Case reports

Isolated endocarditis of the pulmonary valve with fragmentation haemolysis

D P NAIDOO, M A SEEDAT, S VYTHILINGUM

From the Department of Medicine, University of Natal, Durban, Natal, South Africa

SUMMARY An unusual case of isolated endocarditis of the pulmonary valve complicated by fragmentation haemolysis resolved on antibiotic treatment.

Isolated infective endocarditis of the pulmonary valve is uncommon. It usually occurs with endocarditis of the tricuspid valve in heroin addicts or in patients with congenital heart disease.¹⁻³ However, in a recent report of 12 cases, only the pulmonary valve was affected in seven patients.² We report a further case of isolated infective endocarditis of the pulmonary valve. Fragmentation haemolysis was an unusual complication. It resolved during antibiotic treatment.

Case report

A 23 year old man presented with a two week history of dyspnoea on exertion and generalised swelling of the body. He had been treated at another hospital for a sore throat two weeks before. There was no family history of anaemia.

He was feverish, pale, and had slight ankle oedema. The pulse rate was 72 beats per minute and it was regular and bounding in character. The blood pressure was 130/60 mm Hg. The jugular venous pressure was elevated to 5 cm and there was hepatomegaly (3 cm). At the heart apex there was a soft, short, systolic murmur, and at the left sternal border there was a harsh, low pitched, early diastolic murmur that increased on inspiration. There were no splinter haemorrhages, clubbing of the fingernails, splenomegaly, or petechial haemorrhages. The chest was clear and the fundi were normal. The initial

assessment was aortic regurgitation with possible infective endocarditis.

Initial investigations showed: haemoglobin 71 g/l, white cell count $10.7 \times 10^9/l$, and platelet count $219 \times 10^9/l$. There was a reticulocytosis of 8.9% and the smear showed many fragmented red blood cells, spherocytes and schistocytes consistent with fragmentation haemolysis. Because of the red cell structure, the diagnosis was changed to aortic incompetence with haemolytic anaemia and the patient was investigated accordingly. The serum concentration of fibrin degradation product was $<12 \mu g/l$, the urine stain did not show haemosiderin, and the serum bilirubin concentration was $8 \mu mol/l$. The direct antiglobulin test, antinuclear factor, and serum complement were normal. There was a slight increase in erythrocyte osmotic fragility in 0.5% saline.

Repeated blood cultures during the first week showed no growth. The proportion of reticulocytes increased to 14% and the erythrocyte sedimentation rate to 95 mm/hour. Echocardiography performed one week after admission showed normal mitral and aortic valves, and large vegetations were detected on the pulmonary valve, the leaflets of which were thin and mobile (fig 1). Treatment was started with intravenous penicillin 5 megaunits six hourly and gentamicin 80 mg eight hourly. In the following week the patient's condition improved, the proportion of reticulocytes fell to 2.4% and the erythrocyte sedimentation rate to 55 mm/h. His haemoglobin concentration subsequently rose to 115 g/l. Repeat echocardiography showed vegetations still attached to the pulmonary valve. Three weeks later, while he was being treated with antibiotics, the patient became febrile and complained of right sided chest

Requests for reprints to Dr D P Naidoo, Department of Medicine, University of Natal, PO Box 17039, Congella 4013, Natal, South Africa.

Fig 1 Cross sectional short axis echocardiogram showing vegetations (arrows) attached to a pulmonary valve (PV) of normal appearance. AO, aorta; PA, pulmonary artery.

pain. A chest radiograph confirmed that there was consolidation of the right lower lobe of the lung (fig 2). The erythrocyte sedimentation rate had accelerated to 95 mm/h. Antibiotics were continued and his temperature fell in the next 24 hours. Echocardiography at this stage did not show vegetations on the pulmonary valve.

The patient refused further investigation and was discharged. At follow up three months later he was symptom free but still had the murmur of pulmonary regurgitation. His haemoglobin concentration was 133 g/l and his white cell count was $5.5 \times 10^9/l$.

Discussion

When the pulmonary valve is affected in infective endocarditis, there is usually evidence of underlying congenital heart disease,¹⁻³ such as ventricular or atrial septal defect, pulmonary stenosis, and patent ductus arteriosus. More recently it has been described in heroin addicts, and necropsy studies in these patients show that the tricuspid valve is ten times more likely to be affected than the pulmonary valve.^{4,5} Chronic alcoholism, skin disease, and lymphoma with depression of the immune response have been cited as other factors in the development of right sided endocarditis. None of these predisposing factors was present in this patient. A recent review noted that of 28 cases reported since 1960, only three patients had neither a predisposing event nor an underlying cardiac lesion.¹

It is difficult to make a clinical diagnosis of infective endocarditis of the pulmonary valve: when there is pulmonary regurgitation, this is usually misdiagnosed as aortic regurgitation, or the signs may be masked by coexistent heart disease. Furthermore, the clinical picture is often dominated by

Fig 2 Chest radiograph showing consolidation after embolisation from the pulmonary valve.

pulmonary manifestations caused by emboli from the infected valve. In the later stages, as pulmonary hypertension supervenes, the murmur is indistinguishable from aortic regurgitation.⁶ In our patient diagnosis was made more difficult because earlier treatment with antibiotics meant that blood cultures were negative. When endocarditis of the pulmonary valve is suspected in this situation, echocardiography is extremely useful. In the appropriate clinical setting, the demonstration of vegetations on the pulmonary valve confirms the diagnosis. Echocardiography should be performed early to avoid delay in starting antibiotic treatment. In this patient, echocardiography also helped to confirm that the findings on the chest radiograph three weeks after treatment were the result of pulmonary infarction caused by embolisation of vegetations from the pulmonary valve.

In our patient fragmentation haemolysis was an unusual feature which initially directed attention away from infective endocarditis. Anaemia is common in infective endocarditis, and it is usually normochromic anaemia, but hypochromic microcytic anaemia is also well recognised. Fragmentation haemolysis has been described only in rare instances, and pathogenic mechanisms include disseminated intravascular coagulopathy, thrombotic thrombocytopenic purpura, and mechanical damage to red cells by vegetations. Disseminated intravascular coagulation that resolved immediately after valve replacement was described in a patient with vegetations on the mitral and aortic valves.⁷ In another report there was fragmentation haemolysis in two patients who had signs resembling those of thrombotic thrombocytopenic purpura that remitted with

antibiotic treatment.⁸ Our patient did not have signs of a bleeding tendency and his serum concentration of fibrin degradation product was $< 12 \mu\text{g/l}$. Nor did he have thrombocytopenia, renal impairment, or neurological manifestations—features indicative of thrombotic thrombocytopenic purpura. Two possible mechanisms may explain the haematological picture in this patient. Firstly, the presence of a large vegetation occluding and narrowing the valve orifice accompanied by turbulent blood flow was a contributory factor but not the only one, and secondly haemolysis seemed to be related to the infective process itself, because it began to resolve soon after antibiotic treatment was started and while the vegetation was still attached to the pulmonary valve.

References

- 1 Cassling RS, Rogler BS, McManus BM. Isolated pulmonary valve endocarditis: a diagnostically elusive entity. *Am Heart J* 1985;109:558–67.
- 2 Cremieux A, Witchitz S, Malergue M, *et al.* Clinical and echocardiographic observations in pulmonary valve endocarditis. *Am J Cardiol* 1985;56:610–3.
- 3 Nakamura K, Satomi G, Sakai T, *et al.* Clinical and echocardiographic features of pulmonary valve endocarditis. *Circulation* 1983;67:198–204.
- 4 Ramsey RG, Gunnar RM, Tobin JR. Endocarditis in the drug addict. *Am J Cardiol* 1970;25:608–18.
- 5 Banks T, Fletcher R, Ali N. Infective endocarditis in the heroin addict. *Am J Med* 1973;55:444–51.
- 6 De Pace NL, Nestico PF, Iskandrian A, Morganroth J. Acute severe pulmonic regurgitation: pathophysiology, diagnosis and treatment. *Am Heart J* 1984;108:567–73.
- 7 Stuart JJ, Breyer R. Subacute bacterial endocarditis causing disseminated intravascular coagulation: resolution after valve replacement. *J Thorac Cardiovasc Surg* 1984;88:454–6.
- 8 Bayer AS, Theofilopoulos AN, Eisenberg R, Friedman SG, Guzol LB. Thrombotic thrombocytopenic purpura-like syndrome associated with infective endocarditis. A possible immune complex disorder. *JAMA* 1977;238:408–10.