Retinal arterial occlusion associated with resistance to activated protein C

EDITOR,—Resistance to activated protein C (APC) was described in 1994 as a thrombophilic factor responsible for deep venous thrombosis and pulmonary embolism.1 It represents the best-known cause of thrombophilic disorder in the normal population (5%). APC is an important component of the physiological anticoagulant system that inhibits factors V and VIIIa. The resistance to APC is related to a single mutation in the factor V gene which causes the switch from arginine to glutamine at position 506. This mutation of V blocks the site of cleavage by APC.2 Reported thrombophilic manifestations include venous thrombosis and one case of central retinal vein thrombosis.3 Arterial occlusion is not clearly associated with resistance to APC. Ischaemic stroke was recently reported in three patients with resistance to APC.2 We described a case of branched retinal artery occlusion associated with resistance to APC.

CASE REPORT
A 35-year-old non-smoking man was referred for a sudden decrease of visual acuity in the left eye. He had no familial or personal history of thrombophilic disorders. At examination, his best corrected visual acuity was 20/20 in the right eye and 20/30 in the left. A superotemporal branched retinal artery occlusion was noted in the left eye with an oedematous, whitish, retinal infarction in the affected vessel. A fluorescein angiogram confirmed the branched retinal artery occlusion (Fig 1). After 2 months, visual acuity improved to 20/25 in the left eye. He had a permanent visual field defect in the area of damaged retina. He was treated with aspirin. Cardiac rhythm was regular; transthoracic echocardiography, and carotid Doppler studies showed no abnormalities. Red blood cell, white cell, platelet counts, and erythrocyte sedimentation rate were all normal. Platelet aggregation was normal. A search for antinuclear and anticardiolipin antibodies was negative. Prothrombin time and activated partial thromboplastin time (APTT) were normal. Plasma levels of protein C, S, antithrombin III, fibrinogen, plasminogen, plasminogen activator inhibitor were within normal ranges. APC resistance was determined by evaluating the anticoagulant response of plasma samples to APC with an APTT based assay. Results were expressed as the following APC sensitivity ratio (APTT + APC)/(APTT – APC). The cut off value was 2.2. In our patient, the APC sensitivity ratio was 2.1. Heterozygous factor V Leiden mutation was disclosed by molecular analysis.

COMMENT
Occlusion of the retinal artery is more rarely encountered in younger than in older patients.4 Multiple causes of arterial occlusion in the retina were described. In a recent report, the causes of retinal arterial occlusions in 21 young adults were analysed.5 Emboli were identified in 33% of the patients. Cardiac valvular disease, including atrial myxoma, bacterial endocarditis, and mitral valve vegetation due to lupus anticoagulant, was the mainly recognised condition and was present in 19% of the patients. Other associated risk factors for cerebrovascular occlusion such as cigarette smoking, oral contraceptive use, obesity, pregnancy, and Behcet’s disease were found in 91% of the patients. Antithrombin III, protein S, or protein C deficiencies are hypercoagulable conditions that can lead to recurrent venous or arterial thrombotic events.6 Protein S deficiency was associated with a case of bilateral branch retinal artery occlusion.7 This biological abnormality was detected in only one young woman with diabetes mellitus and pregnancy in the series of Greven et al.8 Antiphospholipid antibody syndromes are thrombophilic factors that occur in patients with either lupus anticoagulants or antibodies to antiphospholipid or dissociated syndesmophil serology. Antiphospholipid antibodies can lead to recurrent arterial and/or venous thrombosis.9 APC resistance is clearly related to venous thromboembolism.2 A recent report suggests the possible role of APC resistance in arterial thrombosis.6 APC resistance was not searched for as a thrombophilic factor in retinal arterial occlusions in young adults. In our case, the cause of retinal arterial occlusion could be attributed to the heterozygous mutation of factor V. Owing to the severity of retinal arterial occlusion, long term oral anticoagulant treatment was proposed in our patient for secondary prevention of thrombosis. APC resistance should be considered in patients with retinal arterial occlusion when the usual embolic or thrombotic diseases are ruled out.

Recurrent septic retinal emboli following dental surgery

EDITOR,—Metastatic bacterial endophthalmitis following head and neck surgery is rare.1 To our knowledge, no case of recurrent septic retinal emboli with a presumed dental source has been described before.

CASE REPORT
A healthy 36-year-old white man presented to a dental surgeon with a localised peripapil are abscess at his right upper first molar, which was confirmed by dental x ray. A volume of 0.2 ml of pus was drained after local periapical infiltration with 2% lidocaine. A 1 ml of saline was administered into the root canal 1:80 000 and postoperative irrigation with 0.01% dexamethasone solution. He was prescribed oral amoxicillin/clavulanic acid 375 mg three times daily but did not start antibiotics until 12 hours after the procedure. Three days later, the patient noted sudden blurring of vision and floaters in his right eye but was systemically well.

There was no previous ocular history and the medical history was negative for intravenous drug use, rheumatic fever, or other cardiac disease. He had no risk factors to suggest systemic immunosuppression. A small exudate was noted. Significant, significantly, he was afebrile with no cardiac signs or peripheral stigmata of infective endocarditis. Uncorrected visual acuity was 6/5 in both eyes and there was no relative afferent

Figure 1 Superotemporal branched retinal artery occlusion.