BEHCET'S SYNDROME WITH SAGITAL SINUS AND INFERIOR VENA CAVAL THROMBOPHLEBITIS

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SUMMARY

This report describes a case of Behcet's syndrome in which there was involvement of both the intracranial and systemic veins. The pathogenesis of Behcet's syndrome is discussed.

INTRODUCTION

Behcet's syndrome was originally described as consisting of a triad of recurrent aphthous stomatitis, genital ulceration and hypopyon iritis. Behcet's syndrome is now known to have a wide spectrum of other manifestations including synovitis, [1] cutaneous vasculitis, [2] colitis, [3,4] thrombophlebitis, [5] large artery aneurysm [6] and neurological complications [7] comprising of a brain stem syndrome, a meningomyelitic syndrome and an organic confusional syndrome.

Venous involvement is a recognised feature of Behcet's syndrome and has been reported as high as 44 percent and to be higher than the ophthalmic manifestation. [5] Isolated involvement of the intracranial sinuses [8] and vena cavae have been reported but I have been unable to find any other report where both the superior longitudinal sinus and inferior vena cava were involved in the same patient.

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CASE REPORT

A 20 year old female presented with a two weeks history of labial and mouth ulceration. She had a past history of recurrent mouth ulceration. Subsequently she developed occipital headache without associated nausea, vomiting or blurring of vision.

On examination there were ulcers in the labia and in the mouth. There was neck stiffness and bilateral papilloedema but no other neurological signs.

Her haemoglobin was 14g/dl, E.S.R. 53 mm/hr, W.B.C. 6.5 x 10^9/1 with neutrophils of 77%. Skull X-ray was normal. Technetium 99 scan and bilateral carotid angiography demonstrated a partial thrombosis of the posterior part of the longitudinal sinus (Fig. 1).

The patient improved spontaneously and no specific therapy was instituted at this stage. One month later she developed headache and vomiting and was again found to have bilateral papilloedema. She was commenced on warfarin and A.C.T.H. for the superior longitudinal sinus thrombosis. She remained asymptomatic for seven months, but then developed ankle oedema, ascites, distended abdominal veins and hepatosplenomegaly. Inferior venocavogram showed occlusion of the inferior vena cava above the level of the renal veins (Fig. 2).

DISCUSSION

The pathogenesis of venous thrombosis in Behcet's syndrome is still uncertain. The concept of

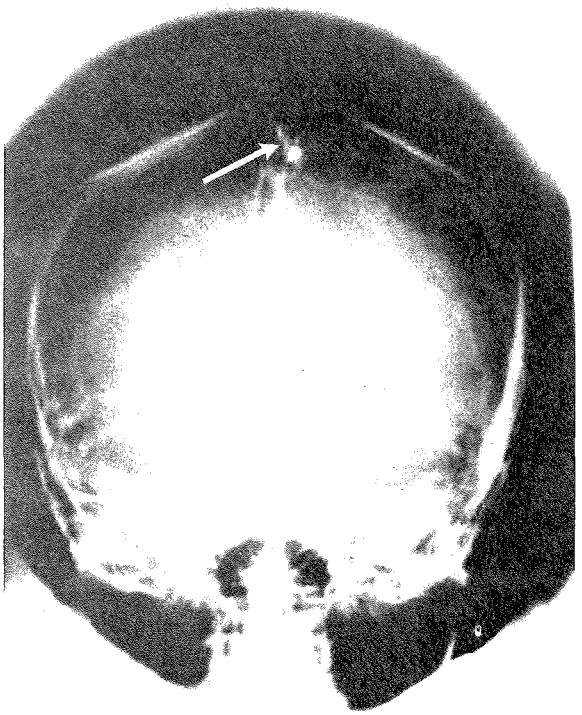

Fig. 1 Carotid angiogram showing partial thrombosis of the posterior part of the longitudinal sinus.

hypercoagulable state is not tenable as there is no abnormality of the haemostatic mechanism. Histological evidence of inflammatory changes in the veins tend to support the concept that thrombosis formation might be due to changes in the venous endothelium. Although increased fibrinogen is usually found in Behcet's syndrome, this is not thought to be the cause of thrombosis. A decreased blood fibrinolytic activity has been reported and it has been suggested that fibrinolytic enhancement therapy might play a role in the control of the vascular manifestation of the disease.

Humoral and cell-mediated mechanisms have been implicated in the pathogenesis of Behcet's syndrome. Immunopathological studies have shown that a vasculitis is the essential lesion in Behcet's syndrome and this might be secondary to immune complexes inducing complement activation and damage. [9]

Recent studies of HLA antigens in patients with Behcet's syndrome suggest that the antigen frequency of HLA-B5 is significantly raised in Behcet's syndrome patients in Japan but not in U.S.A or Britain. [9]

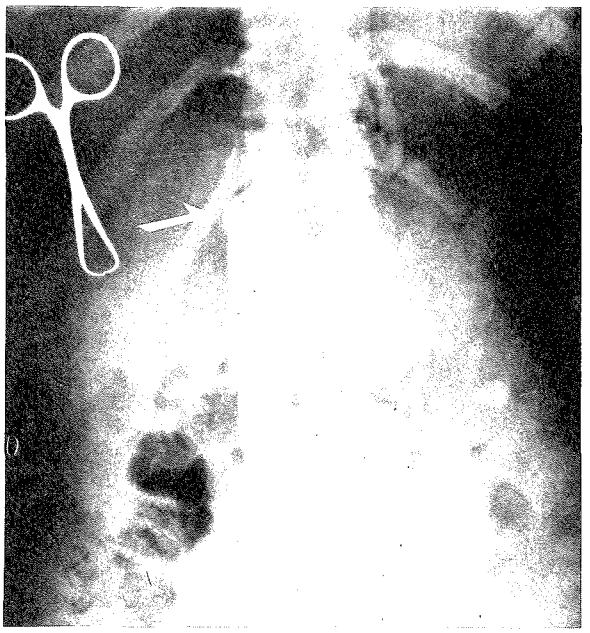

Fig. 2 Venogram showing occlusion of the inferior vena cava.

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