with antiphospholipid syndrome but not satisfying clinical criteria for definite antiphospholipid syndrome.

The concept of probable antiphospholipid syndrome is a relatively new one and the literature is limited. Asherson et al (2007) have reported patients presenting with heart valve lesions, chorea, renal microangiopathy, thrombocytopenia, fetal loss or livedo reticularis. These patients exhibit clear features of a systemic microangiopathy but lack the clinical criteria of large vessel thrombosis or pregnancy morbidity for a diagnosis of definite antiphospholipid syndrome. These patients may develop further clinical signs in the future and eventually be diagnosed with definite antiphospholipid syndrome.

Moyssakis et al (2007) report that valvular abnormalities in antiphospholipid syndrome are usually asymptomatic and often discovered incidentally. However, significant valvular dysfunction and consequent heart failure may occur. Moreover, valvular abnormalities can be complicated by secondary infective endocarditis and embolization from vegetations leading to widespread systemic complications such as strokes, pulmonary and renal infarcts.

This patient underwent a mitral valve replacement with good result and is on lifelong anticoagulation. She is under rheumatology, cardiology and nephrology follow up. **BJHM**

Asherson RA (2006) New subsets of the antiphospholipid syndrome in 2006: "PRE-APS" probable APS and microangiopathic phospholipid syndromes. *Autoimmune Rev* **6**(2): 76–80. https://doi.org/10.1016/j.autrev.2006.06.008

Asherson RA, Pierangeli SS, Cervera R (2007) Is there a microangiopathic antiphospholipid syndrome? *Ann Rheum Dis* **66**: 429–432. https://doi.org/10.1136/ard.2006.067033

Moyssakis I, Tektonidou MG, Vasilliou VA, Samarkos M, Votteas V, Moutsopoulos HM (2007) Libman-Sacks endocarditis in systemic lupus erythematosus: prevalence, associations, and evolution. *Am J Med* **120**(7): 636–642. https://doi.org/10.1016/j.amjmed.2007.01.024

LEARNING POINTS

- In the acute setting it can be difficult to be certain of the cause of a new valvular lesion and often one has to cover for infective endocarditis by starting empirical antibiotics, to prevent irreversible valvular damage.
- Antiphospholipid syndrome treatment remains a challenge both in terms of timely diagnosis and management.
- Consideration of 'probable antiphospholipid syndrome' with the redefining of antiphospholipid syndrome criteria to include important outcomes such as valvular lesions and antiphospholipid syndrome nephropathy is warranted to ensure early diagnosis and prevent long-term sequelae.

Ruiz-Irastorza G, Crowther M, Branch W, Khamashta MA (2010) Antiphospholipid syndrome. *Lancet* **376**(9751): 1498–1509 https://doi.org/10.1016/S0140-6736(10)60709-X

Supraclavicular aneurysm as a presentation of alpha-1 antitrypsin deficiency

A 50-year-old woman presented to the outpatient clinic because of the appearance of a supraclavicular lump. She had experienced several months of weight loss and difficulties swallowing solids. She was on atorvastatin and clopidogrel because of an unexplained ischaemic cerebral stroke. Apart from a deep non-pulsatile tumour which was adherent on examination, physical examination, gastroscopy and abdominal ultrasound scan were normal. A computed tomography scan showed a 4 cm diameter ovoid mass, closely related to the right subclavian artery. Angiography of the aortic and supra-aortic branches was both diagnostic and therapeutic (*Figure 1*).

Apart from a mild cholestatic pattern (bilirubin 1.73 mg/dl and gamma-glutamyl transferase 69 U/litre), blood cell count, proteinogramme, tumour markers, anti-nuclear antibodies, anti-neutrophil cytoplasmic antibodies and viral serologies were normal. Given the clinical scenario of cholestasis, an early acute stroke and the presence of aneurysms there was a broad differential diagnosis ranging from infections, neoplasms, genetic disorders, thrombophilias to autoimmune diseases. All investigations came back negative except alpha-1 antitrypsin which was 10 µmol/litre (normal range 20–53 µmol/litre). A ZZ genotype mutation was found in the alpha-1 antitrypsin gene and vascular phenomena related to the alpha-1 antitrypsin deficiency were diagnosed. **BJHM**

Dr Pablo Ruiz-Sada, Consultant, Internal Medicine Department, Hospital Reina Sofía de Tudela, Spain

Dr Mikel Eskalante-Boleas, Consultant, Internal Medicine Department, Hospital Clínico Universitario de Basurto, Basurto, Bilbao, Spain

Dr Iker Garay-Hidalgo, 5th year resident, Internal Medicine Department, Hospital Clínico Universitario de Basurto, Basurto, Bilbao, Spain

Dr Lara Palacios-García, 4th year resident, Internal Medicine Department, Hospital Clínico Universitario de Basurto, Basurto, Bilbao, Spain

Correspondence to: Dr P Ruiz-Sada (pablitasitas@gmail.com)

Figure 1. Angiography showing partially-thrombosed pseudoaneurysm undergoing supraselective embolization.