palsies, and/or signs related to venous congestion in the orbit. CT scanning alone is usually not useful in demonstrating the DAVM. Superselective angiography can help to determine the exact location and haemodynamic features of DAVM. Venous drainage into the pial venous system via the deep middle cerebral vein can give rise to cerebral pail venous hypertension, which may cause papilloedema, headache, hydrocephalus, and increase the risk of cerebral haemorrhage.[5] Occipital DAVMs may develop carotid-carvenous fistula and cause anterior intracranial venous drainage.[6] The unusual feature of the present case was a DAVM located between the ascending pharyngeal artery and the inferior petrosal sinus, giving rise to severe ocular complications. The development of visual loss and the total ophthalmoplegia may have been because of persistent severe venous hypertension of the ophthalmic veins and cavernous sinus. Visual dysfunction in DAVMs is usually the result of venous hypertension, venous infarct, haemorrhage,[7] or rarely a result of sinus thrombosis.[8] Cerebral angiography can aid prompt diagnosis and treatment.

We would like to point out this rare original location and retrograde pattern of petrosal sinus DAVM. It can mimic the clinical presentations of a cavernous sinus DAVM, and requires cerebral angiography for prompt diagnosis and management. It is also emphasized that although DAVM is not a common cause of visual dysfunction, early diagnosis can avoid serious ocular complications.

Acknowledgements

No public and private support was received for the work and none of the authors have any commercial interest in the material mentioned herein.

References

1 Hofmeister C, Stapf C, Hartmann A, Sciacca RR, Mansmann U, TerBrugge K *et al*. Demographic, morphological and clinical characteristics of 1289 patients with brain arteriovenous malformation. *Stroke* 2000; **31**: 1307–1310.
2 Houser OW, Baker Jr HL, Rhoton Jr AL, Okazaki H. Intracranial dural arteriovenous malformation. *Radiology* 1972; **105**: 55–64.
3 Chaudhary MY, Sachdev VP, Cho SH, Weitzner I Jr, Puljic S, Huang YP. Dural arteriovenous malformation of the major venous sinuses: an acquired lesion. *Am J Neuroradiol* 1982; **3**: 13–19.
4 Houser OW, Campbell JK, Campbell RJ, Sundt Jr TM. Arteriovenous malformation affecting the transverse dural venous sinus—an acquired lesion. *Mayo Clin Proc* 1979; **54**: 651–661.
5 Award IA, Barrow DL. *Cavernous Malformation*. American Association of Neurological Surgeons: Park Ridge, 1993.
6 Huna-Baron R, Setton A, Kupersmith MJ, Berenstein A. Orbital arteriovenous malformation mimicking cavernous sinus dural arteriovenous malformations. *Br J Ophthalmol* 2000; **84**: 771–774.
7 Kupersmith MJ, Berenstein A, Nelson PK, ApSimon HT, Setton A. Visual symptoms with dural arteriovenous malformations draining into occipital veins. *Neurology* 1999; **52**: 156–162.
8 Gelwan MJ, Choi IS, Berenstein A, Pile-Spellman JM, Kupersmith MJ. Dural arteriovenous malformations and papilledema. *Neurosurgery* 1988; **22**: 1079–1084.

C-C Tsai[1,2], Y-C Lin[1,2], H-C Kau[3,4], S-C Kao[1,2], W-M Hsu[1,2] and MMH Teng[5,6]

[1]Department of Ophthalmology
Taipei Veterans General Hospital
No. 201, Sec. 2, Shih-Pai Road
Taipei, Taiwan

[2]Department of Ophthalmology
National Yang-Ming University, Taiwan

[3]Department of Ophthalmology
Taoyuan Veterans Hospital, Taiwan

[4]Institute of Clinical Medicine
National Yang-Ming University, Taiwan

[5]Department of Radiology
Taipei Veterans General Hospital, Taiwan

[6]Department of Radiology
National Yang-Ming University, Taiwan

Correspondence: Y-C Lin
Fax: +886 2 2555 1303
E-mail: linycz@vghtpe.gov.tw

Sir,

Massive subretinal bleed in a patient with background diabetic retinopathy and on treatment with warfarin

Eye (2003) **17,** 649–652. doi:10.1038/sj.eye.6700450

Massive intraocular haemorrhage is a devastating and well-known complication of anticoagulation treatment in eyes with exudative age-related macular degeneration.[1] Thromboembolism, prosthetic heart valves, previous myocardial infarction, and strokes are some of the most common conditions where anticoagulation treatment is indicated.[2] However, intraocular bleed is not common in normal nonpredisposed eyes (without any neovascularization) even with anticoagulation treatment. Background diabetic retionopathy alone is not known to be related to massive retinal bleed. Here we report a case of massive subretinal bleed in a patient with

background diabetic retinopathy who was taking warfarin.

Case report

A 76-year-old woman presented to the eye A&E with sudden painless loss of vision in her right eye of 7 days duration. In the past ocular history, she had an exaggerated subconjunctival haemorrhage and haemotoma in her left eye 3 years ago after a minor blunt trauma, which resolved without any sequlae. She had been diagnosed with NIDDM a year before and blood sugar was well controlled on *diet* alone. She had undergone aortic valve replacement surgery in 1990 and was on a maintainance dose of warfarin, 2 mg/day. She was taking no other medication at the time and was not suffering from any other systemic diseases.

On examination, her VA was perception of light in the right eye and 6/12 with full correction in the left eye, which had early cataract. There was no relative afferent pupillary defect. The intraocular pressures were 16 mmHg in both eyes. The anterior chambers were of average depth and the anterior vitreous was clear in both eyes. A dilated fundus examination of the right eye revealed a slate grey and dark red mass, lying under the major blood vessels of the retina and extending from the macula up to the temporal disc margin. It was slightly elevated from the surface and there was no vitreous haemorrhage (Figure 1a). A clinical diagnosis of right massive subretinal bleed was made.

An FFA was performed, which showed blocked fluorescence in the area with the subretinal bleed with no evidence of leakage (Figure 1b). B-scan ultrasonography did not show any changes attributible to intraocular tumours and also reconfirmed the clinical diagonosis of subretinal bleed (Figure 1c).

A full-blood count (FBC), international normalized ratio (INR) and bleeding, and clotting profiles were urgently carried out. The only positive findings of note were an INR of 3.8 and 4.5 carried out in a span of a week. Her platelet count, WBC count, RBC count, haemoglobin level, and other bleeding and clotting profiles were all within normal limits.

Comment

Warfarin is the most commonly prescribed oral anticoagulant both for prophylaxis as well as treatment of cerebro-vascular and cardio-vascular disorders. Up to 3% of patients on long-term warfarin treatment develop retinal haemorrhage, mostly visually insignificant.[3] Patients with hypertension and older than 75 years are at increased risk of retinal bleed because of warfarin.[3] Clinical indications for warfarin and the therapeutic range of anticoagulation, monitored by the INR is given in Table 1.[2,5]

Warfarin acts by inhibiting the vitamin-K-dependent carboxylation of factors II, VII, IX and X in the liver,[2] and induces a state analogous to vitamin K deficiency.[4] Most patients require a daily maintainence dose of 2.0–7.5 mg of warfarin to remain anticoagulated. About 10% of patients on an oral anticoagulant for 1 year have a serious complication requiring medical supervision and 0.5–1% have a fatal haemorrhagic event (eg cerebral haemorrhage) despite careful medical management.[4]

Superstein *et al*[3] reported the mean duration of warfarin therapy to retinal bleeding to be 1.9 years (range 2 months to 5.5 years). Additional treatment with aspirin in patients receiving warfarin double the risk of bleeding,[6] our patient was on no other medication. Several studies have examined the minimum effective dose of warfarin in a variety of conditions. Optimal risk–benefit appears to be at an INR of 2–3 for almost all conditions, with the exception of prosthetic valves requiring an INR of 3–4.5.[2,5]

El Baba *et al*[7] reported that 19% of the patients with ARMD who developed massive intraocular haemorrhages were taking warfarin or aspirin. Other ocular lesions that can also bleed and present similarly, that is, intraocular tumours particularly malignant choroidal melanoma, need consideration and must be ruled out.[8,9]

Our patient had an INR of 3.8 and 4.5, which although high is an acceptable level of anticoagulation, considering the fact that she had a prosthetic aortic valve. We took advice from haematology and cardiology departments regarding her further management and were advised not to stop or reduce warfarin as these could put her into major risk of thrombo-embolism. She had a few microaneurysms in either eye. We propose that the subretinal bleed was a complication of the long-term warfarin treatment and high level of anticoagulation. We feel it may have been compounded by the coexisting background diabetic changes in the retina through the vascular damages attributible to diabetes.

There are few effective theraupeutic options once the haemorrhage has occurred. The surgical removal (Pars plana vitrectomy with internal drainage) of a massive subretinal haemorrhage has been described by several authors, but the visual outcome is poor because of delays between the onset of the haemorrhage and the actual vitrectomy and the amount of damage to the photoreceptors.[10] Among the newer treatment modalities, low-dose intravitreal tissue plasminogen activator injection and pneumatic displacement using an

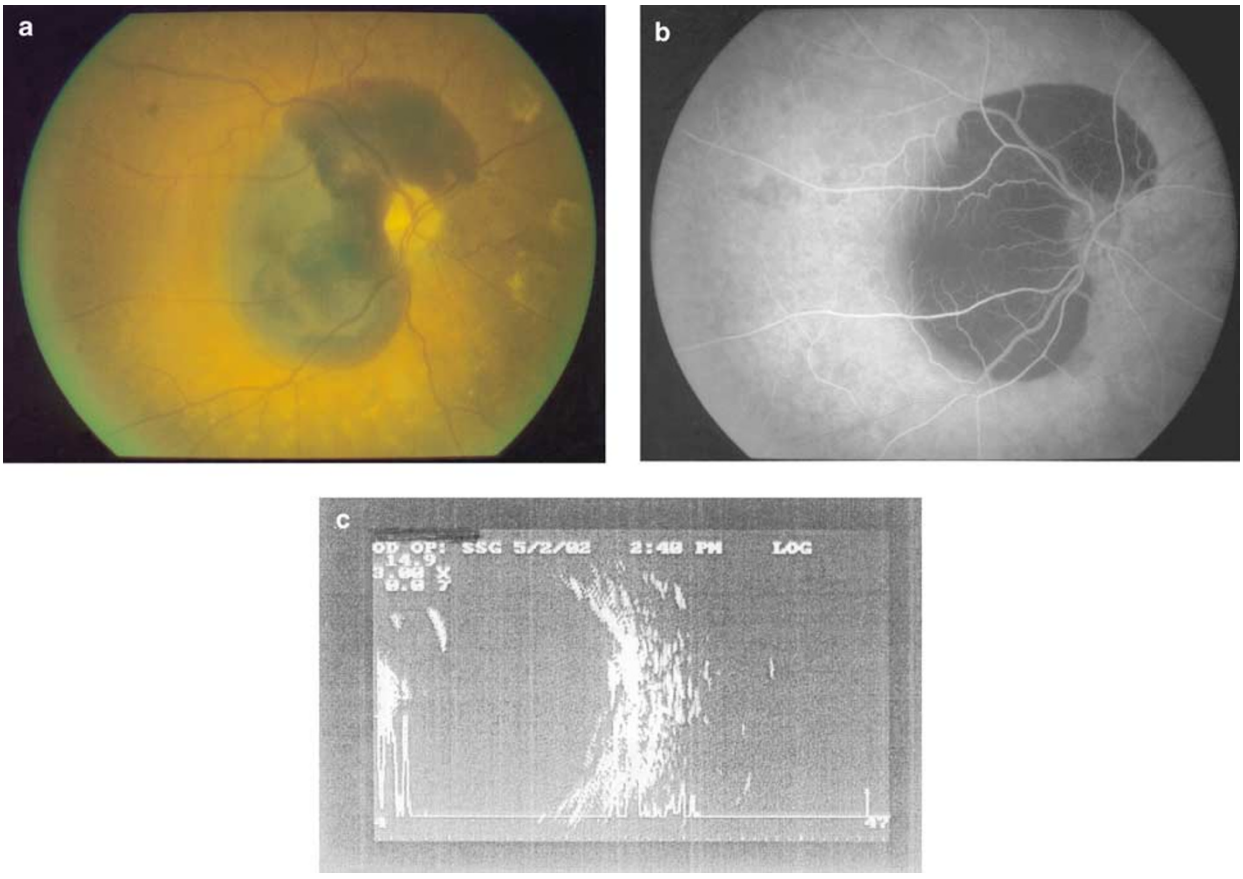

Figure 1 (a, b) Massive subretinal bleed in the right eye, involving the macula and the peripapillary areas and (c) B scan ultrasonography showing subretinal bleed.

Table 1

INR	*Clinical indications*
2.0	Prophylaxis of DVT, high-risk surgeries, for example, hip surgery
2.0–3.0	Treatment of DVT and pulmonary embolism, TIA, atrial fibrillation
3.0–4.5	Mechanical prosthetic cardiac valves, recurrent DVT

Proposed by the British Society for Hematology, 1990.

expansile gas bubble is promising. However, this needs validation with prospective randomized controlled trials.[11] We managed our patient conservatively.

References

1 Tilanus MA, Vaandrager W, Cuypers MH, Verbeek AM, Hyong CB. Relationship between anticoagulant medication and massive intraocular haemorrhage in age-related macular degeneration. *Greafes Arch Clin Exp Ophthalmol* 2000; **238**(6): 482–485.

2 Edwards CRW, Bouchier IAD, Haslett C, Chilvers ER. *Davidsons Principles & Practice of Medicine*, 17th ed. Churchill Livingstone: New York, 1995, pp 840.

3 Superstein R, Gomolin JE, Hammouda W, Rosenberg A, Overbury O, Arsenault C. Prevelance of ocular haemorrhage in patients receiving warfarin therapy. *Can J Ophthalmol* 2000; **35**(7): 385–389.

4 Harrisons. *Principles of Internal Medicine*, 12th ed. McGraw Hill: New York, 1991, pp 1512.

5 Praveen Kumar, Michael Clark. *Clinical Medicine*, 3rd ed. Saunders Co: London, 1996, pp 348–350.

6 Holder R. Spontaneous hyphema as a result of systemic anticoagulation in previously abnormal eyes. *Postgrad Med J* 1991; **67**: 1008–1010.

7 El Baba F, Jarrett WH, Shields JA. Massive sub-retinal haemorrhage complicating age-related macular degeneration: clinicopathological correlation and role of anticoagulants. *Ophthalmology* 1986; **17**: 227–230.

8 The Collaborative Ocular Melanoma Study Group. Accuracy of diagnosis of choroidal melanomas in the Collaborative Ocular Melanoma Study. COMS Report No. 1. *Arch Ophthalmol* 1990; **108**: 1268–1273.

9 Robertson DM, Campbell RJ. Errors in the diagnosis of malignant melanoma of the choroid. *Am J Ophthalmol* 1979; **87**: 269–275.

10 Wade EC, Flynn HW, Olsen KR, Blumenkranz MS, Nicholson DH. Sub-retinal haemorrhage management by Pars plana vitrectomy and internal drainage. *Arch Ophthalmol* **108**: 973–978.
11 Handwerger BA, Blodi BA, Suresh R, Chandra, Olsen TW, Stevens TS. Treatment of submacular haemorrhage with low-dose intravitreal tissue plasminogen activator injection and pneumatic displacement. *Arch Ophthalmol J* 2001; **119**: 28–32.

A Raj, R Sekhri, A Salam and P Priya

Department of Ophthalmology
Hull Royal Infirmary
Anlaby Road Hull, HU3 2JZ, UK

Correspondence: A Raj
Tel: +07763 340 100
E-mail: doc_akash@yahoo.com

Sir,

Wegener's granulomatosis causing lid destruction: a further sight-threatening complication

Eye (2003) **17,** 652–654. doi:10.1038/sj.eye.6700410

Wegener's granulomatosis (WG) is a presumed autoimmune disease causing systemic small-vessel vasculitis, necrotising granulomas of the respiratory system, and a focal necrotising glomerulonephritis. Cytotoxic/steroid combination treatment has improved the mean survival from 5 months to a 20-year survival of more than 80%.[1,2] There is a better prognosis with the renal-sparing form of this disease.

Ear, nose, and throat disease is the most common presenting feature of WG, and is found in 75% of patients at diagnosis. Ocular complications from WG occur in around 50% of patients.[2] We present a patient with an unusual, visually threatening and surgically challenging form of orbital WG.

Case report

A 71-year-old man with a 4-year history of extrarenal ANCA-positive WG was referred for an oculoplastic opinion in January 2002. He had initially presented with epistaxis, sinusitis, and left scleritis. In spite of treatment with Campath-1H monoclonal antibody and pulsed cyclophosphamide and methylprednisolone, disease progression occurred. He developed nasal septum and turbinate loss, bony destruction of the medial maxillary sinus walls and ethmoid sinus floors (Figure 1), and an oro-antral fistula. A progressive bilateral medial lower lid ectropion appeared in early 1999.

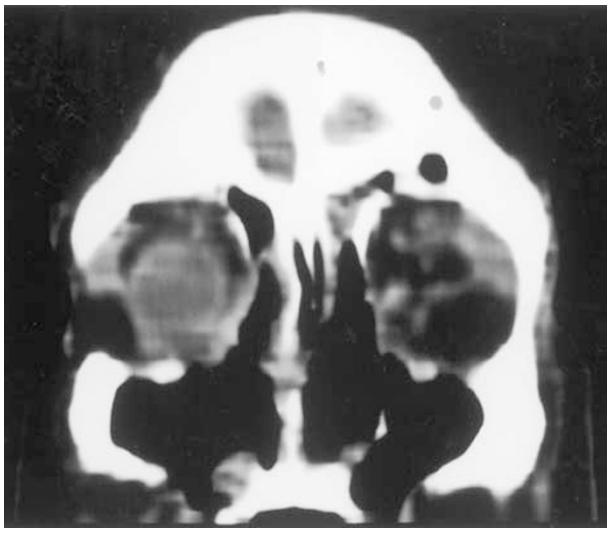

Figure 1 Destructive effects of WG: CT scan, reformatted coronal view. The left eye required enucleation following scleritis-induced perforation. Extensive sinonasal destruction with partial loss of the inferomedial orbital walls and nasal septum.

The left eye was enucleated in 2000, having become painful, blind, and phthisical following a scleritis-induced scleral perforation in August 1999. Abduction of the right eye had also become limited as a further consequence of his orbital WG.

Of primary concern was the progressive destruction of the medial canthal regions bilaterally, with resultant incomplete lid closure. This was causing signs of progressive (right) inferomedial corneal exposure and pannus formation. The bulbar conjunctiva was injected, but there was no active scleritis or marginal corneal thinning. The right lower lid medial attachment was limited to a residual thin strand of canthal tendon. Sclera and adjacent sinus/nasal cavity were visible through the resultant necrotic lid defect, and the surrounding lid skin was injected. The defect extended vertically to involve the upper lid medially, although the upper lid medial canthal attachments appeared secure. Surgery was proposed in view of the risk of progressive corneal damage and possible perforation of this patient's only eye, which had a corrected Snellen visual acuity of 6/12.

With concerns about lid vascular supply/healing potential of the affected lid area and surgery acting as a trigger to local disease flare-up, intervention was restricted to the minimal. The residual medial canthal attachment was divided and a permanent medial tarsorrhaphy was performed, leaving a limited palpebral fissure for axial vision (Figure 2).