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 re-told 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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cavernous fistula. However, angiography revealed with certainty the true nature of the disease.

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

Fig. 2. Case 1. Right carotid angiogram showing the vascular orbital lesion.