Treatment of ophthalmofacial-hypothalamic arteriovenous malformation (Bonnet-Dechaume-Blanc syndrome)

Case report


Department of Neurosurgery, Royal Prince Alfred Hospital, and Departments of Neurosurgery and Radiology, Royal Alexandra Hospital for Children, Camperdown, New South Wales, Australia

A 17-year-old girl with unilateral blindness and exophthalmos was found to have Bonnet-Dechaume-Blanc syndrome without retinal arteriovenous communications. The arteriovenous malformation was managed by combined intracranial resection, ophthalmic artery ligation, and selective embolization of the external carotid component.

KEY WORDS • ophthalmofacial-hypothalamic arteriovenous malformation • Bonnet-Dechaume-Blanc syndrome • embolization • surgical treatment

In 1937, Bonnet, et al., described the syndrome of ophthalmofacial-hypothalamic arteriovenous malformation. Prior to 1973, only 43 cases had been reported in the literature. Since then, a further seven cases have been added. Angiographic confirmation of the diagnosis is available in only a minority of cases, so that Mello and Detoni could list only 17 such cases prior to 1975. The case reported below is the first to be treated by combined intracranial resection, ophthalmic artery ligation, and selective external carotid artery embolization.

Case Report

This 17-year-old aboriginal girl presented with a 10-week history of left retro-orbital headache, left eye blindness, and left-sided proptosis with vascular engorgement.

Examination. She had a pulsatile left proptosis and bruits over the left frontal, temporal, and maxillary regions. Ophthalmological assessment confirmed a blind left eye with an afferent pupillary defect and left optic atrophy associated with distended pulsatile draining veins and dilated left lateral conjuctival vessels.

Angiography via the left internal carotid, left external carotid, and right internal carotid arteries revealed an arteriovenous malformation (AVM) with a 2-cm intracranial component extending from the left optic foramen to the hypothalamus (Fig. 1 left and center), and a left orbital and facial AVM (Fig. 1 right) with a hypertrophied ophthalmic artery. It was difficult to identify the individual feeding vessels, but both external and internal carotid arteries contributed to the malformation.

First Operation. On April 9, 1984, the patient underwent surgery with a left subfrontal approach to the intracranial component of the AVM. The malformation was resected under magnification. It was noted that the AVM followed the course of the optic nerve from the optic foramen to the chiasm. Only at the chiasm was neural tissue identified in the pathway, as there was no evidence of a left optic nerve. From the chiasm, the AVM extended through the lamina terminalis and into the hypothalamus. Most of the intracranial AVM, with its supply arising from the internal carotid and anterior cerebral arteries, was resected, but a small residual clump of malformation vessels was left in the wall of the third ventricle. The AVM along the intraorbital course of the optic nerve and the ophthalmic artery were also left undisturbed as it was thought desirable to repeat the angiography to ascertain what contribution
Bonnet-Dechaume-Blanc syndrome

Fig. 1. Preoperative left internal carotid angiograms (left and center) and left external carotid angiogram (right) showing the extent of the arteriovenous malformation.

Fig. 2. Left internal carotid angiograms, lateral (left) and anteroposterior (right) views, following excision of the intracranial component of the arteriovenous malformation.

the ophthalmic artery made to the lesion. Postoperatively, the patient's recovery was unremarkable apart from transient diabetes insipidus. Left internal and external carotid and right internal carotid angiography was performed on April 18, 1984. This confirmed excision of the suprasellar component with a very small component remaining in the wall of the hypothalamus (Fig. 2). Better definition of the external carotid artery supply revealed an AVM in the infratemporal fossa and oropharynx, fed by small vessels through the inferior orbital fissure that arose from the internal maxillary and proximal superficial temporal arteries. The malformation was also seen to extend superiorly into the temporal fossa.

Second Operation. On May 28, 1984, the left frontal osteoplastic craniotomy was reopened, and the left ophthalmic artery was clipped with a mini Yasargil aneurysm clip. This enabled the orbital component and the external carotid supply to be dealt with by embolization safely and without fear of reflux through the ophthalmic artery into the internal carotid artery circulation.

Embolization. Ivalon and Gianturco spring-coil embolization of the external carotid artery component was performed by selective catheterization of the superficial temporal and internal maxillary arteries via transfemoral catheterization on June 6, 1984. Embolization resulted in 24 hours of facial pain but no other complications. A subsequent angiogram demonstrated obliteration of the extracranial AVM (Fig. 3).

Discussion

The Bonnet-Dechaume-Blanc syndrome was first described in 1937 as an association of retinal cirrhotic aneurysms with facial and cerebral arteriovenous communications. Wyburn-Mason added to the descrip-
tion of the syndrome by demonstrating the anatomical correlations in two cases. Since then, patients described as having this syndrome have presented with a variety of clinical symptoms and signs. The cutaneous manifestations include angiomas of the face, cheek, nose, mandible, palate, pharynx, maxilla, and buccal mucosa. The ocular manifestations include arteriovenous communications seen on funduscopy, visual impairment, and exophthalmos (which may be pulsatile). Neurological manifestations are related to the intracranial component of the lesion and present in the same way as any similarly located AVM (that is, with hemorrhage, epilepsy, and neurological deficits). The common underlying vascular abnormality is an extracranial AVM contiguous with a malformation of the optic pathways. Cutaneous lesions and retinal AVM's are no longer thought essential to the diagnosis of the syndrome.1,2,14

The natural history of this lesion has not been adequately documented. There have been reports of massive oral, nasal, and pharyngeal hemorrhage from the external carotid artery component,3,5,10 while visual impairment is known to occur in association with the internal carotid artery component.2,3,7,12,13,15 The incidence and consequences of hemorrhage from the intracranial component are presumed but not known to be similar to those of other intracranial AVM's.13,15

The discussion of management is scanty in the literature and what can be found is hampered by lack of knowledge of the natural history referred to above. Some authors have advocated radiotherapy.3,13 Malis9 has suggested excision of the intracranial component and possible staged surgery for the extracranial component, although he does not provide details of treatment used in specific cases.

As the patient in the present report was already blind in the left eye, it seemed reasonable to attempt extensive resection of the intracranial component involving the left optic nerve. The combination of surgical excision with selective external carotid artery embolization8 for the extracranial component produced substantial obliteration of the lesion. It remains to be seen whether the small residuum in the hypothalamus is of any significance or whether the vascular channels in the extracranial component will reopen.

References


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Address reprint requests to: Ian H. Johnston, F.R.C.S., Department of Neurosurgery, Royal Alexandra Hospital for Children, P.O. Box 34, Camperdown, New South Wales 2050, Australia.