CASE REPORT

Acute coronary syndrome in a patient with Eisenmenger's syndrome

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SUMMARY

Eisenmenger's syndrome is a complex multisystem disease which occurs as a consequence of large left to right shunt affecting the pulmonary vasculature causing suprasystemic pulmonary artery pressures and subsequent right to left shunt. Usually Eisenmenger's syndrome is characterised by coronary artery dilation but coronary artery disease and occurrence of acute coronary syndrome is a rare association seldom described in literature. We present the case of a 33-year-old man who presented with an anterior wall ST elevation myocardial infarction and was thrombolysed. The occurrence of acute coronary event in a case of Eisenmenger's syndrome is a rare event and needs to be discussed and kept in mind by physicians and cardiologists who deal with such cases so that a high index of suspicion may be developed and emergent management may be possible.

BACKGROUND

Patients with Eisenmenger's syndrome suffer from a complex and multisystem disorder including coagulation disorders (bleeding complications and paradoxical embolisms), renal dysfunction, hypertrophic osteoarthropathy, heart failure, reduced quality of life and premature death. Coronary artery disease is very rarely seen in patients with Eisenmenger's syndrome as the survival to the fifth decade of life is rare¹ and also the reduction in coronary blood flow due to increased haematocrit is offset by the development of coronary anastomosis which results in the enlargement of the calibre of coronary arteries. We present a rare case of coronary artery disease in a patient with Eisenmenger's syndrome. The publication of this case is important to make the physician and the cardiologist aware of this rare but potentially fatal complication of this relatively common disease the timely recognition of which can be lifesaving.

CASE PRESENTATION

A 33-year-old man presented to the intensive care unit with chest pain for 2 h. The patient was diagnosed to have acute anterior wall myocardial infarction (figure 1) and was thrombolysed with injection streptokinase (Lupisl, Lupin Ltd) 1.5 MU given over a period of 1 h.

On routine examination pulse was 100/min and blood pressure 110/80 mm Hg. The patient was cyanosed with clubbing of fingers and a saturation of peripheral oxygen of 83% at room air (figures 2 and 3). Retrospectively the patient gave a history of recurrent respiratory tract infections, and was told

to be suffering from congenital heart disease at the age of 2 years. The patient did not undergo any definite treatment and was on symptomatic management. On cardiovascular system examination the patient had a parasternal heave, loud pulmonary component of second heart sound (P2) and a short systolic murmur at the left parasternal margin.

INVESTIGATIONS

Routine investigations revealed a haemoglobin of 20.8 g/dL, packed cell volume 62.04, platelet count was in the lower range of normal that is, 1.47–2.29 lakh/mm³ at several occasions, total leucocyte count was 9600, mean corpuscular volume 91.8, mean corpuscular haemoglobin (MCH) 30.7, creatinine 0.6 mg/dL, urea 16 mg/dL, total cholesterol 159 mg/dL, triglycerides 193 mg/dL, high density lipoprotein 32.2 mg/dL, creatine kinase (CK-MB) 17 U/L, activated clotting time 70 s, bleeding time 2 min 42 s, uric acid 7.9 mg/dL, homocysteine 13.27 µmol/L, antiphospholipid IgG 0.81 U/mL and antiphospholipid IgM 1.53 U/mL. Baseline activated partial thromboplastin time (aPTT) was increased and was 60.0 s at presentation.

Two dimensional echocardiography revealed regional wall motion abnormalities in the left anterior descending artery (LAD) territory and left ventricular ejection fraction 45%, and a large perimembranous ventricular septal defect with bidirectional shunt. Bilateral lower limb venous Doppler was normal (figure 4).

The patient underwent coronary angiogram subsequently which showed proximal LAD discrete 70% stenotic lesion (figure 5).

TREATMENT

Percutaneous transluminal coronary angioplasty (PTCA) with bare metal stent (multilink vision 3.5×15 mm Abbott vascular) was performed successfully (figure 6).

OUTCOME AND FOLLOW-UP

The patient has been stable at 3 months of follow-up and is maintained on dual antiplatelet therapy.

DISCUSSION

In patients with Eisenmenger's syndrome, with regard to management issues about PTCA and follow-up some points have to be taken care of:

1. Platelet abnormalities due to thrombocytopenia and thrombosthenia.² Platelets are in the lower range of normal or reduced, due to decreased

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To cite: Prakash SS, Gupta K, Agrawal N, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201339

Figure 1 ECG at presentation.

production and survival.³ In our patient platelet count varied from 1.47–2.29 lakh/mm³. Paradoxically, bleeding time was reduced. Other tests of platelet function as whole blood impedance aggregometry was not performed due to non-availability. Aspirin was given as 300 mg loading dose and 150 mg twice daily on follow-up. Clopidogrel was given as 300 mg loading dose 12 h before procedure and then 75 mg once daily for a period of 1 month.

2. Abnormalities of coagulation factors: Production of vitamin K dependent factors (factor II, VII, IX and X) and factor V are reduced. Increased fibrinolytic activity and depletion of largest von Willebrand multimers contribute to bleeding tendency.⁴ In our case baseline aPTT was prolonged, although clotting time (6 min 15 s), and activated clotting time (70 s) were in range. During the PTCA procedure we gave 100 IU/kg body weight heparin. The patient's activated clotting time (ACT) was 195 s after giving 6000 IU heparin which was not in the target range. Since target ACT was not achieved additional 1000 U heparin was given. Final ACT was 250 s (recommended ACT >250 s). ACT was kept at the lower

side of the recommended range to balance the risk of bleeding and thrombosis. These patients not only have increased bleeding risk but also increased risk of thrombosis in the presence of foreign material. The use of anticoagulants and antiplatelet agents in cyanotic patients is controversial due to increased bleeding tendency. They should be used with careful monitoring of degree of anticoagulation.^{5 6}

Coronary artery disease with Eisenmenger's syndrome is a very rare presentation. There has been only one case report of coronary artery disease and the occurrence of an acute coronary

Figure 2 Clinical photograph of the patient showing cyanosis and clubbing. **Figure 3** Clinical photograph of the patient with a pulse oximeter showing a saturation of 83% at room oxygenation.

Figure 4 Two dimensional echocardiogram with colour flow imaging demonstrating large perimembranous ventricular septal defect with bidirectional shunt.

event in a patient with Eisenmenger's syndrome. The patient was a 48-year-old woman who underwent surgery for cerebral abscess. The patient was readmitted with inferior wall myocardial infarction and developed ventricular tachycardia and was successfully resuscitated. The patient was conservatively managed and later discharged.¹ In another case report the patient presented with acute myocardial infarction owing to the compression of the left main coronary artery by distended pulmonary artery.⁷ Ours is the second case report of coronary artery disease with Eisenmenger's syndrome, and in the authors' knowledge, the first case to undergo successful thrombolysis and percutaneous coronary intervention for acute myocardial infarction.

The postprocedure and postacute coronary syndrome follow-up of patients presenting with Eisenmenger's syndrome is controversial and given the procoagulant tendency of the patient due to increased blood viscosity and at the same time the increased bleeding tendency due to deficiency of clotting factors and platelet dysfunction. As of now there are no recommendations about antiplatelet therapy in these patients after acute coronary syndrome and after stent implantation.

Ideally the patients should be maintained on dual antiplatelet therapy and the therapy should be individualised based on bleeding and clotting parameters and the subsequent response

to therapy. The duration of dual antiplatelet therapy is again controversial and there are no standard practice guidelines but preferably the duration of dual antiplatelet should also be individualised according to laboratory and clinical parameters.

The indication of anticoagulation in these patients should be based on laboratory and clinical parameters and should best be avoided in patients already on dual antiplatelet therapy. As per the guidelines, in order to reduce the risk of bleeding we should limit the use of anticoagulation only to indications like atrial fibrillation, recurrent thromboembolic events, mechanical heart valve prostheses while maintaining a meticulous surveillance of anticoagulation. The standard recommendation for target international normalised ratio is between 2.0 and 2.5 (in the absence of a mechanical valve) or to maintain a therapeutic aPTT of 1.5 times of the normal value in patients who are strongly indicated for anticoagulation therapy. Another useful aspect is the prompt treatment of respiratory tract infections with a view to avoiding haemoptysis in anticoagulated patients.

The recommended strategy to reduce the risk of ischaemic events include the avoidance of dehydration, Iron supplementation in patients with iron deficiency or in patients requiring repeated phlebotomies and the use of air filters in intravenous lines and precaution of air embolisation during percutaneous procedures and line insertion.

Figure 5 Angiographic view of coronary arteries showing proximal stenosis of the left anterior descending artery.

Figure 6 Angiographic view of the left anterior descending artery after percutaneous coronary intervention.

Learning points

- ▶ Patients with Eisenmenger's syndrome presenting as acute myocardial infarction is very rare. Ours is the first case which highlights the management issues involved in patients presenting with acute coronary syndrome.
- ▶ Issues regarding antiplatelet agents and anticoagulation are of particular concern as these patients suffer from coagulation and thrombotic abnormalities.
- ▶ There is a paucity of literature and standard recommendations regarding the management of acute myocardial infarction in the setting of Eisenmenger's syndrome.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Passarani S, vignati G, Einaudi A. Eisenmenger syndrome: a case of survival after ventricular tachycardia due to inferior myocardial infarction in a 48-year-old patient with a congenital large ventricular septal defect. *Can J Cardiol* 2004;20:822–4.
- 2 Oechslin E, Mebus S, Schulze-Neick I, *et al*. The adult patient with Eisenmenger syndrome: a medical update after Dana Point Part III: specific management and surgical aspects. *Curr Cardiol Rev* 2010;6:363–72.
- 3 Waldman JD, Czapek EE, Paul MH, *et al*. Shortened platelet survival in cyanotic heart disease. *J Pediatr* 1975;87:77–9.
- 4 Henriksson P, Varendh G, Lundstrom NR. Haemostatic defects in cyanotic congenital heart disease. *Br Heart J* 1979;41:23–7.
- 5 Daliento L, Somerville J, Presbitero P, *et al*. Eisenmenger syndrome. Factors relating to deterioration and death. *Eur Heart J* 1998;19:1845–55.
- 6 Warnes CA, Williams RG, Bashore TM, *et al*. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). *J Am Coll Cardiol* 2008;52:e1–121.
- 7 Koppa T, Mehili J, Hager A, *et al*. Left main coronary artery compression in a young woman with Eisenmenger syndrome. *Heart Asia* 2011;3:13–15.

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