

the morphological changes of intrauterine death do not occur in this placenta. It may be that at about the time of death of the donor twin the cardiac output and blood pressure fall, resulting in the reversal of the shunting of blood, which then flows back from the polycythaemic infant to the donor.

In liveborn cases Klebe and Ingomar⁸ suggest that the second twin receives a larger placento-fetal transfusion than normal at the time of birth because blood from both parts of the placenta is transfused into the baby, particularly if there is a delay in clamping the cord. This would explain our findings in Cases 2 and 3 (Table 2). It seems that FFTS accounts partly for the greater fetal mortality and morbidity seen in monochorial as opposed to dichorial twin gestation.

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Valvulitis—bacterial or rheumatic?

K W MOLES, P MORTON, AND F McKEOWN

Cardiac Unit, Belfast City Hospital, and Department of Pathology, Queens University of Belfast

SUMMARY An 11-year-old girl presented with pyrexia, severe mitral regurgitation, and cardiac failure. The child's condition deteriorated necessitating an emergency life-saving valve replacement. Although the revised Jones's criteria were not fulfilled, histology confirmed acute rheumatic carditis.

Most observers believe that acute rheumatic fever is on the decline in the West. In consequence the classical picture of acute rheumatic fever with carditis is now rare. Acute rheumatic heart disease none the less is not extinct and when it does occur it may be atypical in its clinical manifestations. We report here the case of an 11-year-old girl, mistakenly believed to have subacute bacterial endocarditis.

Case report

The patient, who had previously been well, presented with a 3-week history of lethargy, loss of appetite, and pharyngitis. There was some myalgia and vague complaints of joint pain, but there was no swelling or tenderness and at no time was a rash seen. One week before admission she developed a pyrexia of 39.4°C with rigors, and was treated with erythromycin and ampicillin. Despite this her condition continued

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Correspondence to Dr K M Goel, Royal Hospital for Sick Children, Yorkhill, Glasgow G3 8SJ.

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to deteriorate with persistent pyrexia and the development of dyspnoea.

On admission she was pyrexial and dyspnoeic but no abnormality was found in the respiratory system. Examination of the cardiovascular system showed a sinus tachycardia, systolic thrill, and a pansystolic murmur maximal at the apex. There was no increase in the jugular venous pressure but the liver was enlarged 3 cm and was tender. The girl was pale but there was no lymphadenopathy or splenomegaly, and subcutaneous nodules could not be felt. There were no rashes or splinter haemorrhages. Joints were not swollen or tender, and the throat appeared normal. A working diagnosis of severe mitral regurgitation due to acute rheumatic fever, subacute bacterial endocarditis, or possibly a juvenile arthropathy with valvular involvement, was established.

Initial investigations revealed an erythrocyte sedimentation rate (ESR) of 150 mm in the 1st hour with a haemoglobin concentration of 9.8 g/dl and a white cell count of $11.1 \times 10^9/l$. Throat swabs and blood cultures were negative as was urine analysis. The autoantibody screen and RA latex tests were both negative. Antistreptolysin O titres on admission and 2 weeks later showed no increase. Chest x-ray film showed slight cardiomegaly but no pulmonary

congestion. The electrocardiogram was normal; in particular it showed no changes to suggest carditis.

The patient was presumptively treated as subacute bacterial endocarditis with intravenous antibiotics during a period of 2 weeks, but despite this her condition continued to deteriorate with increasing cardiomegaly. The mitral systolic murmur increased and a new aortic diastolic murmur was heard. Despite digitalisation, diuretic therapy, fluid restriction, and subsequently vasodilators, heart failure became more apparent with pulmonary congestion and a right pleural effusion. Two-dimensional echocardiography at this time showed dilated chambers with a flailing mitral valve and disrupted papillary muscles.

Changes in antibiotic therapy afforded no improvement and it was obvious that surgery was urgently required to correct the valvular defect. Cardiac catheterisation confirmed gross mitral regurgitation with pulmonary hypertension and a resting V wave of 33 mmHg in the wedge pressure. There was also mild aortic regurgitation.

Emergency mitral valve replacement, using a St Jude's prosthesis, was carried out with an immediate and pronounced improvement in the haemodynamic status. Signs of heart failure rapidly resolved and the heart size returned to normal. The

fluctuating temperature settled immediately and the ESR became normal within 2 weeks.

At surgery the mitral valve looked acutely inflamed with thickened cusps and shortened chordae suggesting acute rheumatic valvulitis. Histologically the valve cusps were oedematous and diffusely infiltrated by inflammatory cells, particularly lymphocytes and histiocytes with a few polymorphs. There was a pronounced degree of vascularisation of the leaflets and many of the vessels had smooth muscle in their walls. Aschoff's nodules were also present in the valve substance, showing central fibrinoid degeneration of collagen and peripheral palisading of histiocytes, some of which were bi- and multinucleated. Verrucae composed of platelet thrombi were noted on the line of closure of the cusps, and in other areas a thin layer of thrombus replaced the valvular endothelium and extended over the surface of the chordae tendineae. There were numerous Aschoff's nodules in the endocardium of the adjoining papillary muscle but no myocardial lesions could be observed. No organisms were demonstrated with special stains (Figure).

At this stage a third estimation of an antistreptolysin O titre, some 8 weeks after the onset of symptoms, was raised. The child was treated with prophylactic penicillin and aspirin; her subsequent

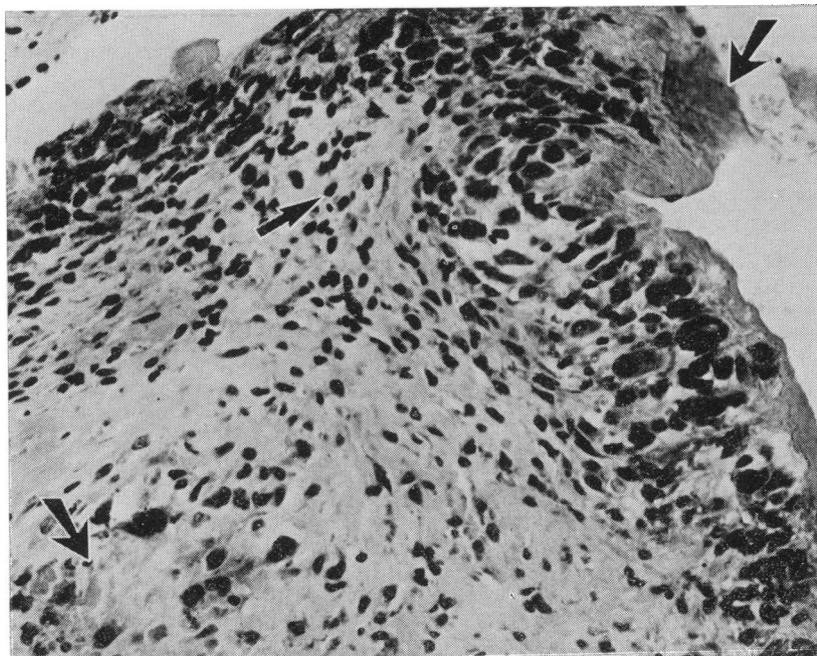


Figure Platelet thrombus deposition on valve surface (upper arrow), deep to which are Aschoff-type cells and one discrete Aschoff's nodule in valve substance (lower arrow). (HE \times 150).

progress was excellent and she remains well one year later.

Discussion

Our case illustrates the persistence of severe rheumatic carditis and emphasises a number of points in the diagnosis and management of a febrile child with valvulitis and significant haemodynamic disturbance.

The differential diagnosis between the two most likely aetiological factors was made difficult by the previous administration of antibiotics and by the absence of laboratory evidence of a recent streptococcal infection. Although our patient had one major and two minor criteria for the diagnosis of acute rheumatic fever, there was no evidence of a preceding streptococcal infection as required in the 1965 revision of the Jones's criteria,¹ until 8 weeks after the onset of symptoms.

It was clear that cardiac surgery was required but our dilemma was that we were unable to exclude subacute bacterial endocarditis and indeed, in view of the comparative rarity of acute rheumatic carditis of this severity, we felt that an infective endocarditis was the more probable cause. We therefore considered it advisable to delay surgery until we could ensure that sufficient antibiotic treatment had been administered to render this

intervention less hazardous. Although our pre-operative diagnosis was shown to be wrong, we believe that faced with the same situation we would still pursue the same course of management.

In accordance with previous reports,²⁻⁴ this case shows that surgical intervention in the acute phase of rheumatic fever is safe and beneficial; in the critically ill patient it may even be life-saving.

We thank Mr J Cleland, senior consultant cardiac surgeon, Royal Victoria Hospital, Belfast, for help in the management of this case.

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Correspondence to Dr Patricia Morton, Belfast City Hospital, Lisburn Road, Belfast BT9 7AB.

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Infantile multicystic encephalomalacia after maternal bee sting anaphylaxis during pregnancy

C ERASMUS, W BLACKWOOD, AND J WILSON

The Hospital for Sick Children, London, Queen Mary's Hospital for Children, Carshalton, Surrey, and Hurstwood Park Hospital, Haywards Heath, Sussex

SUMMARY We report a case of infantile multicystic encephalomalacia after maternal bee sting anaphylaxis in the 30th week of pregnancy. The clinical features and findings at necropsy are described, and it is suggested that these are the result of severe fetal hypoxia secondary to maternal hypotension.

In multicystic encephalomalacia, cerebral necrosis and cavitation occur after cerebral insult. The tendency for the immature brain to undergo cavitation is a function of the timing¹ and nature of the insult.¹⁻³ It is especially likely to occur between the 6th month of gestation and the early postnatal period.¹ Severe asphyxia or circulatory disturbances are of major aetiological importance.¹⁻⁴

Very few neuropathological studies have been reported in which a specific antenatal insult could be identified.^{5,6}

Case report

The pregnancy of a healthy non-consanguineous 31-year-old primigravida progressed normally, under regular obstetric supervision until the 30th week, when she was stung by a bee. She developed severe anaphylaxis and was unconscious for about 2 hours before admission to an intensive care unit. On examination there, she was drowsy and had facial oedema. Her blood pressure was 60/0 mmHg. She responded rapidly to intravenous hydrocortisone acetate and after 3 hours her blood pressure was