False aneurysm of the internal carotid artery in Behçet’s disease: successful combined endovascular treatment with stent and coils

Sir, A 47-yr-old woman, with a 7-yr history of Behçet’s disease, was admitted due to a disabling weakness in her right arm. Behçet’s disease was identified following a history of polyarthralgias, recurrent oral aphthous ulcers, recurrent venous phlebitis and folliculitis. For the last 7 yr, the patient has been successfully treated with acetylsalicylic acid (100 mg/day) and colchicine (1 mg/day). At the time of admission, there was no clinical or biological sign of evolution of the Behçet’s disease. Physical examination identified a left cervical mass; following an angiography and magnetic resonance imaging (MRI), this was shown to be a false aneurysm of the left internal carotid artery, 30 mm in diameter. This false aneurysm, located at the level of the C1 vertebra (Fig. 1), was accompanied by small aneurysms of the right cervical and left vertebral arteries. Its localization precluded any surgical intervention due to the inaccessibility of the vessel at the base of the skull. Therefore, the patient was treated by endovascular placement of a metallic stent combined with the use of coils. The stent was placed at the level of the aneurysm. A microcatheter was then introduced into the aneurysm through the wall of the stent. The implantation of 19 coils resulted in a dense packing of almost the entire aneurysm. Control angiograms showed that the normal calibre of the internal carotid artery had been restored and that the aneurysm remained occluded (Fig. 2).

Renal and pulmonary angiographies did not identify any further aneurysms. Treatment with acetylsalicylic acid and colchicine was stopped, and treatment with azathioprine (100 mg/day) was started. Four years on, Behçet’s disease has not recurred. The angiographic and MRI controls indicate a stable stent position without any change in the small vertebral artery aneurysms and no new aneurysm.

Arterial aneurysms are a classical and severe complication of Behçet’s disease. These aneurysms, whose incidence ranges from 2 to 6% [1, 2], are commonly located in the abdominal aorta as well as in femoral or pulmonary arteries. Aneurysms of internal carotid arteries are seldom reported [1, 2]. Operative therapy is usually recommended for the management of arterial aneurysms since their rupture is the primary cause of death in patients with Behçet’s disease. In this case, we can report the long-term success of treatment by the endovascular placement of a stent in the arterial aneurysm of a patient with Behçet’s disease. This new endovascular treatment

Fig. 1. Angiogram showing a false aneurysm and small aneurysms of the right cervical and left vertebral arteries.

Fig. 2. Control angiogram showing that the normal calibre of the internal carotid artery had been restored and that the aneurysm remained occluded.
[3, 4] could be a good opportunity for treating false aneurysms in patients with Behcet’s disease when the localization precludes any surgical intervention. Adjuvant immunotherapy with azathioprine, with or without high doses of corticosteroids, is usually required to control the formation of new aneurysms [5].

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Combination therapy in rheumatoid arthritis: a comment

Sr, We read with interest the review by Verhoeven et al. [1] on combination therapy in rheumatoid arthritis (RA). With the recent emphasis on aggressive treatment of RA and the almost universal use of combinations of disease-modifying anti-rheumatic drugs [2], this was both timely and thorough. However, while in general we agree with the conclusions of this review, several errors occurred in interpreting the effectiveness of our study of the combination of methotrexate (MTX), sulphasalazine (SSA) and hydroxychloroquine (HCQ), and greatly diminished the magnitude of the long-term benefit of that therapeutic regimen [3].

Our study, as designed and reported, was a 2 yr, double-blind, randomized controlled trial; it was not a 9 month trial, as reported by Verhoeven et al. While it is true that only half of our patients were available for blinded evaluation at 2 yr (the majority of the patients in the MTX-alone and the SSA–HCQ groups having already failed for efficacy reasons by that time), to see which therapy was best able to produce a 50% improvement in composite criteria and maintain that improvement for 2 yr was exactly the point of the study. Two years is, admittedly, a short time in a lifelong disease process like RA, but it is twice as long as the next longest combination trial reported and, therefore, arguably more relevant to clinicians caring for patients with RA.

If our study had been analysed at the 2 yr time point, based on the criteria set down in the review, it would have easily met the criteria for ‘substantially more effective’ (as defined). This comparison at 2 yr is even more significant since benefit was shown for MTX–SSA–HCQ over MTX alone even in the patients who had met 50%-improvement criteria (the only patients left in the study at that point).

A comparison of our study with the MTX–cyclosporine (CSA) study [4] may be illustrative. The addition of CSA to patients who have had a suboptimal response to MTX (mean 10.2 mg/week) was shown to produce a 20% improvement from baseline in 32% more of the active-treated patients compared to placebo-treated patients [48% (20% response) in active minus 16% response in placebo-treated patients] in a short-term trial (6 months). The combination of MTX–SSA–HCQ for 2 yr, on the other hand, has been shown to result in 77% of the MTX–SSA–HCQ-treated patients receiving a 50% composite improvement compared with 33% of the patients in the MTX-alone group (mean dose of MTX 16.6 mg/week).

The implication that CSA and low-dose MTX has stronger evidence to support clinical efficacy than the combination of MTX–HCQ–SSA is misleading to readers.

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Use of prophylaxis for corticosteroid-induced osteoporosis in hospital practice

Sr, We were interested in the recent abstract of Jordan and Witherington: ‘A questionnaire survey of the use of prophylaxis for corticosteroid induced osteoporosis in hospital practice’ [1].

We have already conducted both an internal audit and subsequently a national survey on this topic specifically looking at ophthalmologists. An internal audit of practice in one ophthalmology department [2] showed that little consideration was given to the problem of osteoporosis, and that little or no advice was given to the patients commencing corticosteroids. We subsequently conducted a survey of all UK ophthalmologists with a response rate of 81% [3]. While 75% of respondents regularly prescribed prednisolone in
dosages of >5 mg for at least 3 months, only 25% gave
patients advice on osteoporosis. This dosage is a recom-

mended level for consideration of bone densitometry in
a recent government report [4]. Few consultants in our
survey used bone densitometry.

We agree that this lack of awareness is a serious
problem and one that needs highlighting. It is crucial
for hospital-based specialists from any specialty to liase
with the GP, who should be aware of the implications
and problems associated with steroid usage. This is
especially important where that specialist is unfamiliar
with the problems of steroid usage. Consideration should
be given to the following. (1) Direct osteoporosis advice
in the form of a discussion and a leaflet at that visit.
Simple measures such as calcium supplementation in
the diet, e.g. milk, dairy products and fish. Post-
menopausal women should consider starting HRT. (2) A
letter to the GP stating directly the potential of this
problem. (3) If the course of steroid treatment is likely
to be prolonged or indeed the patient has already been
treated for some time, provision should be made for
bone densitometry measurement and further treatment
if necessary following recent guidelines [5]. This may
involve referral to another service, directly or via the GP.

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1. Jordan AC, Witherington RH. A questionnaire survey of the use
of prophylaxis for corticosteroid induced osteoporosis in hospital
2. Hodgkins PR, Hull RG, Vakalis A et al. Long term oral cortico-
steroids and osteoporosis prevention in an ophthalmology clinic.
3. Hodgkins PR, Hull RG, Evans AR, Jeffrey MN. Osteoporosis: a
4. Barlow DH. Osteoporosis. London: Chief Medical Officers
5. National osteoporosis society—Guidance on the prevention and
management of corticosteroid induced osteoporosis. Bath: National

An unusual cause of blindness in Wegener’s granulomatosis

Sr, Although ocular involvement is the presenting
feature in 16% of cases of Wegener’s granulomatosis
(WG), sight-threatening complications are rare. Retinal
artery occlusion is very unusual, with only three cases
reported since 1960.

A 53-yr-old Caucasian woman was admitted with
sudden loss of vision in the right eye. Fundoscopy
revealed severe right arteriolar occlusion, resulting in
macular infarction. Two months previously, inflammation
of both tympanic membranes had been noted at the
otorhinolaryngology clinic and topical corticosteroid
therapy commenced. On admission, she had haematuria
and proteinuria, CRP 100, ESR 103 and elevated liver
enzymes (ALT 160 IU/l, GT 160, alkaline phosphatase
175). A chest radiograph was normal. WG was sus-
pected, and renal biopsy showed focal segmental necro-
tizing glomerulonephritis with a few immunological
deposits, characteristic of WG. Treatment with cyclo-
phosphamide (15 mg/kg) and methylprednisolone
(15 mg/kg) was commenced. Subsequently, her serum
showed antineutrophil cytoplasmic antibodies (ANCA),
of which 83% were specific for proteinase 3 (PR3). Tests
for lupus anticoagulant activity and for anticardiolipin
antibodies were normal. She improved clinically, but
fundoscopic examination showed optic atrophy, and
signs of old, extensive retinal arterial occlusion.

A 55-yr-old man complained of cough and lethargy.
Progressive muscle weakness led to hospital admission,
where pyrexia (38°C), splinter haemorrhages and
one episode of haemoptysis were noted. Urinalysis
showed proteinuria and haematuria, white cell count
15.0 × 109/l, CRP 350 IU/l and proteinuria 2.48 g/24 h.
Coagulation and biochemistry were normal. He
developed multiple sensori-motor neuropathies.
Immunofluorescence detected a cANCA pattern, crude
ELISA level 25% (normal: <16%), PR3 20%. Renal
biopsy showed pauci-immune focal segmental necrotiz-
ging glomerulonephritis, again characteristic of WG. He
developed painful scleritis of the left eye and loss of
vision, fundoscopy showing central retinal artery occlu-
sion. He commenced pulse therapy, with good resolution
of his inflammatory markers and neurological deficits,
but visual acuity did not recover.

Untreated, classical WG can be rapidly fatal, and
combination treatment with corticosteroids and cyclo-
phosphamide has considerably improved survival, with
variable recovery in affected organs. Exophthalmos and
optic neuritis are mentioned in the early case descriptions
of WG [1]. Further ocular manifestations of WG have
since been documented [2–4], occurring as a presenting
feature in 16% [5]. However, serious sight-threatening
complications attributable to WG are rare, noted in
those with orbital disease, central retinal vein occlusion
and corneo-scleral inflammation. Rubeciosis irides and
neovascular glaucoma are also known to cause blindness
in WG [6]. There have been only three previous reports
of loss of vision due to retinal artery obstruction [7–9].
In two of these cases [8, 9], impending occlusion in the
other eye was reversed with immunosuppressive therapy.
The cases we have described had multi-organ involve-
ment which led to the rapid diagnosis of WG. Tests for
ANCA and their specific targets have facilitated the
diagnosis of WG, although current criteria do not
include ANCA (see [10] for a review). PR3 specificity,
its strongly associated with WG, has been shown
prospectively to be present in 32% of cases with eye
involvement [11]. Nonetheless, the diagnosis of WG still
relies on a clinical pattern and on histological appear-
ances, both of which rely in turn on clinical suspicion
of the condition. Therefore, it is important to highlight
that WG must be considered in the differential diagnosis
Letters to the Editor

of retinal artery occlusion, recognizing that eye disease may be a presenting feature of this systemic disease.

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Alfuzosin is not associated with dermatomyositis

Sir, In the recent paper by Vela-Casasempere et al. [1], the evolution of the patient described is not consistent

with the characteristics of an agent-induced disease, especially regarding the need for corticosteroids to improve evolution. When a drug induces a disease, withdrawal of the drug is usually sufficient to manage the patient and improve his condition.

In the case in question, the evolution of the disease with time is particularly important, since it is known that dermatomyositis is a condition which could appear as the first manifestation of an occult neoplasm. Consequently, it is pertinent to ask how much time elapsed between diagnosis and the date the paper was submitted for publication. In this case, I know for a fact that very little time elapsed because I work in the same hospital and department as the author, who previously held my post. Diagnosis was carried out in February and the paper was accepted in May. Moreover, in my opinion, there is no justification for considering the disease to be the consequence of any drug.

The patient described in the paper was admitted to hospital 7 months after initial diagnosis because he had haemoptysis, weight loss and a subclavicular nodule. His dermatomyositis remained active even with corticosteroids, and the X-ray showed a nodule in the middle of the left lung. Biopsy of the nodule confirmed a microcytic carcinoma of the lung. The patient is now undergoing oncology treatment. Therefore, there is no reason in this case to ascribe the dermatomyositis to the drug alfuzosin, but to the lung carcinoma.

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