Mechanisms of presentation of carotico-cavernous fistulae

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The clinical features of carotico-cavernous fistulae are too well known for detailed description to be necessary. The proptosis, chemosis, venous engorgement in and around the orbit, oculomotor palsy, pulsation of the globe, and the subjective and objective bruit over the orbit and forehead are familiar to all. It is perhaps not always appreciated that the bruit may, on occasions, precede the appearance of the other features by weeks or even months. There are, however, several modes of clinical presentation, and a number of different aetiological processes responsible. The lesion may present as the conventional unilateral carotico-cavernous fistula; the bilateral fistula; the contralateral fistula, in which the pulsating exophthalmos is on the side opposite to the actual fistula; or the alternating fistula. This has been written in these words advisedly, because there is a tendency to accept such expressions too readily and too literally without giving enough thought to their meaning. For instance, does bilateral pulsating exophthalmos mean that the patient has two symmetrical carotico-cavernous fistulae? Or does the alternation of ocular symptoms mean that one fistula has healed but by coincidence the same patient has developed another on the opposite side? It is because this must very rarely be the case that this paper has been written, to consider the mechanism by which clinical presentations of different types comes about, and also to describe two unfamiliar aetiological processes.

Anatomical considerations (Fig. 1a, opposite)

The cavernous sinuses lie lateral to the pituitary fossa between two layers of dura. They connect anteriorly with the superior orbital veins, and through these with the supra-orbital, angular, and inferior orbital veins. Posteriorly they are connected to the superior petrosal sinuses, and the inferior petrosal sinuses, and through the latter to the jugular veins. They are not isolated structures, however, but are joined together by the intercavernous sinuses, anterior and posterior, before and behind the pituitary fossa, and also through the basal venous plexus lying on the clivus, and other less important communications. The degree of patency and the relative importance of these connections varies. The sinuses themselves are not just flaccid containers of blood, but are named from the fibrous trabeculations which traverse them giving the appearance of cavernous tissue. They are the only places in the body where a major artery lies bathed in venous blood.

When considering the mechanism of carotico-cavernous fistulae, the important structures involved are the superior orbital veins and the intercavernous sinuses, particularly the anterior. Their anatomical relationships vary, as does the relationship of the carotid artery to them.

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FIG. 1 Diagrammatic representation of cavernous sinus and connecting veins

- (a) normal
- (b) ipsilateral carotico-cavernous fistula
- (c) arterial blood crossing by the intercavernous sinus gives impression of bilateral fistulae
- (d) ipsilateral ophthalmic vein thrombosed but intercavernous sinus patent allowing contralateral syndrome to develop.

Clinical material

The material for this study comes from a series of eleven consecutive cases of carotico-cavernous fistulae, ten of which were female, one of which was "bilateral", one contralateral, and three "alternating". All were non-traumatic in origin.

Aetiological factors

According to Walsh (1957), trauma is responsible for 75 per cent. of all cases and men are most frequently affected. In the present series, however, the chief cause of the condition is the rupture of a hypertensive atherosclerotic internal carotid artery, or of a pre-existing intracavernous aneurysm, or a combination of aneurysm, hypertension, and atheroma. Women predominate, and most of them are elderly (*e.g.* Case 1). Spontaneous

fistulae may on rare occasions occur in younger patients, however, in which neither trauma nor aneurysm play any part, and two of these unusual mechanisms are mentioned later in this paper.

Mechanism of production of clinical symptoms

Fig. 1 (*b-d*) is intended to illustrate in diagrammatic fashion the principal events which occur. When rupture occurs, arterial blood escapes from the carotid artery, enters the sinus, tracks through the trabeculae (which may delay the development of the full clinical picture), and then enters the superior ophthalmic vein (Fig. 1*b*), which becomes engorged, enlarged, and tortuous. These changes are passed forwards to the supraorbital, angular, and inferior ophthalmic veins, and arteriography will, in the lateral view, show the dye escaping into the sinus and along the distended superior ophthalmic veins to the orbit (Fig. 2). This is the mechanism of the conventional unilateral fistula with ipsilateral pulsating exophthalmos. It depends upon the superior ophthalmic vein being patent, and the intercavernous sinus being relatively unimportant.

FIG. 2 *Internal carotid arteriogram (subtraction technique) showing dye entering cavernous sinus and passing along dilated superior ophthalmic vein to the orbit*

THE "BILATERAL" FISTULA (Fig. 1*c*)

If both superior ophthalmic veins are patent and in the normal position, and if the intercavernous sinus is patent, or becomes so as a result of the pressure of arterial blood in the cavernous sinus, this blood can not only pass into the superior ophthalmic vein on the side of the fistula, but can also pass across the mid-line and enter the other sinus and the other superior ophthalmic vein, thus causing bilateral pulsating exophthalmos and bruit. There is however still only one fistula, which is a point of great importance, for if this can be demonstrated arteriographically only one carotid artery need be sacrificed for both eyes to be cured.

THE CONTRALATERAL FISTULA (Fig. 1*d*)

If the superior ophthalmic vein on the side of the fistula is an unimportant structure in that particular patient, or has an unusual mode of communication, or becomes thrombosed

by damage to its walls from the rush into it of arterial blood; and if the intercavernous sinus is patent or becomes so, the arterial blood can track across to the other side, entering the other superior ophthalmic vein, and so cause unilateral pulsating exophthalmos on the side opposite to the fistula. That this is not merely hypothetical was shown by Dandy and Follis (1941); there have been others in the literature, and the following patient from this series illustrated the point very clearly.

Case 1

A woman aged 58 suddenly developed a sensation of something having snapped inside her head. For 48 hours she had severe headache and developed a pulsating noise inside the head. Over the next 6 months a right-sided unilateral pulsating exophthalmos gradually became apparent with a loudly audible systolic bruit. However, when the right carotid arteriogram was carried out by Dr. Philip Moxon, no abnormality was seen. The procedure was repeated on the left side, and here it was clearly demonstrated that there was a left-sided carotico-cavernous fistula, and the dye could be seen to be tracking through the anterior intercavernous sinus to the right side, and along the right superior ophthalmic vein to the orbit. The ipsilateral ophthalmic vein did not appear to be patent. Immediately after arteriography all the symptoms ceased and all the physical signs subsided. A very careful repetition of the left carotid arteriogram was then carried out and this showed that, whereas the dye still escaped from the artery into the left cavernous sinus, the intercavernous connection had become occluded at its mid-point. This patient has remained perfectly well for over 5 years.

THE ‘‘ALTERNATING’’ FISTULA

If, after a conventional carotico-cavernous fistula syndrome has developed, the superior ophthalmic vein thromboses, the symptoms will settle down. It has been seen, however, in Case 1 that the fistula may remain open, and this may cause the intercavernous sinus to open up gradually, until finally arterial blood is allowed to enter the contralateral superior ophthalmic vein (Fig. 1*d*), so that the pulsating exophthalmos will re-appear, but on the opposite side. It is still only the one fistula which is present and which may require treatment.

Much of what has been said may be hypothesis, but it is clearly proven in Case 1, and it seems so much more likely that a sequence of events of this type may take place than that identical fistulae should develop coincidentally in the two carotid arteries at the same or different times.

Some additional aspects of aetiology

The commonest causes of non-traumatic carotico-cavernous fistulae have been mentioned earlier, but in the younger, non-atheromatous, normotensive patient, there must be some unusual defect in the wall of the carotid artery. The type of intracavernous aneurysm which affects the older patients is uncommon in the young, and haemorrhagic disorders rarely cause such symptoms. There may occasionally, however, exist a hereditary fragility of the vessel wall, or the vessel may come to be eroded from outside.

Case 2

A 21-year-old female patient had suffered a thrombosis of the right internal carotid artery after an acute infection of the middle ear and petrous bone, and 2 years later she suddenly developed a left-sided intracranial bruit which after 2 months was followed by pulsating exophthalmos on that side. Arteriography proved the presence of a carotico-cavernous fistula, but there was no aneurysm. She gave a history, however, of having had many spontaneous haemorrhages in previous years in the

subcutaneous tissues, the muscles, and around both orbits, and any interference with her arterial walls (such as during carotid arteriography) had produced very troublesome haemorrhage. Her mother had shown exactly the same type of vascular fragility. Their blood coagulation factors were all quite normal, and sections of her arterial wall examined by light and electron microscopy showed no recognizable abnormality. The nature of this fragility was quite obscure but it seemed possible that the excessive demands made upon the one vessel to supply both hemispheres was such that in the presence of this fragility spontaneous rupture occurred.

Case 3

A mildly hypertensive male patient aged 50 suddenly developed the typical signs and symptoms of a left-sided carotico-cavernous fistula. On examination, however, he presented in addition the full clinical picture of marked hypopituitarism, which had previously passed unnoticed, and without visual field defect. Plain x rays of the skull showed a ballooned sella turcica, and by contrast radiography the presence both of a pituitary tumour and of a carotico-cavernous fistula was confirmed. The tumour was removed and the fistula occluded (by Mr. Jack Small) at two operations, and it was seen that there had been a lateral projection of the pituitary tumour which had infarcted and presumably eroded the carotid artery wall.

Two other cases in which lateral projections of pituitary tumours have almost encircled the carotid artery have recently been seen and which, if a more advanced stage had been reached, might have been expected to erode the artery wall and result in a fistula.

Conclusion

It has been the aim of this paper to stress the need for viewing the cavernous sinus and its communicating veins not as rigid structures, but as part of the dynamics of the cerebral circulation. When a fistula occurs, some vessels open up, others close down, and new channels may appear, but throughout all this the patients themselves can respond by showing only a limited clinical picture. It is therefore most important in such cases, if their age and physical condition justifies it, that the most meticulous arteriographic studies should be carried out bilaterally, in order that the precise potential for surgical correction of the lesion may be assessed. Another interesting aspect shown in this series, which is well-known, and is illustrated clearly in Case 1, but for which no very adequate explanation has been arrived at, is that either the process of arteriography, or in other cases the trial period of carotid compression usually required before the carotid is interrupted surgically, may result in closure of either the fistula or its associated channels, so that the symptoms settle down without further interference being required.

Summary

Mechanisms are described by which the so-called bilateral, contralateral, and alternating carotico-cavernous fistulae may be produced by a single arteriovenous communication. Attention is also drawn to two unusual pathological processes which may be the cause of such fistulae.

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