A case of primary cerebral venous thrombosis

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Summary

A case of primary cerebral thrombophlebitis is described in a young man, who had a thrombotic diathesis. The patient initially presented with a right phlegmasia alba dolens which was treated with anticoagulants, surgery and thrombolytic therapy. Over 2 years later he developed a further phlegmasia and what was clinically thought to be a meningitis. Despite anti-coagulation he died from a progressive cerebral venous thrombosis.

Introduction

Primary sinus thrombosis is rare and occurs mainly at the extremes of life. Predisposing factors are anaemia, increased coagulability of the blood, slowing of the blood stream and dehydration (Brain, 1969). Cerebral thrombophlebitis is most commonly seen in pregnancy and the puerperium (Carroll, Leak and Lee, 1966).

Case report

R.T., aged 23. The patient, a previously fit man, was first admitted on 14 June 1970, with a right phlegmasia alba dolens and a right basal pulmonary embolism. There was no history or evidence of trauma to the leg. A right ilio-femoral thrombectomy was performed and at operation there was a clot

extending from the right common iliac to the femoral and profunda veins.

Despite heparinization the patient developed progressive phlegmasia again after 6 days. This was treated with streptokinase and the leg was almost normal within a week. Previously the patient had had oral ulceration, but there was no other evidence to suggest Behçet's syndrome.

Laboratory investigations at this time were normal, apart from an ESR of 100 which fell to 3 over the next 6 weeks and a 10% clot lysis time (Fearnley, Balmforth and Fearnley, 1957) of over 24 hr.

At this time the patient was thought to have an inadequate endogenous fibrinolytic system.

Subsequent treatment consisted of long term warfarin therapy and Fearnley's regime (Fearnley, Chakrabarti and Evans, 1968) (phenformin S.A. 50 mg b.d. and Orabolin 4 mg b.d.). Progress was satisfactory apart from a further deep venous thrombosis in November 1971. In October 1972, warfarin was tailed off, the phenformin S.A. and Orabolin being continued.

The patient was readmitted on 18 November 1972 with pain in the right leg and fever of 2 weeks' duration. On the day before admission he had complained of nausea, headaches and lethargy, but there was no history of head injury.

FIG. 1. See 'Post-mortem findings'.

On examination he looked ill, was confused and had a temperature of 38.8°C. Recurrent right phlegmasia alba dolens was present with painful neck stiffness. There were no other focal signs in the central nervous system nor papilloedema.

Investigations

Hb 13.0 g%; white cell count 5000; blood sugar 106 mg%; lumbar puncture revealed a CSF pressure of 150 mm with free rise and fall. The fluid was turbid and analysis revealed red blood cells 4300/ml, white cell count 1000/ml—mainly polymorphs, protein 200 mg%, sugar 65 mg%, Gram stain—no definite organism seen—possibly a few Gram-positive cocci present.

A diagnosis of meningitis was made at this stage, and the patient was treated with intravenous penicillin and sulphadimidine. Heparin was given intravenously for the right phlegmasia.

Thirty-six hours later the patient suddenly deteriorated with vomiting and rapidly progressing coma with bilateral ankle clonus and extensor plantar responses. Two hours later respiratory arrest occurred which required assisted ventilation. Left carotid angiography was performed and this showed a very slow circulation. Right fronto-parietal burr-hole revealed low pressure in the right lateral ventricle and the CSF fluid was blood-stained with 19,000 red blood cells and 60 white blood cells/ml, protein 77 mg%, and normal sugar content. Six hours later the patient died.

Post-mortem findings

The brain. (Fig. 1). There was left temporo-occipital thrombophlebitis of the large superficial vein with adjacent haemorrhagic cortical infarction. Antemortem thrombus was present in the lateral sinus.

Thrombus was present in the common and external iliac veins but there was no evidence of fresh pulmonary embolization.

Comment

There is little doubt that this patient had a thrombotic diathesis. He had three separate episodes of spontaneous venous thrombosis and an abnormal endogenous fibrinolysis as measured by the 10% clot lysis time. The venous thrombosis and oral ulceration may have been manifestations of Behçet's disease, but other features were absent. The diagnosis of cortical vein thrombosis was only made post mortem and, initially on his final admission in November 1972, the patient was thought to have a bacterial meningitis. However, there was no evidence of

meningitis or a focus of infection at post-mortem examination.

Carroll *et al.* (1966) found that 14% of patients with cortical vein thrombosis had a raised white cell count and 24% had a raised red cell count in the CSF. Forty per cent of the cases they reviewed had a raised CSF protein. The CSF pressure may be raised and the fluid may be turbid or heavily blood stained. Electroencephalography may show a generalized abnormality and carotid angiography may show an abnormality in the venous phase. The mortality of cerebral venous thrombosis associated with pregnancy and the puerperium is 33%, and 19% of the survivors have persistent neurological deficits.

Treatment at present is undecided and the value of anticoagulants is not proven in cases associated with pregnancy. In cerebral venous thrombosis associated with the contraceptive pill good results have been obtained with anticoagulants (Fairburn, 1973). In the above case anticoagulants were indicated in view of the patient's thrombotic diathesis. The development of a recurrent right phlegmasia and cortical thrombophlebitis in November 1972 were probably precipitated by the withdrawal of anticoagulants in October 1972. Heparin was given in the final illness and was of no avail and may have contributed to the haemorrhagic cortical infarction. The use of fibrinolytic therapy for the recurrent phlegmasia in November 1972 was considered but this was thought inadvisable in view of the provisional diagnosis of bacterial meningitis. Possibly streptokinase may have prevented the fatal outcome if it had been given early enough in the terminal illness.

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