Haemolytic-Uraemic Syndrome in an Adult with Pericarditis and Pleurisy

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The haemolytic-uraemic syndrome was first described in children, and since then numerous cases have been reported. Brain reviewed the subject in 1969. There have been few reports of adult cases (Dunea *et al.*, 1964; Waddell and Matz, 1966; Clarkson *et al.*, 1970; Giromini *et al.*, 1972). The usual manifestations are renal failure and intravascular haemolysis, and, less commonly, neurological disturbances and liver damage. There are a few reports of cardiac and pulmonary involvement, mostly appearing to be due to complications of the syndrome—for example, uraemia or infection—than to microangiopathy. We present a further case in an adult, who developed pericarditis and pleurisy as a primary manifestation of the haemolytic-uraemic syndrome and also showed a very favourable response to a new form of therapy.

Case Report

A 38-year-old woman presenting with oliguria, generalized myoclonic twitching, and grand mal convulsions was found to have the haemolytic-uraemic syndrome—haemoglobin 9.3 g/100 ml; reticulocytes 10%; platelets 110,000/mm³; blood film: anisocytosis, poikilocytosis, spherocytes, numerous fragmented red cells; fibrin degradation products (F.D.P.) 640–1,280 µg/ml (normal <10 µg/ml); haptoglobins absent; blood urea 255 mg/100 ml, creatinine 11.1 mg/100 ml. Haemodialysis was begun and she was treated with streptokinase 750,000 U eight-hourly for 10 days, followed by heparin 40,000 U daily. After 12 days' anuria and four haemodialyses she had a diuresis and her blood urea fell slowly to normal. Fibrin degradation products fell to 10–40 µg/ml and haptoglobins rose to 402 mg/100 ml (see fig.).

One month after admission, while still on heparin, she developed a loud pericardial friction rub and signs of cardiac tamponade. L.E. cells were absent. Fibrin degradation products rose to 2,560–5,120 µg/ml, a higher level than at any time in her illness, and haptoglobins fell to 80 mg/100 ml. Signs of tamponade increased, necessitating aspiration, and 300 ml of heavily blood-stained fluid was removed. No organism could be isolated from this fluid or from throat swabs and faeces collected at the same time. Paired sera showed no change in viral antibody titres. Over the next few days a large left haemothorax developed, from which 900 ml of blood was removed, and a loud pleural friction rub was heard over the whole right chest. Because she had relapsed while receiving full doses of heparin treatment was changed to aspirin 3 g daily and dipyridamole (Persantin) 300 mg daily, since this combination has been found to reduce platelet adhesiveness and aggregation and had been effective in a similar clinical situation (Giromini *et al.*, 1972). There was no reaccumulation of pericardial or pleural fluid and she was well enough to be discharged.

All treatment was stopped after discharge and she remained well, with normal renal function and no sign of red cell destruction, six months after the beginning of her illness. While receiving dipyridamole she complained of alopecia; this may have been a side effect of the drug, since after its withdrawal her hair grew.

Comment

We believe that the haemolytic-uraemic syndrome was the underlying cause of this patient's pericarditis and pleurisy. Friction rubs developed when the blood urea was normal and there was no evidence of collagen disease, infection, or neoplasia. We could find no reports of fibrin degradation product levels in

Blood values and urine excretion during treatment.

patients with pericarditis or pleurisy, but we found no rise in six of our patients with pericarditis from other causes.

Scheer and Jones (1968) described a 24-year-old woman with pericarditis, but its relation to the blood urea is uncertain. Ray *et al.* (1971) described two children with the haemolytic-uraemic syndrome and myocarditis associated with Coxsackie virus infection, one having 20 ml of blood-stained fluid aspirated from the pericardium. Although electrocardiograms were consistent with pericarditis there was no record of friction rub. Gianantonio *et al.* (1964), in their review of 58 cases of haemolytic-uraemic syndrome in children, found five with electrocardiographic abnormalities, but none showed clinical signs of cardiac involvement. Hence cardiac and pulmonary changes in the haemolytic-uraemic syndrome are usually due to complicating factors such as uraemia and septicæmia. No cases with prominent friction rubs due to the microangiopathy have previously been described.

Treatment remains a problem. Heparin has not been uniformly successful (Brain *et al.*, 1968), and our patient was started on streptokinase in the hope that stimulating fibrinolysis would help to recanalize thrombosed vessels (Rosen *et al.*, 1970; Bergstein *et al.*, 1972). After 10 days this was changed to heparin in the hope of preventing further intravascular coagulation, but the failure of this treatment was shown by the sharp rise in fibrin degradation products and the occurrence of fresh clinical features. The subsequent satisfactory response to aspirin and dipyridamole confirms the experience of Giromini *et al.* (1972), and we feel that this combination should be considered in the treatment of future cases, possibly after initial streptokinase therapy.

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