A 71 year–old retired mechanic with previous asthma and hypertension presented with a one week history of progressive left homonymous hemianopia, expressive aphasia and dyspraxia. His wife reported that he had developed black spots on his ear, neck and hands over the last 10 days. He had multiple raised black lesions over his neck, ear, hand and limbs, which bled easily on contact. A serum inflammatory, infective and vasculitic screen as well as a CT chest, abdomen and pelvis were normal. CT brain found multiple haemorrhagic lesions with surrounding oedema. MRI head demonstrated multiple lesions with modest enhancement with gadolinium.

Skin biopsy of two typical lesions showed vascular haemangiomatous type lesions with organising thrombus. Special stains for organisms were unremarkable. Brain biopsy revealed similar changes with a non–specific multifocal reactive process with perivascular inflammation but no vasculitis. Dexamethasone therapy was started with transient benefit but 1 month later he developed a subacute onset right hemiparesis, dyspnoea and new skin lesions. CT chest demonstrated pulmonary emboli, originating from a proximal lower limb deep vein thrombosis. Repeat MRI of the brain demonstrated a new lesion in the left
fontal lobe. FDG whole body PET was normal. A low titre IgG kappa paraprotein was found 6 months after presentation. Propranolol was added and the dexamethasone phased out. There was steady improvement neurologically and radiologically with imaging at 4 and 14 months demonstrating gradual resolution of the cerebral lesions, no new skin lesions and an almost complete clinical recovery.

Reactive angiomas are benign vascular skin growths characterised histologically by intravascular and extravascular hyperplasia of endothelial cells and pericytes. Reactive angioendotheliomas are rare lesions classically described in association with systemic diseases such as subacute bacterial endocarditis, cholesterol emboli, monoclonal gammopathy and rheumatoid arthritis. They can histologically mimic neoplasia, however, in reactive angioendotheliomas vascular hyperplasia stops once the hypoxic stimulus has been withdrawn. Treatment includes steroids to suppress neoangiogenesis and therapy for the underlying systemic disease. More recently β-adrenergic receptor antagonists have been proposed to block angiogenesis by decreasing expression of vascular endothelial growth factor. Published literature implies these lesions only occur in the skin.

We present a patient with biopsy proven reactive angiomatosis with histologically identical symptomatic lesions in the skin and brain that both followed the same time course–neurocutaneous angiomatosis. The timing of the improvement following treatment with propranolol is intriguing.

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A CASE OF THE BLACK SPOT

Kanchan Sharma, Paul Craig, Eduardo Calonje, Nikki Cohen, James Milne and Geraint Fuller

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