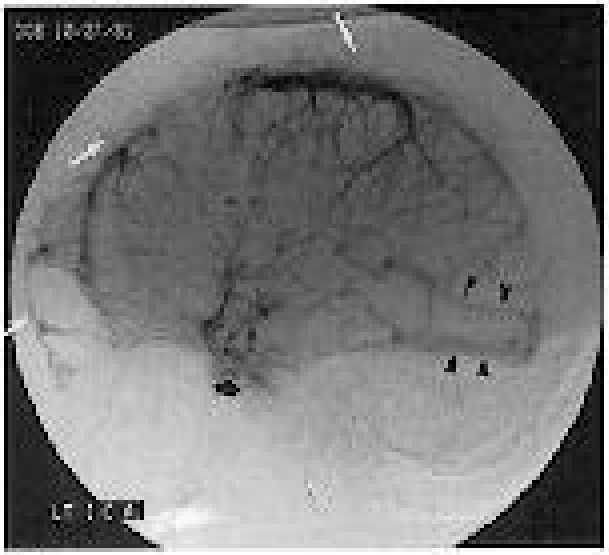

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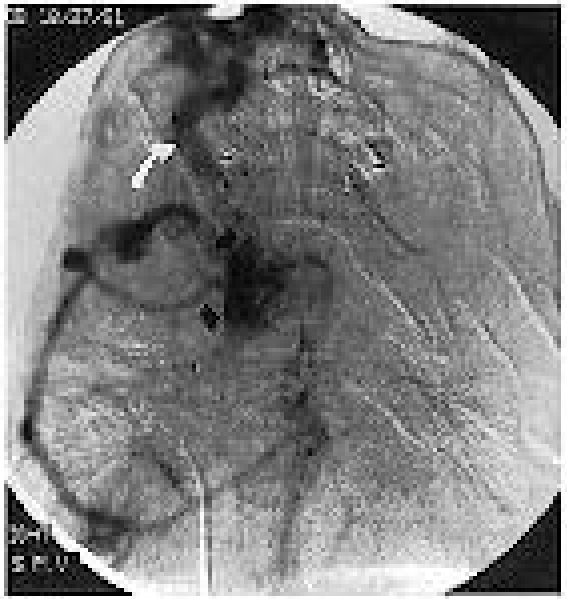

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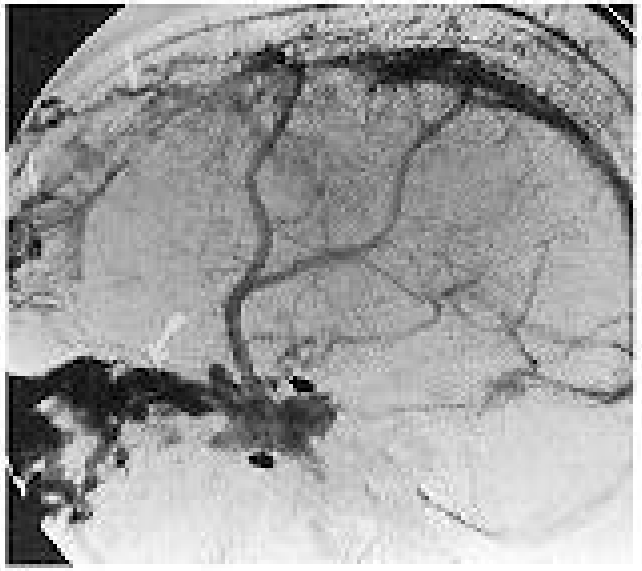

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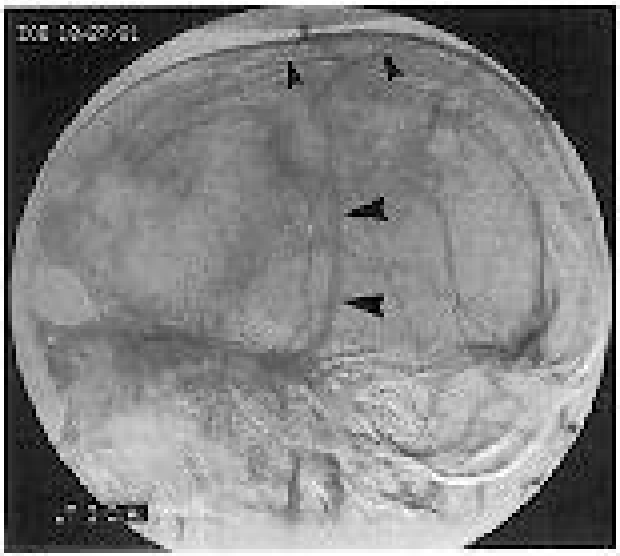

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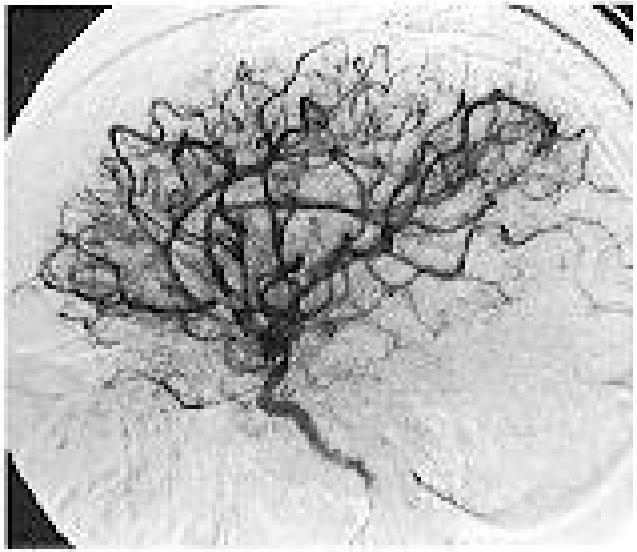

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