AJNR

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AJNR Am J Neuroradiol 1983, 4 (3) 741-744
http://www.ajnr.org/content/4/3/741

Cavernous Hemangiomas of the Orbit: Value of CT, Angiography, and Phlebography

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Neuroradiologic studies performed in 18 cases of surgically verified intraorbital cavernous hemangioma are reported. Skull films usually showed enlargement of the orbit and evidence of soft-tissue mass. Phlebography rarely demonstrated filling of the cavernous hemangioma or enlarged draining veins. On angiography, in addition to displacement of vessels, pooling of contrast medium in the cavities of the hemangioma was observed in more than half the cases. Computed tomography (CT) demonstrated a rounded, hyperdense, enhancing lesion usually in the superior segment of the intraconal space. Although CT may be sufficient for planning the surgical approach, in most cases a combination of CT and angiography will give better, more specific preoperative diagnosis.

Cavernous hemangiomas are by far the most common vascular malformation of the orbit and are among the most common of all the mass lesions that develop within the orbit [1, 2]. In 256 consecutive operations performed on the orbits at the Istituto Neurologico of Milan, 18 (7%) cavernous hemangiomas were found. Excluding orbital involvement of intracranial meningiomas, they were (together with the heterogeneous group of lacrimal gland tumors) the most common lesion.

Computed tomography (CT) has enormously increased diagnostic capabilities in the pathology of the orbits; however, reports of the pre-CT era have suggested the possibility of a specific diagnosis of cavernous hemangiomas of the orbit by angiography and phlebography [3, 4]. We reviewed the radiologic findings in a series of 18 cavernous hemangiomas of the orbit.

Materials and Methods

Eighteen consecutive surgically proven cases of cavernous hemangioma of the orbit were reviewed. The series included seven men and 11 women. The average age was 44 (range, 11–72), with 15 cases concentrated from 31 to 58 years of age. The length of the histories varied from a few weeks to 30 years (average, 5 years). Exophthalmos was the presenting symptom in all but two patients. This was accompanied by decreased visual acuity, largely due to hyperopia caused by the indentation of the globe [1], and rarely by central scotoma.

All the patients had skull films, sometimes supplemented by tomograms; one radionuclide study was positive. Thirteen patients had orbital phlebography performed by puncture of the frontal vein. Internal or common carotid angiography was performed in 14, magnified × 2 in nine cases. In four cases selective external carotid injection was performed. CT was available in the last 14 cases. It was usually performed with an EMI 1010 scanner with 5 mm cuts, a 320 × 320 matrix, and, when feasible, with coronal cuts. The radiologic findings were compared with the description of the surgical intervention.

Results

Skull films were completely normal in three cases, and in two others only minimal enlargement of the involved orbit was observed. Definite enlargement was found in 11, often with evidence of increased soft-tissue density. In two cases, enlargement and poor definition of the margin of the optic canal were observed. In one of these the cavernous hemangioma, 4 mm in diameter, had grown within the optic nerve at the apex of the orbit. Enlargement of the superior orbital fissure was found in two cases. In one of these the cavernous hemangioma (already reported [5]) had eroded the anterior clinoid, growing both into the orbit and into the middle cranial fossa, but remaining extradural. Calcifications with the appearance of phleboliths were found in two cases, but in one of these they indicated the position of a hemolymphangioma coexisting with the cavernous hemangioma.

Orbital phlebography was abnormal in all 13 cases. In 10, displacement of orbital veins was observed on the involved side, sometimes accompanied by compression and poor filling of the veins adjacent to the cavernous hemangioma. In the case involving the anterior clinoid with intracranial growth, lack of filling of the posterior part of the superior ophthalmic vein proximal to the apex of the orbit and of the anterior part of the homolateral cavernous sinus was observed. In two cases varicose dilatations close to the superior ophthalmic veins were present, while in two other cases filling of the cavities of the cavernous hemangioma with sedimentation of the contrast medium for several seconds was observed (fig. 1).

Internal carotid angiography was normal in only two cases. In the other 12 cases stretching and displacement of the branches of the ophthalmic artery were present. In eight of these, pooling of the contrast medium in one or a few small cavities of the cavernous hemangioma, with sedimentation through the venous phase, was demonstrated (fig. 2).

External carotid angiography was always normal. It was obtained either by selective injection (in four cases) or by injection of the common carotid artery using stereo views (in several others).

In 11 of the 14 cases in which it was performed, CT demonstrated

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AJNR 4:741–744, May/June 1983 0195–6108/83/0403–0741 $00.00

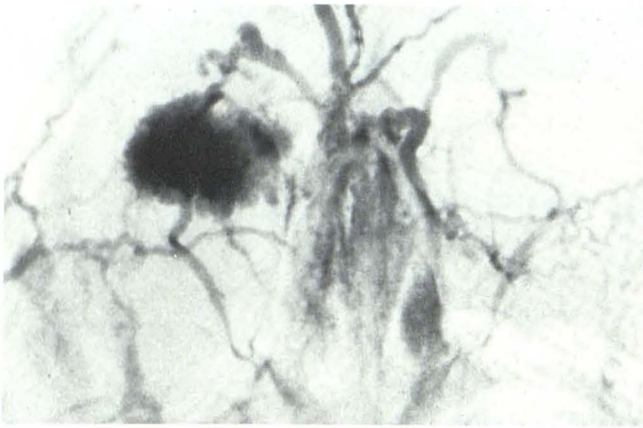


Fig. 1.—Orbital phlebography. Retrograde filling of intraconal cavernous hemangioma of right orbit.

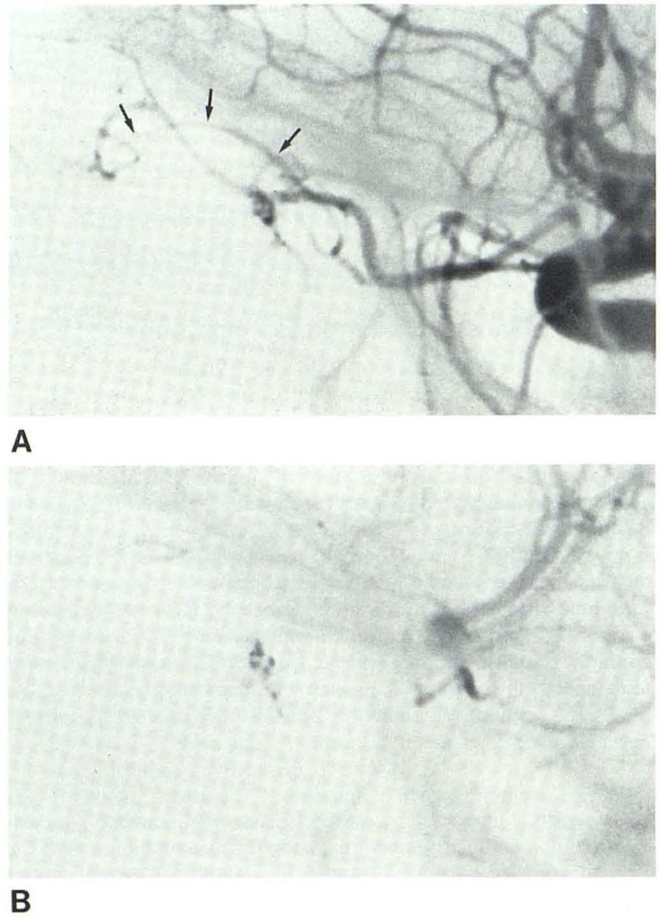


Fig. 2.—Carotid angiography. **A**, Stretching and displacement of lacrimal branch of ophthalmic artery (*arrows*) surrounding cavernous hemangioma. **B**, Pooling of contrast medium in several cavities at posterior pole of hemangioma.

an intraconal rounded lesion, homogeneous and with smooth margins, slightly hyperdense with respect to the optic nerves and extraocular muscles, homogeneously enhancing to various degrees (figs. 3 and 4). In another case CT demonstrated a large lesion that occupied all the retrobulbar space; the extraocular muscles were not distinguishable. At operation this cavernous hemangioma was also found to be intraconal.

CT was available in two other cases. In a small cavernous hemangioma, partially embedded in the optic nerve at the apex of the orbit, CT demonstrated only a slightly enlarged and tortuous optic nerve. In the intraorbital-intracranial cavernous hemangioma, CT showed erosion of the lesser wing of the sphenoid by the enhancing lesion, which protruded both into the apex of the orbit, compressing the optic nerve, and into the middle cranial fossa [5].

Of the intraconal lesions, CT demonstrated a medial or superomedial lesion in three cases, a superolateral lesion in three, a central superior lesion in two, and a lesion inferior to the optic nerve in one. In one case the lesion was in the central part of the intraconal space, apparently surrounding the optic nerve. In two cases with large lesions, the cavernous hemangioma occupied the whole intraconal space. In these two cases the lesion reached the apex of the orbit, while in all the other cases the posterior pole of the cavernous hemangioma was clearly demonstrated and the apex of the orbit appeared free (fig. 3). Demonstration of the displaced optic nerve was obtained only in the cases with rather small lesions (fig. 5). In nine cases the orbit was enlarged, and in only one case was a small calcification demonstrated within the cavernous hemangioma.

The review of the operative reports demonstrated a perfect correlation between the location of the lesion demonstrated by radiologic studies and that found at surgery, with obvious superiority of CT over both angiography and phlebography in depicting the boundaries of the lesion. Phlebography was in fact quite inaccurate in demonstrating the size of the lesion, and sometimes uncertain also about its location. The only difference between surgical and radiologic findings regarded the size of the lesion; surgical reports sometimes described a smaller size for the cavernous hemangioma compared with that observed on CT.

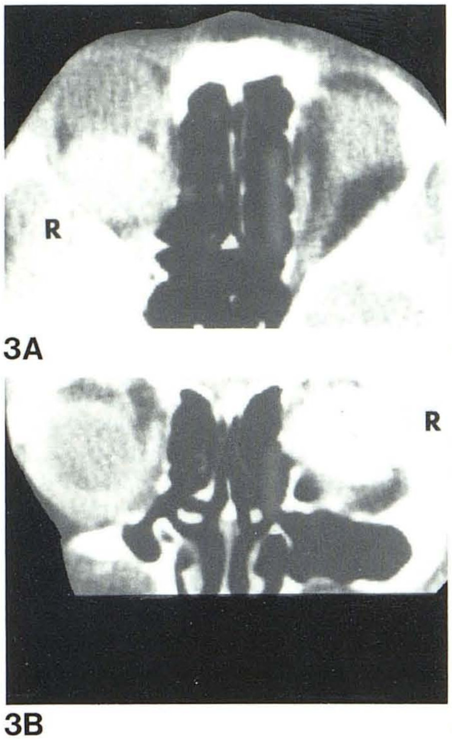
On the whole, 15 cases were intraconal (all but four in the superior part of this space); one was embedded in the optic nerve; one was intraorbital, intraosseous, and intracranial; and one developed superiorly to the ocular globe, extending into the eyelid. This patient was also operated on for a coexisting retrobulbar hemolymphangioma.

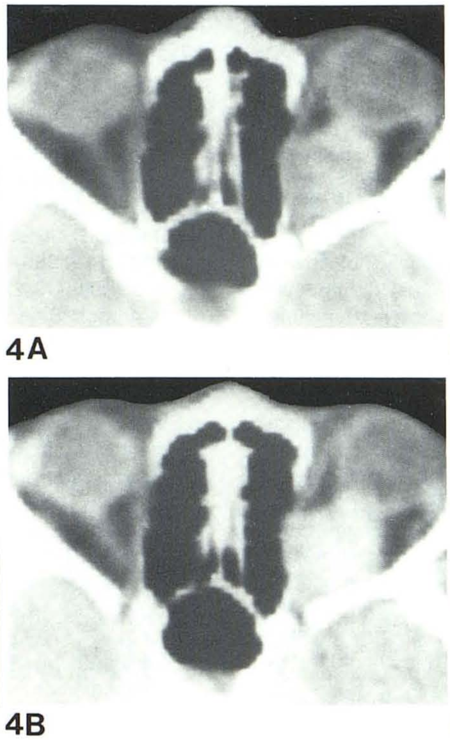
Discussion

Cavernous hemangiomas of the orbit are common, long-standing mass lesions that usually present in middle age with exophthalmos that progresses extremely slowly over the years [1]. Prevalence in women has been reported [1]. They are second in frequency to arteriovenous malformations in the brain [6], and are the most common vascular malformation of the orbit. They are composed of thin-walled enlarged vascular spaces without elastic membrane or muscular tissue, and are well delineated by a fibrous capsule that usually allows easy, complete surgical removal. Although they are usually detected in middle age, it is likely they have been present since a young age, as indicated by orbital expansion. Orbital enlargement was a very common finding on our skull films, but of little value, as was the demonstration in only two cases of pheboliths that pointed to a vascular pathology.

Radionuclide studies, always positive in our series of intracranial cavernous hemangiomas and in six cavernous hemangiomas of the orbit included in a larger series by Davis et al. [7], were performed in only one case, which was positive.

Phlebography was reported by Aron Rosa and Doyon [4] to have demonstrated frequent partial opacification of the cavernous hemangiomas in their series. Lloyd [8], however, in a series of 10 hemangiomas, found local draining vessels in only one case without





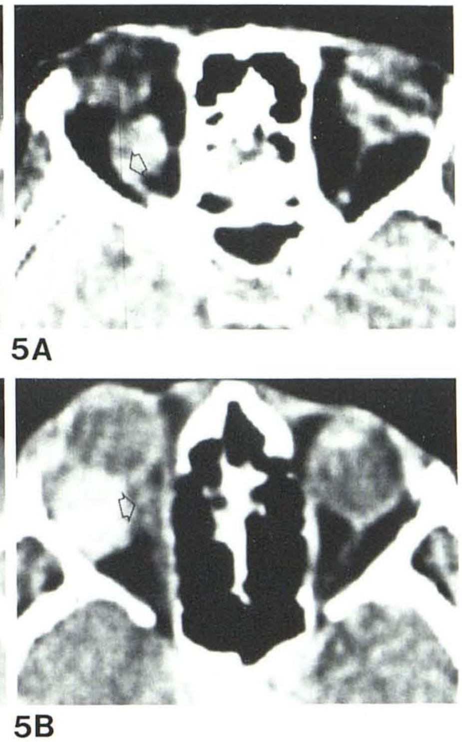


Fig. 3.—Postcontrast CT. Axial (**A**) and coronal (**B**) cuts in large intraconal cavernous hemangioma.

Fig. 4.—Pre- (**A**) and post- (**B**) contrast CT. Intraconal cavernous hemangioma enhances homogeneously. Note enlarged orbit.

Fig. 5.—Different displacements of optic nerve. **A**, Medial cavernous hemangioma displaces nerve laterally (*arrow*). **B**, Large lateral cavernous hemangioma pushes nerve (*arrow*) against medial wall of orbit.

filling of the malformation itself. Our findings lie between these, since, of 13 cases, two had filling of the cavities of the cavernous hemangioma and two had evidence of varicose dilatations that could very well have been part of the draining veins of the hemangioma.

Phlebography may differentiate cavernous hemangiomas from true varices, but differentiation is easily made on clinical grounds. Postural changes affect the exophthalmos in varices, while changes in exophthalmos in cavernous hemangiomas are extremely rare [1]. It is quite possible that the demonstration of the cavernous hemangioma by the venous route is related to the technique, to the quantity of contrast medium injected, and to the length of the series of films.

Differences in technique (amount of contrast medium, rate of injection, and length of the angiographic series) may also account for the variability of visualization of pathologic circulation on carotid angiography. In our series [9] there was a great difference between intracranial cavernous hemangiomas, which very rarely exhibited pooling of contrast medium even with an increased amount and slow injection, and intraorbital cavernous hemangiomas, which showed accumulation of contrast medium in eight (57%) of 14 cases, even with normal injection (fig. 2).

Dilenge [3] reported a constant visualization of angiomatous spots or stains in his cases of cavernous hemangiomas of the orbit. He considered this a typical sign. Davis et al. [7] observed this sign in five of six cases in which angiograpy was performed. In any event, the opacification of cavities of the cavernous hemangiomas is usually greatly inferior to the size of the lesion as demonstrated by vascular displacements and by CT.

The cavernous hemangiomas that involve the bony orbit show a greater vascularity than those located in the intraconal space. In these cases, participation of branches of the external carotid artery is common, while the external carotid artery never supplied tumor stain in our cases located in the usual intraconal position.

The pooling of contrast medium is not unique to cavernous hemangiomas. We have also observed it in cases of hemangiopericytoma. Cases of capillary angioma presented a uniform tumor blush that delineated the whole lesion.

Unfortunately, reports regarding vascular malformations of the orbit frequently contain no precise indication of the histologic type, referring to them all as "hemangiomas." In addition, these malformations sometimes consist of a mixture of several histologic types. Therefore, it is impossible to draw an accurate conclusion about different patterns of pathologic circulation in different types of hemangiomas of the orbit.

CT findings of cavernous hemangiomas of the orbit have been reported by several authors [2, 7, 10]. They all agree that the most common presentation is that of an intraconal, homogeneous, high-attenuation mass with rounded margins, which enhances after contrast medium injection.

Newton (cited in [7]) pointed out the frequency of lack of involvement of the orbital apex. Extension to the apex was found in four of 18 cases in the series reported by Davis et al. [7] and in four in our series, including the small cavernous hemangioma embedded in the optic nerve at the apex of the orbit. Forbes et al. [11] also demonstrated cases of hemangiomas developing at the apex and extending intracranially, but in their series there is no distinction between different histological types.

Within the muscle cone the cavernous hemangioma may be located in any quadrant, but all the series agree on the prevalence of a lateral or superolateral location [1, 7]. In our cases, superomedial location was equally common, with the inferior section definitely being the less involved one.

Enhancement was usually marked and was present in 100% of the cases in all series [7, 9, 12]. In our series, enhancement was demonstrated in all cases, but in two it was minimal. It was not recognizable in the small cavernous hemangioma, which itself was not detectable, being embedded in the optic nerve.

CT appearance of cavernous hemangioma is highly indicative of the diagnosis but not specific, since several benign and even malignant tumors may occasionally present the same CT picture. Davis et al. [7] examined the differential diagnosis of cavernous hemangiomas and lymphangiomas, which are more diffuse and occur at a younger age [1]. In the latter, sonography may help in demonstrating larger lakes and broader septae. Lacrimal gland tumors have a characteristic superolateral and anterior location, are easily palpable, and usually envelop the lateral rectus muscle, and only half of them show contrast enhancement [12].

Neuromas and neurofibromas may grow anywhere in the retrobulbar space and may be indistinguishable from cavernous hemangiomas. Optic nerve sheath meningiomas and optic nerve gliomas frequently extend to the apex of the orbit and even intracranially [13]. In addition to their characteristic pattern of growth, early diminished visual acuity or loss of vision are features that differentiate them from cavernous hemangiomas.

Lymphomas usually do not have well defined margins but rather a diffuse infiltrative aspect, and are often associated with periorbital edema [12]. Metastatic lesions almost never enter into the differential diagnosis. The primary lesion is usually manifest when the orbital metastases occur and destruction of the bony orbit is frequently present.

In conclusion, CT may be considered the only necessary examination since it demonstrates the exact boundaries of the lesion and its relation to the optic nerve, thus providing all the elements necessary to the surgeon for planning the approach. However, a preoperative specific diagnosis may be more frequently reached with the combination of CT and angiography, while the information obtained by phlebography is too limited to recommend its inclusion in the investigation of these lesions.

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