CONGENITAL INTRAORBITAL ARTERIOVENOUS ANEURYSM

BY

RAMÓN JACAS, ADOLFO LEY, and DOMINGO CAMPILLO

From the Neurosurgical Department of the Seguro Obligatorio de Enfermedad, Barcelona, Spain

Aneurysms or malformations of the intraorbital vessels are lesions which in spite of the large number of cases cited in the medical literature have rarely been verified, the larger part being merely clinical descriptions.

In 1823, Guthrie described the first case of intraorbital aneurysm proved by post-mortem examination. Pulsating exophthalmos was the most striking feature of this case, and for many years after this description pulsating exophthalmos was thought to be pathognomonic of intraorbital aneurysm (Dempsey, 1886; Nunneley, 1859). In 1865 Nunneley extended the pathological possibilities by describing two cases of pulsating exophthalmos due to an aneurysm of the internal carotid in one case and to an intracranial extension of a thyroid carcinoma in the other.

In 1870, Delens reviewed the whole problem of pulsating exophthalmos and identified a carotid cavernous fistula as the most common cause, and from that time intraorbital aneurysms seem to have been almost forgotten. This can be seen in the publications of Locke (1924) and Martin and Mabon (1943) followed in 1920 by Sattler who referred to five supposed arteriovenous aneurysms, not verified, as a cause of pulsating exophthalmos.

In 1949, Heimburger, Oberhill, McGarry, and Bucy reviewed the published cases of intraorbital aneurysm and found that only in six was the lesion verified. After studying the descriptions given, they came to the conclusion that in many cases the ocular symptoms were probably due to carotico-cavernous fistulas or vascular lesions of endocranial origin. They added one case of their own of non-pulsating exophthalmos of the left eye of rapid development. It was accompanied by congestion, pain, conjunctival oedema, amblyopia, increase of ocular pressure, and dilatation of the veins of the retina, followed later by haemorrhages. This case was explored and a great dilatation of the lacrimal artery was found, without any evidence of other vascular abnormalities. Ligation of the enlarged artery was followed by recession of the clinical symptoms.

The following case presents a clinical picture of an intraorbital arteriovenous aneurysm, verified by angiography.

Case Report

E.C.F. (H.Cl., S.O.E.41.402), a woman, 31 years of age, was first seen on September 18, 1957. She stated that in the last two years, without any pathological antecedent or trauma, she noticed on bending the head a feeling of tension around the left eye. The various ophthalmologists who had seen her could not find any changes in the visual acuity, fields, or fundi.

During the last three months these troubles had increased, appearing with any forward inclination of the head. Two months previously she had suffered an acute episode accompanied by diplopia and transient amblyopia lasting half an hour. The diplopia coincided with periods of intense palpebral oedema. These crises passed on lifting the head. The patient did not complain of headache or subjective cranial bruit at any time.

Examination showed a normal young woman, bright and alert. The superficial venous network of the eyelids was perhaps more obvious than usual, especially on the left side. A mild exophthalmos was present on the same side, the protrusion being 6 mm. more than on the right side. With the head erect, the left palpebral fissure was slightly narrower than the right (Fig. 1A), and there was also mild conjunctival congestion. The pupils were equal and reacted normally. Pressure on the left jugular vein, or bending of the head, produced palpebral swelling and a slight increase of the left exophthalmos (Fig. 1B).

The visual acuity was 90/100 in the right eye, and 50/100 in the left eye. With proper correction for his myopic astigmatism, the visual acuity in both eyes rose to 100/100. Ocular motility was normal, but there was a mild intermittent imbalance of the muscular equilibrium, due to deviation of the left eye. The visual fields showed in the right eye a mild upper temporal depression. In the left eye there was a concentric reduction for white and colours and a depression in the upper nasal quadrant.

The fundi were normal except for a mild venous congestion on the left. Otherwise, the neurological and systemic examinations were normal. The blood pressure was 130/85 mm. Hg. No bruit was audible over the head or eyeballs.

No abnormalities were found in the x-ray examination of the head, including the orbits and the optic canals. Bilateral carotid angiography was carried out. No abnormalities were observed on the right side. On the
CONGENITAL INTRAORBITAL ARTERIOVENOUS ANEURYSM

Fig. 1.—Palpebral fissures at rest (A) and during jugular compression (B).

Fig. 2.—Carotid angiogram, lateral projection. Note arteriovenous aneurysm in posterior part of orbit.

Fig. 3.—Carotid angiogram, early (A) and late (B) stage in axial projection. Note arteriovenous aneurysm in upper part of orbit.
left a lesion was demonstrated in the postero-superior part of the orbit. Its appearance was typical of an arteriovenous aneurysm, of which the arterial supply probably came from the lacrimal artery, a branch of the intraorbital portion of the ophthalmic artery (Figs. 2 and 3). The venous drainage of the lesion was by way of the superior ophthalmic vein. No abnormality was seen in the intracranial vessels.

Discussion

This case demonstrates an intermittent exophthalmos. The first description of this type of exophthalmos was published by Schmidt in 1805, and although they are rare, in 1944 Walsh and Dandy collected 111 published reports of which the majority were merely clinical observations. The commonly accepted aetiology of intermittent exophthalmos (Cordero, 1935; Hippius, 1936; Olivella, 1946) is that of an orbital varicocele, caused according to Birch-Hirschfeld (1930), by a complete or partial blockage of the venous drainage of the orbit into the jugular vein. In 1944 Walsh and Dandy published one of their own cases of intermittent exophthalmos due to an extensive arteriovenous aneurysm situated within and behind the sphenoidal fissure and extending through the dura mater to the Gasserian ganglion and the cavernous sinus. The lesion was demonstrated at operation, and was the first case of verified aetiology. Later, Walsh (1947) published another case in which a great varicosity was found situated in the sphenoidal ridge. Senà in 1955 cites a series of published cases of known aetiology, namely, diffuse angioma, congenital vascular tumour, lymphosarcoma, ethmoiditis, pituitary tumour, cavernous angioma, etc. None of these cases was angiographically verified, but Huber (1951) published a case similar to our own in which the lesion was so verified.

Concerning the treatment, we must confess that we have no previous experience with intraorbital arteriovenous aneurysms. From our experience with similar intracranial lesions, we had the impression that the lesion in the present case could be removed surgically. However, we could not guarantee that in doing so there would not be the possibility of damaging the function of the optic or oculomotor nerves, either by direct injury or as a result of the surgical interference with the blood supply of the eye or the orbit. We discussed the matter with the patient, and as she had not had any pain yet and her visual function was so well preserved, she decided not to accept the risks of an operation at present unless new circumstances arose.

The authors wish to express their sincere thanks to Dr. F. Palomar for his ophthalmological cooperation.

BIBLIOGRAPHY


Schmidt (1805). Cited by Senà, J. A.


Williams and Wilkins, Baltimore.