Dural arteriovenous fistula and spontaneous choroidal detachment: new cause of an old disease

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SUMMARY A case is presented in which bilateral spontaneous choroidal detachments appear to be the direct result of bilateral dural arteriovenous fistula of the cavernous sinus region. Rapid resolution of the clinical signs followed bilateral orbital decompression via the transfrontal approach. Similarities in clinical presentation of both entities are reviewed and a premise for their cause-and-effect relationship elaborated. A literature search for similar unrecognised cases is discussed. The paper suggests that this association may be more frequent than published reports would imply.

Spontaneous choroidal detachment and dural arteriovenous fistulae of the cavernous sinus are uncommon clinical entities. Each produces a relatively characteristic syndrome with a constellation of ocular signs and symptoms. Their occurrence together has been previously reported on only a single occasion (Woillez *et al.*, 1967).

A patient with this rare clinical combination recently provided the opportunity to study extensively their association. The results of this investigation suggest the association may not be as unusual as the current literature would suggest. A plausible basis for the relationship of spontaneous choroidal detachment with dural arteriovenous fistulae in the region of the cavernous sinus is now proposed.

Case history

A 70-year-old White male professor emeritus of internal medicine noted irritation and a foreign body sensation of both eyes associated with lacrimation, conjunctival injection, and mild lid oedema in October 1973. Before the onset of symptoms he admitted to much stress in his work as well as a minor fall in which he struck the right side of his head without apparent injury. Initial therapy with topical steroids by his private ophthalmologist provided no improvement.

A thorough medical investigation with neurological consultation was undertaken in June 1974. No

history of additional neurological, ophthalmological, endocrine, or medical symptoms was elicited. He specifically denied headache, ocular pain, diplopia, visual loss, or cranial bruits. A review of his past medical history revealed hypertensive cardiovascular disease, coronary artery disease with angina pectoris, diverticulosis and diverticulitis, Dupuytren's contractures, and transient ischaemic cerebral vascular disease characterised by transient global amnesia and left hand inco-ordination of infrequent occurrence.

Physical examination showed the visual acuity to be 20/20 OU. Both pupils were noted to be dilated, the right responding poorly to direct light. Ocular motility was normal and there was no ptosis, although mild lid oedema persisted. Hertel exophthalmometry registered 20 mm OU. Marked dilatation of conjunctival vessels was now present. No cranial or ocular bruits were detected and there was no increase in resistance to retropulsion of the globe. Slit-lamp examination was normal, and applanation tensions were 21 mmHg OU. Retinal changes were characterised funduscopically by mild venous engorgement, small dot haemorrhages, and arteriovenous nicking. No significant findings were recorded on general physical or neurological examination.

Laboratory studies giving normal results at that time included: total and differential blood counts, ESR, urine analysis, BUN, glucose, serum electrolytes, calcium, phosphorus, alkaline phosphatase, LDH, SGOT, cholesterol, bilirubin, total protein and A/G ratio, creatinine, uric acid, serum protein electrophoresis and immunoglobulins, cryoglobulins, tuberculin and histoplasmin skin tests, and

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Fig. 1 Top view shows conjunctival vascularity at the time of admission. Bottom view shows resolution of conjunctival vascularity

toxoplasmin serum titres. Chest and skull x-rays were normal. The electrocardiogram revealed a right bundle branch block, left anterior hemiblock, occasional ventricular ectopic beats, and left ventricular hypertrophy.

No diagnosis was then established, though a hyperviscosity syndrome was considered.

At 2 subsequent ophthalmic examinations no new diagnostic suggestions were provided. On 2 August 1974 visual acuity OD had dropped to 20/40, and an intraocular mass was noted temporally from 7 to 10 o'clock. A choroidal melanoma was suspected. A second opinion was requested of the authors before proposed enucleation.

Examination on 7 August confirmed the presence of a pigmented mass extending from 7 to 10 o'clock temporally OD. The mass transilluminated. Fluorescein angiography showed only the previously noted vascular changes. A nasal visual field loss was noted. The remainder of the ocular examination was unchanged. In view of this examination the mass was thought to be a choroidal detachment, and the patient was begun on oral prednisone 60 mg daily. By 19 August 1974 the choroidal detachment was more extensive OD, involving both the temporal and nasal segments. A temporal and inferior choroidal detachment was now present OS.

Three days later abrupt proptosis OD, with limitation of motility and ptosis, occurred over a period of 15 minutes. The patient was placed at bed rest, and by 25 August the proptosis, motility defect, and ptosis had resolved.

On 3 September 1974 the patient was admitted to the Neuro-Ophthalmology Service at the Medical

College of Virginia. On admission conjunctival vascular changes (Fig. 1) with mild chemosis and lid oedema were re-affirmed. Visual acuity OD had fallen to light perception. Acuity OS was 20/50, could not be improved, and was associated with a moderate red/green colour perception loss. The pupils were both widely dilated; OD remained unresponsive to light, OS normally responsive. Hertel readings were 23 mm OD and 24 mm OS.

Ocular motility was restricted in all fields without ptosis. Fundusoscopic examination demonstrated previously documented bilateral choroidal detachments. Visual field examination revealed both temporal and nasal defects OS; OD could not be effectively plotted (Fig. 2). General physical and neurological examination was normal. No bruit was heard about the head or neck. The differential diagnosis included endocrine exophthalmos and arteriovenous fistula. A low-flow dural fistula was thought to be more likely than a conventional high-flow carotid-cavernous fistula.

In the early morning of 7 September the patient experienced the onset of severe orbital pain. Examination showed increase in proptosis, with Hertel readings now of 28 mm OD and 26 mm OS. There was increased chemosis and lid oedema. Ocular motility was restricted in all fields (Fig. 3). The choroidal detachments were nearly complete, and visual acuity had fallen to bare light perception OU. Intraocular pressure was recorded at 46 mmHg OU with fairly shallow anterior chambers.

The patient was immediately begun on acetazolamide and subjected to selective cerebral angiography. This showed bilateral 'low flow' dural arteriovenous

Fig. 2 Tangent screen visual field at time of admission

Fig. 3 Marked proptosis, chemosis, conjunctival injection, and restriction of ocular motility

fistula draining into the cavernous sinus (Fig. 4). Because of the exacerbation of the orbital signs and symptoms attributed to venous stasis and hypoxia, it was decided to decompress both orbits in an attempt to reverse the course of events and regain vision. Bilateral transfrontal orbital roof decompression was therefore undertaken shortly after angiography.

The immediate postoperative course was uneventful. The patient experienced progressive improvement, and by December 1974 both eyes were quiet and white (Fig. 1). Visual acuity was 20/30 OS and 20/40 OD, and ocular motility full (Fig. 5). The choroidal detachments were largely resolved. At this point it was considered that both fistulae had undergone thrombosis. He has continued an asymptomatic course since then.

Discussion

The initial clinical description of spontaneous choroidal detachment has been attributed to von Graefe in 1854 (McDonald *et al.*, 1964). Ursin (1965) found 100 reported cases. Small series (McDonald *et al.*, 1964; Preisler, 1965; Rosen and Lyne, 1968; Davis *et al.*, 1973; Scheie and Morse, 1974), as well as single cases (Horay, 1935; Karaske, 1935; Csillag, 1937; Mathews and Moodie, 1955; Lewallen, 1957; McClure, 1967; Woillez *et al.*, 1967; Brockhurst and Lamb, 1973) of spontaneous exudative or serous choroidal detachment continue to be reported, emphasising the infrequency of this clinical entity.

By definition spontaneous choroidal detachment excludes separation of the choroid related to intra-

Fig. 4a Lateral view of the left common carotid artery injection. There is early filling of the posterior cavernous sinus (curved arrow)

Fig. 4b Frontal view of left common carotid artery injection with manual cross-compression of right carotid artery. Early filling of cavernous sinus is seen medially (arrow)

Fig. 5 Complete resolution postoperatively of proptosis, chemosis, conjunctival injection, and oculomotor restriction

ocular surgery, trauma, obvious focal infection or haemorrhage, and traction effects in a phthisical eye. It has been associated with many local ocular as well as systemic diseases (Schepens and Brockhurst, 1963; Gupta *et al.*, 1965). Common among these has been ocular inflammation: iridocyclitis, tenonitis, and scleritis.

Spontaneous choroidal detachment most commonly presents over the age of 55, with a predilection

for males. Presenting symptoms consist of visual impairment often associated with pain, photophobia, and epiphora. Visual loss of significant degree is commonly associated with shallowness of the anterior chamber, hypotony, retinal venous stasis, and contraction of the visual field. Additional signs included: increased intraocular pressure, proptosis, conjunctival injection, and non-rhegmatogenous retinal detachment. The course is

often characterised by remission and recurrence of symptoms.

A number of theories have been considered with regard to the aetiology of spontaneous exudative or serous choroidal detachment. Although its cause remains elusive, it is generally accepted that a vascular process producing choroidal congestion is the source of transudation, which results in the choroidal separation (Verhoeff and Waite, 1925; Capper and Leopold, 1956; Lewallen, 1957; Cogan, 1960; Davis *et al.*, 1973; Scheie and Morse, 1974). This concept is experimentally supported by Aaberg's work in the owl monkey (Aaberg, 1974). Most authors agree that many diseases can produce this 'vasculopathy'. This has led Velzeboer (1960) to suggest that choroidal detachment should be considered a symptom rather than a separate clinical entity. Perhaps spontaneous choroidal detachment should be considered a secondary clinical entity symptomatic of a primary disease productive of significant alterations of intraocular haemodynamics which include changes in choroidal vascularity capable of inciting transudation.

Carotid-cavernous sinus fistulae with their high-flow arteriovenous communication have been easily recognised since Traver's initial description in 1811. In recent years, with the advent of such major advances in the technology of cerebral angiography as selective catheterisation, magnification, and subtraction, a second type of arteriovenous communication has been recognised (Hayes, 1963; Takekawa and Holman, 1965; Mingrino and Moro, 1967; Clemens and Lodin, 1968a; Newton and Hoyt, 1968; Rosenbaum and Schechter, 1969; Newton and Hoyt, 1970; Taniguchi *et al.*, 1971; Houser *et al.*, 1972; Aminoff, 1973; Schlezinger and Schatz, 1973; Katsioris *et al.*, 1974). These dural arteriovenous fistulae are 'low flow' communications between small meningeal branches of both the external and internal carotid arteries and basal venous sinuses, frequently the cavernous sinus or its tributaries. Although the exact pathophysiology of the dural arteriovenous fistula is obscure, current theories include the spontaneous opening of congenital arteriovenous shunts (Clemens and Lodin, 1968b; Aminoff, 1973) and the rupture of thin-walled dural arteries within venous sinuses secondary to minimal trauma (Katsioris *et al.*, 1974) or straining (Taniguchi *et al.*, 1971), with pre-existent vascular disease playing an uncertain role (Newton and Hoyt, 1970).

A clinical entity generally distinct from the more florid ocular syndrome of carotid-cavernous fistulae results from the 'low flow' character of these small dural arteriovenous communications. The presentation is commonest over the age of 50, with an

apparent predisposition in women. The most frequent presenting symptoms are headache or orbital pain of unilateral occurrence and often severe nature. Diplopia is usual and an ipsilateral abducens paresis typically the source. Mild non-pulsating proptosis, conjunctival injection, chemosis, and increased intraocular pressure are generally present.

Misdiagnosis early in the course is the rule. Migraine, cluster headaches, endocrine exophthalmos, chronic conjunctivitis, episcleritis, iritis, and orbital tumour have been the most frequent diagnostic errors. The absence of an objective bruit is widely accepted as the major pitfall in diagnosis. A significant number of patients without an objective bruit, however, will provide historical evidence for one at some point in the clinical course. The presentation of the fistula is spontaneous and the course frequently characterised by remission and recurrence of typical signs and symptoms. It may be bilateral but most frequently is unilateral. Some cases have been asymptomatic and incidentally identified.

If spontaneous choroidal detachments and dural arteriovenous fistulae have more than a chance association, common signs and symptoms should be definable, providing a plausible cause-and-effect relationship. The cohesive elements must then gain support from clinical observations. These criteria appear to be satisfactorily confirmed in the following discussion.

It is clear from the review of cases available in the literature of both spontaneous choroidal detachment and dural arteriovenous fistulae of the cavernous sinus that a group of signs and symptoms are common to selected cases of both clinical entities. These include: mild proptosis, conjunctival injection, orbital pain, moderate increase in intraocular pressure, and evidence of retinal venous stasis. Further, both share a spontaneous mode of onset and a course frequently characterised by remitting and recurrent symptomatology. Finally, both tend to resolve spontaneously after periods measured in weeks to months.

A single case reported by Woillez *et al.* (1967) provides the only previous documentation of the association of spontaneous choroidal detachment and carotid-cavernous sinus fistulae. Their case differs from ours in that it was a unilateral post-traumatic carotid-cavernous fistula with the choroidal detachment occurring in the ipsilateral eye following a third surgical procedure directed at obliteration of the 'high flow' fistula. Our case more clearly substantiates a significant association, if not cause-and-effect relationship, of spontaneous choroidal detachment and 'low flow' dural arteriovenous fistulae. Another case report of interest in the

Table 1 *Tabulation of documented and possible cases of associated dural arteriovenous fistula and serous choroidal detachment*

	Author	Case	Age	Sex	Clinical diagnosis	Proptosis	Conjunctival injection	Oculomotor abnormality	Pain
D O C U M E N T E D	Harbison <i>et al.</i>	1	70	M	Bilateral choroidal detachment and bilateral dural A-V fistulae	Yes	Yes	Yes	Yes
	Cogan 1960	1	53	F	Bilateral retinal detachment and carotid-cavernous fistula (presumptive)	Yes	Yes	Yes	Yes
	Wuillez <i>et al.</i> , 1967	1	38	M	Carotid-cavernous fistula, choroidal and retinal detachment ipsilaterally	Yes	Yes	Not recorded	Yes
P O S S I B L E	Preisler 1964	1	63	M	Tenonitis-scleritis with ipsilateral choroidal detachment	Yes	Yes	Yes	Yes
	Lewallen 1957	1	103	M	Orbital cellulitis or tumour, with bilateral exudative retinal detachments	Yes	Yes	Not recorded	Yes
	Csillag 1937	1	34	F	Exudative retinal detachment. Tentative scleritis and MS	Yes	Yes	Yes	Yes
	Horay 1935	1	46	M	Exudative retinal detachment. Tentative bilateral scleritis	Yes	Yes	Not recorded	Yes
	Scheie 1974	1	28	M	Spontaneous choroidal detachment	No	Yes	No	Yes
		2	64	F	Spontaneous choroidal detachment episcleritis	Yes	Yes	No	No
	McClure 1967	1	66	M	Bilateral choroidal detachment	No	No	No	Yes
	Ursin 1965	1	63	F	Acute glaucoma with choroidal detachment	No	Yes	No	Yes
	McDonald 1964	1	61	M	Choroidal detachment and dysthyroid exophthalmous	Yes	Yes	No	No
		1	42	M	Choroidal detachment R/O melanoma	Not recorded	Not recorded	Not recorded	Not recorded
	Fleischer 1921	2	35	M	Choroidal detachment, R/O intraocular tumour	Not recorded	Yes	Not recorded	Not recorded
		1	32	F	Posterior scleritis, retrobulbar neuritis and choroidal detachment	No	Yes	Yes	Yes

present discussion is that of Cogan (1960). He presented an instance of bilateral exudative non-rhegmatogenous retinal detachment occurring in association with a presumed but unproved carotid-cavernous sinus fistula. One wonders if a spontaneous choroidal detachment did not precede the retinal separation. In addition the clinical picture reported suggests a low-flow dural fistula as opposed to a high-flow carotid-cavernous fistula.

In reviewing published reports of spontaneous choroidal detachment one can identify a number of cases based on the previously noted signs and symptoms in which an occult dural arteriovenous fistula of the cavernous sinus can reasonably be suspected as the cause (Lewallen, 1957; McDonald *et al.*, 1964; Preisler, 1965; Ursin, 1965; McClure, 1967; Scheie and Morse, 1974). These cases have been tabulated and recorded in Table 1 along with the apparently

proved cases as possible cases of choroidal detachment secondary to dural arteriovenous fistula.

It appears relatively clear that a dural arteriovenous fistula in which the venous drainage is directed anteriorly into the superior and inferior orbital veins will result in increased orbital as well as ocular venous pressure. This in conjunction with reduced mean arterial pressure resulting from arterial shunting will produce tissue hypoxia. This sequence of events clearly provides the intraocular haemodynamic basis necessary for choroidal congestion, transudation, and finally detachment, as well as the additional clinical signs and symptoms common to both entities (Sanders and Hoyt, 1969). If these mechanisms are accepted, it would seem established that dural arteriovenous fistulae of the cavernous sinus region are one of the primary disease entities productive of secondary choroidal detachments.

Intraocular pressure	Retinal venous stasis	Course
Increased	Yes	Bilateral resolution over 4 months following transcranial orbital decompression
Not recorded	Cloudy vitreous	Spontaneous resolution in 4 months
Decreased	Yes	Spontaneous resolution in 3 days
Not recorded	No	Treated with local and systemic steroids, mydriatic and antibiotics. Resolved in 4 weeks
Increased	No	Spontaneous resolution in 2 weeks. Treated with antibiotics
Normal	Yes	Spontaneous resolution after waxing and waning course
Not recorded	Yes	Spontaneous resolution after waxing and waning course
Decreased	No	Waxing and waning course
Normal	No	Resolved in 1 month
↑ OD ↓ OS	No	Treated with steroids. Spontaneous resolution in 5½ months
Increased	No	Spontaneous resolution in 2 weeks
Increased	No	Eye enucleated for suspected melanoma
Increased	Yes	Eye enucleated for suspected melanoma
Increased	Yes	Waxing and waning course. Patient died of other disease
Normal	Disk and retinal oedema	Waxing and waning course with spontaneous resolution

The case presented as well as those identified and suspected in the literature lends clinical support to this contention.

If this premise is accepted, it would seem reasonable to consider selective cerebral angiography in appropriate cases presenting spontaneous choroidal detachments. These cases should exhibit a significant number of those features common to dural arteriovenous fistulae. We would suggest these include: proptosis, conjunctival injection, increased intraocular pressure, pain, and diplopia. If a realistic diagnosis approach to the investigation of spontaneous choroidal detachment is pursued, we may expect the incidence of inciting dural arteriovenous fistulae to be appropriately determined.

Presented to the American Ophthalmological Society, Hot Springs, Virginia, 29 May 1975.

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