

## LETTERS TO THE EDITOR

### Myocardial infarction caused by an aneurysm of the left main coronary artery without evidence of Kawasaki disease

SIR,—We read the case report from Pfafferoth and coworkers<sup>1</sup> about atypical Kawasaki syndrome with great interest. The authors report the case of a 20 year old woman with acute myocardial infarction and a huge aneurysm of the left main coronary artery that was occluded by a large thrombus. Because other vascular and systemic diseases were excluded, atypical Kawasaki syndrome was diagnosed. Diagnostic criteria of Kawasaki disease include fever for five or more days unresponsive to antibiotics, bilateral congestion of the conjunctiva, peripheral limb changes including indurative oedema, and erythema followed by membranous desquamation of the fingertips, erythema of oral and pharyngeal mucosa and a strawberry tongue, a polymorphous exanthema of the trunk, and cervical lymphadenopathy. Furthermore, the diagnosis is accepted when coronary aneurysms are present in addition to four of these major symptoms.<sup>2</sup> Thrombosis of coronary aneurysms, however, often leads to myocardial infarction in this disease and therefore has major influence on long term prognosis.<sup>3</sup>

In November 1996, a 19 year old male patient was transferred from a primary care hospital to our institution. The day before he had had an acute anterior myocardial infarction. He had been treated with intravenous recombinant tissue plasminogen activator (rt-PA), and both chest pain and ST segment elevation vanished. Some hours later a new ST segment elevation in the anterior leads was diagnosed accompanied by severe chest pain. Therefore, further thrombolysis with rt-PA was given and, owing to haemodynamic instability, the patient was transferred to our institution. On the following day the patient underwent coronary angiography. It revealed a huge aneurysm of the left main artery and total occlusion of the left anterior artery. Because of major akinesia and dyskinesia of the anterior wall as well as unfavourable anatomic conditions angioplasty was not done. An intra-aortic balloon pump and mechanical ventilation because of pulmonary oedema were necessary on the subsequent eight days. The patient was weaned without complications.

In total the patient stayed 20 days in our hospital. During this time extensive examination of his medical history revealed no major illness; in particular, high fever, cervical lymphadenopathy, conjunctivitis or desquamation of the skin had not been present at any time. Furthermore, screening for laboratory signs of collagenosis was negative. Examination of peripheral vessels for aneurysmatic changes were all negative. We did find a resistance to activated protein C (APC resistance) because of mutation of factor V Leiden.<sup>4</sup> Screening of family members revealed the same genetic defect in his older



Figure 1 Anterior-posterior view of the left coronary artery with 30° caudal angulation showing huge aneurysm of the left main (LM) coronary artery, total occlusion of the left anterior descending artery (LAD), and a normal left circumflex artery. The LAD shows slight signs of reperfusion with the intra-aortic balloon pump during angiography.

brother but not his mother. His father had died some years before. Therefore, before discharge oral anticoagulation was started, and medication included a  $\beta$  blocker, an ACE inhibitor, and digoxin.

The patient was seen regularly in our outpatient department. He had no signs of congestive heart failure. Ventricular function was stable within the first 10 months. In November 1997 echocardiography showed an increase of ventricular dimensions while the patient was free from symptoms. Therefore, a diuretic was added to the therapeutic regimen.

In January 1998 the patient had ventricular fibrillation without preceding symptoms. Resuscitation was successful and he was transferred to the same primary care hospital as in 1996. Acute reinfarction with ST segment elevation in leads V5 and 6 was diagnosed and systemic thrombolysis using rt-PA was given before the patient was transferred to our institution by helicopter. Coronary angiography revealed an increase of the known left main aneurysm while the other arteries were unchanged (fig 1). The patient was haemodynamically stable but nuclear magnetic resonance imaging of the brain revealed severe hypoxic encephalopathy.

This case of a young patient with huge left main coronary aneurysm combined with APC resistance raises the question whether Pfafferoth and coworkers screened their patient for haemostatic abnormalities. We do not believe that our patient suffered from atypical Kawasaki syndrome as his medical history was negative for typical signs and symptoms of this disease. Although our case is very similar to that of Pfafferoth *et al* we think that Kawasaki disease should not be diagnosed when there is coronary aneurysm without other diagnostic criteria of this disease.

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- 1 Pfafferoth C, Wirtzfeld A, Permanetter B. Atypical Kawasaki syndrome: how many symptoms have to be present? *Heart* 1997;78:619-21.
- 2 Dajani AS, Taubert KA, Gerber MA, *et al*. Diagnosis and therapy of Kawasaki disease in children. *Circulation* 1993;87:1776-80.
- 3 Katayama F, Hiraishi S, Takeda N, *et al*. Intracoronary urokinase and post-thrombotic regimen in an infant with Kawasaki disease and acute myocardial infarction. *Heart* 1997;78:621-2.
- 4 Bertina RM, Koelman BPC, Koster R, *et al*. Mutation in blood coagulation factor V associated with resistance to activated protein C. *Nature* 1994;369:64-7.

### Inappropriate management of polycythaemia in adults with cyanotic congenital heart disease

SIR,—The editorial by Thorne<sup>1</sup> highlights misconceptions in the management of polycythaemia in adults with congenital heart disease. Inappropriate venesection is not only encountered in patients with inoperable cyanotic heart disease. Venesection and bone marrow suppression with radioactive phosphorus are also used in patients with operable congenital heart disease. Two recent cases are illustrative.

Case 1 is a 38 year old man with double outlet right ventricle, a straddling tricuspid valve, and pulmonary atresia who had a classic Blalock-Taussig shunt at age 11 years. After many years of venesection for his "inoperable congenital heart disease" he suffered a cerebrovascular accident from which he made a good recovery. Following this he received two courses of radioactive phosphorus in an attempt to suppress red cell production. After being referred to our unit he had a fenestrated Fontan operation with an increase in his exercise tolerance. He is currently awaiting transcatheter occlusion of the atrial fenestration.

Case 2 is a patient with classic tricuspid atresia, a restrictive ventricular septal defect, and pulmonary stenosis. After repeated venesections, he was referred to our unit at the age of 45 years, after he had become unwell during a venesection. He had a fenestrated Fontan operation, which was followed by transcatheter occlusion of his fenestration. Four years later he had oxygen saturations of 95% and a greatly improved exercise tolerance. (On a lighter note, his aged but sprightly mother "complains" that she is no longer able to keep up with him when walking—a reversal of the situation before surgery.)

These cases are typical of patients who have been "lost to follow up" from congenital heart disease centres and are seen only in general medical, adult cardiology, or haematology departments. Adults with congenital heart disease should be managed in conjunction with congenital heart disease units. In particular, patients labelled as "inoperable" or with "complex" congenital heart disease need to be reviewed in light of modern approaches to treatment.

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- 1 Thorne SA. Management of polycythaemia in adults with cyanotic congenital heart disease. *Heart* 1998;79:315-16.