Thorax, 1979, 34, 686–687

Massive intravascular haemolysis due to mitral Björk-Shiley paraprosthetic regurgitation

AJAIB S SOORAE

From the Cardio-Thoracic Unit, Walsgrave Hospital, Coventry CV2 2DX, UK

A mild degree of haemolysis after insertion of intra-cardiac prostheses and other foreign material is not uncommon, but massive, life-threatening haemolysis necessitating reoperation is, fortunately, rare.

Case report

A 68-year-old man, who had undergone closed valvotomy for mitral stenosis in 1966, was admitted with restenosis of the mitral valve, which was confirmed by cardiac catheterisation.

On 15 December 1975 a densely calcified mitral valve was excised and replaced by a 27 mm Björk-Shiley prosthesis, which was fixed in place with interrupted Ethiflex sutures. The calcification extended into the mitral ring near the lateral commissure, and the sutures in this area were reinforced by three rectangular pledgets of Teflon. Immediately after the operation the patient passed adequate amounts of clear urine, but 10 hours later he produced dark burgundy-coloured urine. On the next day he was jaundiced, and a mitral systolic murmur could be heard.

Investigations showed a severe haemolytic anaemia, with absent serum haptoglobins, persistently raised plasma haemoglobin, which was lactic dehydrogenase levels above 1400 units/l, methaemalbumin in the plasma, and continuous haemoglobinuria. The haemoglobin level fell at the rate of about 1·5 g/dl a day. A peripheral blood smear showed fragmented red cells and a reticulocyte count varying from 12 to 18%. The serum bilirubin reached a peak of 98·5 mmol/l, the blood urea 30 mmol/l, and the serum creatinine 250 mmol/l. Coombs's test and repeated blood cultures were negative.

Forty-six units of packed cells were infused intravenously over a period of seven weeks to maintain a satisfactory haematocrit. Ventriculography showed a contractile left ventricle and paraprosthetic regurgitation into an enlarged left atrium.

On 9 February 1976 the mitral prosthesis was explored. Two small defects about 2×3 mm and a larger one about 3×7 mm, were found at the site of the lateral commissure, where the sutures had cut out of the calcified area. The pledgets of Teflon felt were still intact and were seen to be arching over the paraprosthetic defects in the path of the regurgitant jets. The sutures and the Teflon pledgets were removed and the defects closed by mattress sutures, taking a part of the supra-annular portion of the left atrium in each bite.

After operation the urine was clear, and there was no clinical or biochemical evidence of haemolysis. The patient was discharged after four weeks. Two and a half years later he was asymptomatic. He had normal prosthetic sounds, no murmurs, and a stable haematocrit.

Discussion

Haemolytic anaemia of cardiac origin is generally believed to be due to mechanical trauma to the red blood cells caused by turbulence, resulting in shearing forces sufficient to fragment the cells (Rodgers and Sabiston, 1969). Therefore, any condition that results in increased turbulence, such as increased cardiac output, ball variance, high transvalvar pressure gradients, and paraprosthetic regurgitation, may give rise to haemolysis, and hence anaemia.

The two main factors responsible for haemolysis in the reported case are believed to be (1) the turbulence caused by forceful contact of red cells with the calcium-lined rough and hard surfaces of the narrow paraprosthetic defects and (2) the impact of the jets of blood at high velocity on the pledgets of bare Teflon, which were lying in the path of the regurgitant jets. Severe haemolytic anaemia, due to a similar set of circumstances to this case, was first reported by Sayed et al., in 1961. Their patient developed anaemia, after repair of an ostium primum defect with a Teflon patch, and a regurgitant jet through the mitral cleft striking the unepithelialised patch. This was successfully treated by covering the bare area with endocardium. Since then, nine other cases of haemolysis of a similar type have been reported (Hines et al., 1978).

The main diagnostic features of severe mechanical haemolysis are considerably raised serum concentrations of lactic dehydrogenase, anhaptoglobinaemia, haemoglobinemia, methaemalbuminaemia, haemoglobinuria, haemosiderinuria, and bilirubinaemia. In addition to these a rapidly falling haematocrit, reticulocytosis, and red cell fragments in the peripheral blood smear are seen. Red cell survival studies have shown a shortened survival in these cases, and the Coombs's test is negative.

Under favourable circumstances, spontaneous remission of anaemia may occur as a result of endo-
Massive intravascular haemolysis due to mitral Björk-Shiley paraprosthetic regurgitation

The lialisation of the rough surfaces. Medical treatment in the form of oral iron treatment, folic acid, and limitation of physical activity is effective in most patients with mild haemolytic anaemia. Treatment with steroids has been shown to be of no benefit. Less often, a blood transfusion may be necessary to correct refractory anaemia. In our case the haemolysis continued relentlessly until repair of the paraprosthetic leak, after which it ceased as dramatically as it had started. It is concluded that all cases of severe, persistent haemolysis from prosthetic dysfunction who do not show signs of spontaneous regression, or are unresponsive to adequate medical treatment, should have an early assessment by cardiac catheterisation and angiocardiography followed by exploration of the prosthesis and surgical repair of the intracardiac abnormality.

I wish to thank Mr W G Williams for allowing me to report this case.

References


Requests for reprints to: A S Soorae, FRCS, Thoracic Surgical Unit, Fazakerley Hospital, Longmoor Lane, Liverpool L9 7AL.
Massive intravascular haemolysis due to mitral Björk-Shiley paraprosthetic regurgitation.

A S Soorae

Thorax 1979 34: 686-687
doi: 10.1136/thx.34.5.686

Updated information and services can be found at:
http://thorax.bmj.com/content/34/5/686.citation

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/