THE VALUE OF ORBITAL ANGIOGRAPHY FOR DIAGNOSIS OF UNILATERAL EXOPHTHALMOS

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(Received for publication August 28, 1961)

It is the aim of the Cushing tradition in neurosurgery to elaborate the preoperative diagnosis of intracranial tumor as accurately as possible. The same should be true for the diagnosis of a space-occupying lesion in the orbit. The value of various roentgenological demonstrations of the orbit is uncontested, but in the opinion of the majority of neurosurgeons angiography has been a disappointing tool for the diagnosis of most orbital lesions. Our personal experience does not support this general opinion. The purpose of this paper is to present our experience in the differential diagnosis of space-occupying orbital lesions by the method of orbital angiography. Curtis, Schurr, Decker, and Decker and Schlegel were amongst the first to describe arteriography of the ophthalmic artery and the choroidal plexus of the eye. But the angiographic demonstration of pathological orbital structures is a field that is still largely unexplored. In their study on unilateral exophthalmos Van Buren et al. showed tracings of arteriograms from 2 cases of proptosis indicating an unusual intraorbital vessel, one of which proved microscopically to be a vein. In addition Yaşargil first reported in a monograph the various roentgen-ray examinations for unilateral exophthalmos, and our own contribution to this subject appeared in 1958.

The disappointing study of most orbital lesions by arteriography, noted especially by Matson and by Gros et al., obviously has been caused by the use of weakly concentrated contrast medium. Since we have been using the 60 per cent contrast medium Urografin, it has been possible to demonstrate the ophthalmic artery in 98 per cent of the cases. Its course as a rule is fairly constant. It is seen to arise from the internal carotid artery as it emerges from the cavernous sinus either at the horizontal portion of the carotid artery or close to the anterior clinoid process (Figs. 1 and 2). According to Schurr, the artery pursues a slightly tortuous course largely because of the freedom of movement which must be allowed to its branches in order to permit rotation of the eyeball. Furthermore it passes along the medial wall of the orbit, at times forming a loop. It is unlikely, therefore, that displacements of the vessel by space-occupying lesions within the orbit would be easy to detect with the exception of tumors arising from the sinuses and penetrating the orbit, which always displace the artery. The recognition of the different branches of the ophthalmic artery and of the choroidal plexus is not easy, especially when it comes to distinguishing them from the outlines of the bone that overlies them. In our experience the stereoscopic views of the angiograms or the study of angiographic pictures produced by the method of Subtraktion according to Ziedes des Planes are a great help in the demonstration of the normal or abnormal vessels. The method of subtraction consists of making a preliminary negative roentgenogram of the skull, from which a positive is made. A second roentgenogram is made after injection of Urografin into the carotid artery. The original positive is now superimposed on the negative arteriogram. The positive of the first film blots out the negative of the second film leaving, in sharp contrast, only the shadows not common to both films, namely, the vessels filled with contrast medium (Fig. 3).
In many instances the venous phase is even more important than the arterial phase. It is obtained either by the late phase of an angiogram of the external carotid or by direct percutaneous injection of the angular vein with contrast medium (Figs. 4–6). This special method, which has been developed by Yaşargil, has some technical difficulties. If the angular vein is very thin it must be punctured with the head in a downward position and with compression of the facial and frontal veins. The course of the orbital veins is to be seen in the phlebogram. The displacement of the superior ophthalmic vein usually is well demonstrated in the anteroposterior view.

CASE REPORTS

Case 1. B.L., a 15-year-old girl, had suffered from gradually increasing protrusion of the right eye for 3 years. There was marked right-sided exophthalmos of 5 mm. (Fig. 7), right papilledema of 6 diopters and right visual loss (0.2). Ocular movements were full. Roentgenogram of the right orbit and right optic foramen did not show anything abnormal. Right carotid angiography revealed a retrobulbar slightly stained tumor surrounded by a thin capsule, the size of a cherry (Fig. 8). The tumor was removed by the lateral,
temporal approach of the orbit. It was attached to the optic nerve which entered the tumor at its posterior pole. It had been cut in the course of complete removal of the tumor. Histologically the tumor was a neurinoma. The postoperative course was uneventful. The right exophthalmos subsided completely, but the patient was blind in her right eye. Vision in the left eye was normal.

Case 2. A.N., a 28-year-old woman, had suffered from moderate protrusion of the right eye for 6 months. There was proptosis of 5 mm. of the right eye (Fig. 9), with right papilledema and visual loss (0.2). Right carotid angiography showed a normally placed right ophthalmic artery and a tumor the size of a cherry behind the eyeball surrounded by a thin arterial vessel in the first phase (Fig. 10) and in the second phase a finely and diffusely stained tumor (Fig. 11). By a right frontotemporal approach the thin lateral wall of the orbit was removed and underneath the superior rectus muscle a cherry-sized round tumor was disclosed and removed. It was slightly adherent to the optic nerve. Histological sections revealed a neurinoma. Three months after operation the exophthalmos subsided completely and the
right optic nerve showed primary atrophy, and some vision for movements of the hand at a distance of 30 cm. remained.

**Case 3.** H.W., a 54-year-old man, had sustained a motorcycle accident with a basal fracture of the skull 4 years before, and had been suffering from tension in the right eye, swelling of the right lower eyelid, and diplopia for 6 months.

He had right exophthalmos of 7 mm. (Fig. 12), upward displacement of the right eyeball, and slight weakness of the 6th nerve. At the base of the orbit a tumor was palpated and tomography revealed destruction of the orbit. In the capillary phase of the right external carotid angiogram (Fig. 13) a huge tumor was seen with peripheral capsular vessels of the internal maxillary artery and upward displacement of the choroidal plexus.

The diagnosis of a periorbital and maxillary tumor was made, and the tumor was removed radically from the maxillary sinus by Prof. Rüedi. Histological sections showed it to be a neurinoma of the right infraorbital nerve.

**Comment.** This group comprising 3 cases of nerve tumors is demonstrated angiographically by an unstained or only very slightly and diffusely stained space-occupying lesion surrounded by very thin peripheral capsular vessels. Such an angiographic finding favors the diagnosis of a benign orbital tumor; it is fairly typical of neurinomas, such as acoustic neurinomas.

**Case 4.** P.H., a 1½-year-old boy, had onset of gradually increasing right-sided proptosis and downward displacement of the right eyeball 8 to 12 months before. On examination there was right-sided exophthalmos of moderate degree, with downward and temporal displacement of the right eyeball (Fig. 14). Roentgenograms revealed lefornity and calcifications in the right orbit. Right carotid angiography demonstrated down-
ward displacement of the peripheral portion of the ophthalmic artery by a retro- and suprabulbar diffusely stained tumor (Fig. 15). The tumor, an ossifying fibroma, was removed by the lateral approach. On control examination 3 years after operation there was no proptosis, but there was definite impairment of upward movement of the right eye.

Comment. The very fine staining of this retro- and suprabulbar tumor was also in favor of a benign lesion.

Case 5. R.W., a 6-year-old girl, had been suffering from right-sided headache, diplopia, and progressive protrusion of the right eye for 4 weeks. There was considerable proptosis of the right eye with downward displacement and impairment of all ocular movements (Fig. 16). Roentgenograms of the skull, including tomography of the orbit, were normal. The right carotid angiogram (Fig. 17) showed a slightly stained retrobulbar tumor, the size of a cherry, without displacement of the ophthalmic artery. In the venous phase of a right external carotid angiogram the facial, angular, nasofrontal and superior ophthalmic veins were demonstrated as well as the downward-displaced choroidal plexus (Fig. 18). At operation (orbitomy) the tumor was found to be well encapsulated and was removed radically. Histologically it was a retinoblastoma.
A postoperative course of roentgen-ray treatment was given, but the child died 1½ years later from metastases.

Case 6. V.d’A., a 2-year-old boy, began to have left-sided parenchymatous keratitis and gradually increasing left-sided proptosis after an alleged injury of the left eyeball with a pencil 8 weeks before. Numerous ophthalmologists were consulted, and a diagnosis of primary or secondary glaucoma and of abscess of the lens was made. Nine months after onset of the illness the left eyeball finally was removed and a prosthesis was implanted. But there was progressive protrusion of the prosthesis, and a retrobulbar tumor or abscess was diagnosed.

The left carotid angiogram (Fig. 19) showed a huge tumor with numerous, very fine pathological vessels in the left orbit and no displacement of the ophthalmic artery. The tumor was removed and proved to be a retinoblastoma. Postoperative roentgen-ray treatment was instituted.

Comment. These 2 cases of retinoblastoma are demonstrated angiographically by a more or less intense staining of the lesion with contrast medium. The vessels are very fine and much more numerous than in the group of neurinomas. This is also the case with neuroblastomas, the malignant tumor of the peripheral sympathetic nervous system, as shown by the following example.

Case 7. S.R., a 7-month-old girl, had suffered from gradually increasing protrusion of the left eye with downward displacement for 7 weeks (Fig. 20). There was a swelling of the left fronto-temporal region with bony destruction of the lateral supraorbital margin. Left carotid angiography (Fig. 21) demonstrated a highly vascular tumor in the left lateral orbital and frontal region with downward displacement of the ophthalmic artery and posterior and upward displacement of the anterior cerebral and frontopolar artery. Biopsy of the tumor revealed a highly malignant tumor, a neuroblastoma. Irradiation was instituted; but the child died 8 months later from the primary abdominal tumor and general metastases in different organs and bones.

Case 8. M.N., a 32-year-old woman, had suffered from diplopia and slight protrusion of the left eye for a few months. There was exophthalmos of 4 mm. with downward displacement of the
left eye (Fig. 22), as well as impairment of adduction and abduction, and hypesthesia in the left trigeminal division. Roentgenograms of the skull and orbit were normal. The left carotid angiogram (Fig. 23) showed a normally placed left ophthalmic artery which gave rise to abnormal, rather thick branches surrounding a tumor the size of a cherry behind the eyeball. The forward displacement of the choroidal plexus is clearly visible. As the general condition of the patient was not good, no operation was performed. The patient died 6 months later from a bronchogenic carcinoma of which the retrobulbar tumor was a metastasis.

Case 9. H.S., a 48-year-old man, had suffered from headache for 2 years; 5 months before admission he was operated on for a metastasis of a bronchogenic carcinoma in the left temporal lobe. Two months later left-sided proptosis developed. On examination there was left-sided exophthalmos of 4 mm. with swelling of the eyelids as well as complete palsy of the oculomotor nerve and partial palsy of the 6th nerve. The phlebogram of the angular vein (Fig. 24) demonstrated a diffusely and roughly stained, retrobulbar tumor, the size of a cherry, and general venous engorgement of the orbit. Roentgen-ray treatment was instituted, but the patient died 1½ months later from the bronchogenic carcinoma.

Case 10. F.M., a 73-year-old male, had noticed gradually increasing swelling of the left eye for 3 months and loss of vision on the left side for 2 months. On examination he had left-sided exophthalmos of moderate degree, and left-sided optic atrophy with impairment of vision (0.3). Roentgenogram of the skull and left-sided carotid

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**Fig. 20. Case 7. Child with neuroblastoma of left orbit.**

**Fig. 21. Case 7. Highly vascular tumor in lateral orbital and frontal region.**

**Fig. 22. Case 8. Patient with left-sided retro-orbital metastasis of a bronchogenic carcinoma.**

**Fig. 23. Case 8. Left carotid arteriogram demonstrating a cherry-sized round tumor behind the eyeball.**
angiogram were normal. The phlebogram of the left angular vein (Fig. 25) showed a roughly stained vascular mass in the superior retrobulbar region of the orbit. Otorhinological examination revealed a tumor of the left ethmoid sinus which histologically proved to be an adenocarcinoma. The tumor was removed, the left eye was enucleated and postoperative roentgen-ray treatment was instituted.

Comment. Malignant tumors differ angiographically from the other groups in that there is a high degree of vascularization of the tumor. The diffusely stained tumor also shows a much rougher pattern than the retinoblastomas.

Case 11. R.T., a 47-year-old woman, had been suffering from right-sided migraine for years and from diplopia and progressive proptosis of the right eye for six weeks. There was moderate exophthalmos of the right eye (Fig. 26), with impair-
ment of outward and upward movement of the right eyeball as well as right-sided papilledema. The phlebogram of the right angular vein demonstrated a medially displaced right superior orbital vein (Fig. 27), and a very finely stained tumor. At operation, the lateral approach of Krönlein, a round, reddish tumor, the size of a cherry, was found in the lateral region of the orbit and was removed radically. Histological sections showed it to be a cavernous hemangioma. On control examination nothing abnormal could be detected; the right-sided exophthalmos has subsided completely.

Case 12. T.d.B., a 23-year-old woman, had been suffering for 3 years from progressive right-sided proptosis which suddenly increased in size during labor. It diminished gradually to a certain extent after birth. But on admission there was still moderate exophthalmos on the right side. There was some stretching of the ophthalmic artery but no evidence of tumor or abnormal vessels (Fig. 28). At operation an angioma, the size of a walnut, was removed completely from the dorsolateral region of the orbit. Histologically it proved to be a cavernous hemangioma. Two months later the right exophthalmos had subsided completely and the ocular movements were full.

Case 13. Ch.Z., a 6-month-old girl, began to have a gradually increasing, bluish, left-sided proptosis 4 months after birth. There was moderate proptosis on the left side with bluish swelling of the eyelids and vascular engorgement of the left temple (Fig. 29). No bruit could be detected. But there was a hemangioma to be seen on the iris. On the left-sided carotid angiogram an enormous vascular neoplasm was detected with increase of vascularity in the capillary and venous phase. The tumor is fed by a huge ophthalmic artery (Fig. 30). No treatment has been given as yet since the condition has remained stationary.

Comment. This group of orbital hemangiomas shows, as in cases of intracerebral hemangiomas, that the cavernous type usually is not visualized directly by angiography, but that the capillary type is demonstrated beautifully as a very fine vascular network.

The next problem to be illustrated is the unilateral exophthalmos secondary to congenital arteriovenous malformation of orbital and intracranial vessels.

Case 14. M.Z., a 43-year-old woman, was

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Fig. 29. Case 13. Infant with vascular neoplasm in left orbit.

Fig. 30. Case 13. Lateral arteriogram demonstrating vascular neoplasm of orbit with huge ophthalmic artery (I).
admitted because of left-sided headache and progressive prominence of the left eye over a period of 6–8 months. There was swelling of the upper and lower eyelid, with exophthalmos of 4.5 mm. on the left side (Fig. 31). Extraocular movements, and visual acuity and fields were normal. Both optic nerves were normal.

Left carotid angiography (Fig. 32) disclosed an arteriovenous malformation in the orbit with dilated feeding arteries (ophthalmic and internal maxillary) and enormously enlarged superior ophthalmic and angular veins. The left external carotid and internal maxillary arteries were ligated in the neck. The left-sided proptosis subsided gradually and nearly completely in the course of 6 months' time. The patient was in perfect health 3½ years later.

Case 15. S.A., a 16-year-old boy who was first noted at the age of 10 to have protrusion of the left eye, was admitted because of repeated bleeding from the nose over a period of 3 years. On examination there was left-sided exophthalmos of moderate degree (Fig. 33) with dilated pulsating retinal veins. Extraocular movements were normal; visual acuity and fields were also normal. Examination of the nose, pharynx and epipharynx revealed huge blood-vessel dilatations on the left side. Left carotid angiography disclosed an enormous extra- and intracranial arteriovenous malformation of the orbit, palatal region and left Sylvian region (Figs. 34 and 35). The left external and internal carotid arteries were ligated at the neck.

Fig. 31. Case 14. Patient with left-sided exophthalmos from retro-orbital vascular malformation before (left) and after (right) operation.

Fig. 32. Case 14. Lateral arteriogram (subtraction method of Ziedses des Plantes) demonstrating arteriovenous malformation.

Fig. 33. Case 15. Boy with left-sided exophthalmos from enormous extra- and intracranial arteriovenous malformation.

Fig. 34. Case 15. Lateral arteriogram showing intra-orbital and intracranial vascular malformation, and huge ophthalmic artery (1).
Comment. Unilateral exophthalmos occurred in this group spontaneously and was secondary to congenital, arteriovenous malformation of orbital vessels in Case 14, and of orbital, palatal and intracranial vessels in Case 15. The malformation is demonstrated beautifully by orbital angiography, especially by the subtraction method of Ziedses des Plantes. Pulsating exophthalmos as a manifestation of post-traumatic carotid-cavernous sinus fistula is not considered here; its diagnostic and angiographic features are well recognized. But a last group of vascular orbital lesions has to be mentioned, the so-called orbital varix of the ophthalmic veins.

Case 16. B.v.d.M., a 38-year-old woman with an injury of the right eye at the age of 5, was admitted because of a 2 months' history of progressive protrusion and outward displacement of the right eye. There was a bluish swelling of the inner portion of the right upper and lower lid and an exophthalmos of moderate degree. The swelling of the eyelids seemed to be under pressure (Fig. 36). Puncture of the angular vein disclosed dark fluid blood. The angular phlebogram showed a huge dilatation of the angular and superior ophthalmic vein (Fig. 37). After aspiration of 6 cc. of blood the swelling collapsed completely and remained so.

Case 17. R.St., a 35-year-old woman, was experiencing slight swelling of the left eye with impairment of vision on bending the head forward. There was slight left proptosis with slight increase of the exophthalmos on forward inclination of the head (Fig. 38). Roentgenograms of the skull and orbit, and right carotid angiogram were normal. Percutaneous angiography of the left angular vein demonstrated a cherry-like enlargement of the left superior ophthalmic vein (Fig. 39), and an orbital varix was diagnosed. The slight proptosis has been unchanged for 8 years.

Comment. In these 2 cases the orbital varix was disclosed by angular phlebography. In other cases of cystic retrobulbar tumor the tumor may be punctured directly and visualized by injection of contrast medium. This is true especially for the simple dermoid cyst
in the outer portion of the eyebrow and orbit.

DISCUSSION

The results of our angiographic experience with the orbit indicate that there is a definite place for orbital angiography in the diagnosis and treatment of unilateral exophthalmos, but it goes without saying that careful clinical examination still is required.

With the technique of percutaneous injection of the carotid artery and of the angular vein with highly concentrated contrast medium the circulation in the orbit and in a space-occupying lesion, namely, arterial, capillary and venous, are well demonstrable. Aside from the obvious vascular lesions such as hemangiomas, vascular malformations and orbital varix, which can be diagnosed definitely by this method, it is also of greatest possible value in the diagnosis of many mass lesions. The angiographic results are particularly encouraging in the differential diagnosis between benign and malignant orbital tumors. As a rule malignant tumors possess an excessive circulation permitting visualization even in an unfavorable location, such as in the retrobulbar space. Semimalignant and malignant tumors show an increased density of vascularization. Benign tumors, such as fibroma and neurinoma, are visualized by a very fine vascular network and peripheral, capsular vessels. Vascular tumors and malformations show the same typical patterns as the intracranial vascular lesions. The dis-placement of the ophthalmic artery and vein is demonstrated only in exceptional cases. As a result of the upward and downward displacement of the choroidal plexus the exophthalmos is visualized in a direct way.

SUMMARY

In the present review attention has been focused upon the angiographic features that seem to be of importance for the understanding of the mechanism of production of unilateral exophthalmos. The angiographic appearances of the ophthalmic artery and the choroidal plexus of the eye are demonstrated, and the visualization of the orbital veins by the direct method of angular phlebography is described. The value of orbital angiography is discussed with regard to the various vascular patterns of orbital space-occupying lesions.

REFERENCES


