The Value of Angiography in the Differential Diagnosis of Pulsating Exophthalmos

A Report of 3 Cases

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In his monograph on carotid-cavernous fistula, Dandy (1937) wrote “medical literature can scarcely claim more accurate and thorough studies than upon this subject.” Several years later the same author, referring to the comprehensive monograph of Sattler on carotid-cavernous fistula, stated: “The last word... has probably been said... on pulsating exophthalmos.”

In spite of all that has been “told and retold and most admirably” about carotid-cavernous fistula, there remain among the medical profession two outstanding false impressions. First, that the clinical diagnosis of carotid-cavernous fistula presents little difficulty. Secondly, that its treatment by carotid ligation in the neck is highly successful. The few patients in whom this treatment is not satisfactory are cured by a “trapping” operation.

That the treatment of carotid-cavernous fistula is, more often than not, a perplexing surgical problem has been made clear by Echols and Jackson.

Neither is the diagnosis of carotid-cavernous fistula, on clinical evidence, always so simple as might be expected in a lesion with such special features. Pulsating exophthalmos, the single important feature of carotid-cavernous fistula, may be produced by various other lesions.

The purpose of this paper is twofold: (1) to show that angiography is the only reliable method of diagnosing carotid-cavernous fistula, and (2) to record an unusual case of post-traumatic bilateral pulsating exophthalmos caused by head injury of a patient harboring basal meningioma, and closely simulating carotid-cavernous fistula. There is yet another aspect of angiography in carotid-cavernous fistula. This is the valuable information that could be obtained from careful study of these cases by angiography once their diagnosis is settled beyond any doubt. This will be the subject of a future communication.

Three representative case histories will now be described to show the difficulties that may be encountered in the clinical diagnosis of carotid-cavernous fistula.

Case Reports

Case 1. A 35-year-old woman while walking on rough ground stumbled and hit her head against the ground. She did not lose consciousness but immediately she became aware of a swishing noise in her head. Her right eye started to protrude at about the same time.

When seen 10 days later, there was right pulsating exophthalmos (Fig. 1). A murmur synchronous with the heart beat, was heard over the right eye and the right temple. Its intensity was diminished markedly but not abolished entirely by ipsilateral carotid compression. The diagnosis of carotid-cavernous fistula was made. Angiography revealed a vascular orbital tumor (Fig. 2).

Left carotid angiography was done. There was adequate collateral circulation. The right internal carotid artery was ligated in the neck.

Marked improvement in exophthalmos and pulsation was noted. The murmur became continuous and barely audible. When seen 4 years postoperatively there was practically no change in her condition.

Comment. There is little doubt that this patient has been harboring her orbital tumor for some time prior to the accident, but there was no history obtainable in this direction. The alleged appearance, for the first time, of her pulsating exophthalmos and murmur after head injury leaves little room for the diagnosis of a lesion other than carotid-

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Fig. 1. Case 1. Post-traumatic right pulsating exophthalmos caused by a vascular orbital lesion. Clinical diagnosis: carotid-cavernous fistula.

What actually happened to this vascular orbital lesion as a result of trauma is a matter for conjecture. The injury may have attracted the patient’s attention to a pre-existing lesion, or it may have established an arteriovenous communication in a congenital vascular anomaly. In view of little or no progress over 4 years it is more probably a vascular anomaly rather than a neoplasm. There was no means of verifying the diagnosis since the patient had had serviceable vision.

There is little doubt that, without angiography, improvement after carotid ligation would have been taken as corroborative evidence in favor of carotid-cavernous fistula.

As a matter of fact, careful reading of case histories in the older literature gives the impression that the diagnosis of carotid-cavernous fistula in some cases is doubtful. Very few patients came to autopsy\(^5,12\) and improvement after carotid ligation was considered sufficient evidence of correct diagnosis.

Case 2. A 40-year-old woman complained of left exophthalmos of 12 years’ duration. There was no history of trauma. Examination showed marked pulsating exophthalmos of the left eye, which was blind (Fig. 3). A bruit was heard over the left eye. It was diminished by ipsilateral carotid compression. The diagnosis of spontaneous carotid-cavernous fistula was made. Angiography disclosed a vascular orbital tumor (Figs. 4 and 5). The left eye was enucleated. Meningioma of the optic nerve sheath was diagnosed on pathological examination.

Comment. Absence of trauma and a long history are in favor of tumor. Pulsating exophthalmos with a bruit markedly diminished on carotid compression makes the diagnosis of carotid-cavernous fistula a high probability. Angiography proved an easy and reliable method in clearing this doubt. Incidentally, it was of material help in the treatment of this patient. Dual supply of blood from both internal and external carotid arteries was demonstrated (Figs. 4 and 5). Controlling the supply of the external carotid artery before commencing

Fig. 2. Case 1. Right carotid angiogram showing the vascular orbital lesion.
enucleation of the eye proved very helpful.

Case 3. A 23-year-old man complained of bilateral exophthalmos and progressive visual failure of 5 years’ duration. The disease started in the left eye and a few months later the right eye was involved similarly. When seen, there was bilateral nonpulsating exophthalmos, advanced bilateral primary optic atrophy and poor vision, all more marked on the left side. Angiography revealed a bilateral vascular subfrontal tumor. Operation was refused and the patient was discharged.

A year later, he was involved in a street-car accident. He did not lose consciousness. Shortly after the accident, he became aware of a swishing noise in his head. It was noted that his exophthalmos had increased markedly, more so on the left side. The next morning he had a sharp attack of profuse epistaxis. It continued for 24 hours and was, with difficulty, controlled by packing of the nose. Massive blood transfusion had to be administered.

When seen a few days later, there was bilateral pulsating exophthalmos, more marked on the left side. The left eye was virtually outside its bony socket. It was totally blind. Vision in the right eye was poor. A bruit was heard over both eyes and

![Fig. 3. Case 2. Left pulsating exophthalmos caused by a vascular orbital tumor. Clinical diagnosis: carotid-cavernous fistula.](image)

![Figs. 4 and 5. Case 2. (Left) Internal carotid angiogram showing blood supply of the tumor from the ophthalmic artery. (Right) External carotid angiogram (oblique view), showing blood supply of the tumor from the internal maxillary artery.](image)
both temples. Its intensity was about equal on both sides. It was diminished, but not abolished totally by digital compression of either carotid. Bilateral carotid angiography revealed the subfrontal tumor already diagnosed 1 year before (Figs. 6 and 7). At operation, a fairly large olfactory groove meningioma was removed subtotally. One week later his left eye was enucleated. Recovery was uneventful.

**Comment.** On his first admission this patient had had nonpulsating exophthalmos. Following the accident, his exophthalmos increased in degree and assumed its pulsating character. With this history of trauma, followed by pulsating exophthalmos and bruit, it was difficult to resist the temptation of diagnosing carotid-cavernous fistula, even though we were aware of the presence of his subfrontal tumor. Pulsating exophthalmos caused by injury of a pre-existing basal meningioma is, no doubt, a very uncommon finding. We are not aware of similar cases reported previously. Unfortunately, the angiograms before and after the accident are not comparable because of the difference in the angle of exposure.

The modus operandi of trauma is not altogether clear, but a reasonable explanation is as follows: Injury to the tumor has started bleeding. Blood has trickled to both orbital cavities through the superior orbital fissures and probably also through fractured orbital roofs. The fracture has lacerated the nasal mucosa and a communication was established between the nasal cavity and the bleeding tumor, hence his profuse epistaxis. In support of this view is the finding of a huge retro-orbital clot when the left eye was enucleated 1 week following the accident. A tear in the subfrontal dura mater would account for the pulsating character of exophthalmos which made its appearance only after the accident.

**Discussion**

The clinical picture of carotid-cavernous fistula has been described classically as consisting of 3 cardinal elements: (1) exophthalmos, (2) pulsation of the protruding eye and (3) bruit. Most authors lay special emphasis on bruit or the effect of digital compression on it, as the one distinguishing feature of “pulsating exophthalmos” caused by carotid-cavernous fistula from that resulting from other causes. **Jefferson** had this to say on the relative importance of the three clinical signs: “‘pulsating exophthalmos’ indeed is a term so picturesque that it gives a wrong impression of a condition in which

![Fig. 6. Case 3. Left carotid angiogram a few days following the accident. There is backward displacement of the carotid and anterior cerebral arteries. Abundant blood supply to the tumor is demonstrated.](image1)

![Fig. 7. Case 3. Tracing of frontal view of left carotid angiogram a few days after the accident. Suprasellar extension of the tumor is shown.](image2)
more often than not the exophthalmos is minimal and the pulsation is scarcely perceptible. The one constant feature is the bruit."

Review of our cases will show that bruit was present in all 3 cases. It was diminished, in varying degrees, by ipsilateral carotid compression.

Analysis of large series of carotid-cavernous fistulas, as well as the continuous reporting of individual cases, is convincing that one or more of this favorite triad may be missing. Even the entire gamut of these classical signs may be absent in some cases. It serves no useful purpose to tread once more the well-worn path of the differential diagnosis of carotid-cavernous fistula. There are, however, some points that will need comment.

It seems that carotid-cavernous fistula, a well-known and thoroughly studied lesion for many years, is claiming little attention from the modern neurosurgeon. Most of the basic work on the subject has been done in the pre-angiography days. The exhaustive monograph of Sattler (1920), the review by Locke of 588 cases, the monograph by Dandy and his notable advance in treatment by intracranial "clipping" and the more recent review by Martin and Mabon are important landmarks in our knowledge of this lesion and have no parallel in modern literature. This lack of interest may be ascribed to a mistaken belief that this long-known disease has been described completely and there is little left for further study.

Curiously enough the introduction of angiography did very little to advance our knowledge of this lesion. The noteworthy contribution was made by Wolff and Schmid who studied the venous drainage of the cavernous sinus. Their work was confirmed later by List and Hodges.

Apparently, Terry and Mysel's case of carotid-jugular fistula was the first one in which angiography was used.

It was the opinion of Dandy that the clinical picture was "sharply defined and unmistakable." He described Terry and Mysel's angiographic demonstration as "far from convincing". He concluded "since the clinical picture of these aneurysms is unmistakable, one cannot be justified in unnecessary procedures merely to display them more graphically."

It is clear from the review of our 3 cases that other diseases may present the full picture of "classical" carotid-cavernous fistula. Clinical diagnosis, under these conditions, is unreliable, and there is no easy means of verifying the true situation. Angiography is the only method of making a confident diagnosis. Not only is it necessary to establish irrefutably the presence of a fistula, but also to indicate its exact site. Exophthalmos may be on the opposite side of the fistula, it may be bilateral in a unilateral case or even two fistulae may be present at the same time.

The angiogram of carotid-cavernous fistula is easy to interpret. There is only one precaution to take. A large intracavernous nonfistulous carotid aneurysm may simulate superficially carotid-cavernous fistula, but this is distinguished readily by the absence of dilated ophthalmic veins.

The merits of angiography are so obvious that no surgeon could afford to handle a case of "pulsating exophthalmos" without its valuable help.

**Summary**

The diagnosis of carotid-cavernous fistula on clinical evidence alone is sometimes difficult. Angiography affords a simple and safe method of making a definite diagnosis. Three case histories are cited to illustrate the diagnostic difficulties. One of them is a rare example of head injury to a patient harboring basal meningeoma.

**References**

4. **Dandy, W. E.** Cited by Hamby.
5. **Dandy, W. E.** The treatment of carotid cavernous


