Behçet's disease presenting as benign intracranial hypertension

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Summary: Two siblings with Behçet's disease presenting as benign intracranial hypertension are reported and the literature is reviewed. We suggest that raised intracranial pressure should be added to the clinical criteria considered for the diagnosis of Behçet's disease.

Introduction

Benign intracranial hypertension is a diagnosis by exclusion; intracranial pressure is raised in the presence of normal size ventricles and in the absence of a space occupying lesion. In most cases the cause is unknown. Behçet's disease classically comprises a triad of oral, genital and eye lesions but since Behçet's description in 1937 a number of other clinical features, including neurological involvement, have been added. The diagnosis depends upon satisfying major and minor clinical criteria; however, since all the manifestations may not occur simultaneously, the diagnosis may be overlooked. The recognized neurological presentations of Behçet's disease are an acute confusional state, meningoencephalitis, brain stem lesions and transverse myelitis (Lehner & Barnes, 1979; Kocen & Mathews, 1983). We describe the case histories of two patients, a brother and sister, with Behçet's disease who presented with benign intracranial hypertension and suggest that the association is not uncommon.

Case reports

Case 1

A 23 year old caucasian presented in May 1973 with a 3 day history of headache, nausea and blurred vision affecting his right eye. He had been blind in his left eye from the age of 3 following a penetrating injury. Examination revealed right sided papilloedema and left sided optic atrophy. There were no focal neurological signs. Cerebral angiography excluded an intracranial space-occupying lesion. At the time of lumbar puncture, on the seventh day of admission, he had begun to improve: the spinal fluid (CSF) pressure was not raised; microbiological and biochemical

analysis of the fluid was normal. His other investigations showed haemoglobin 15.3 g/dl, erythrocyte sedimentation rate 33 mm in the first hour and normal serum biochemistry. A diagnosis of papillitis secondary to demyelination was made and he received a short course of ACTH (40 units/d for 4 weeks) as an outpatient.

In December 1973 he presented with oral and scrotal ulceration. A diagnosis of Behçet's disease was made and he was treated with chlorambucil 5 mg/d as the sole treatment. This was continued until 1981 when, in view of his maintained remission, it was electively stopped. He has since remained well.

Case 2

A sister of Case 1 presented in June 1984 aged 32 years with severe headache, vomiting and blurred vision. She had suffered intermittent headaches for 3 months and during this time had been troubled by a rash affecting her lower legs consistent with erythema nodosum. On examination she was pyrexial (38°C), had bilateral papilloedema and equivocal left sided weakness. There was no neck stiffness. A computed tomographic (CT) scan excluded a space occupying lesion and demonstrated normal size ventricles. The CSF pressure was raised at 320 mm of water; there were no cells and the glucose and protein concentrations were normal. Her other investigations showed haemoglobin 15.9 g/dl, erythrocyte sedimentation rate 36 mm in the first hour and normal serum biochemistry. Twenty ml of CSF were removed on two occasions and her condition improved.

Closer questioning revealed a 2 year history of recurrent oral aphthous ulceration and on two occasions she had received treatment for genital ulcers. The history of orogenital ulceration and erythema nodosum with a positive family history satisfied the criteria for Behçet's disease.

She was started on prednisolone 45 mg/d and the

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dose was gradually reduced over the next 8 months. She was readmitted in February 1985 on prednisolone 10 mg/day with a recurrence of her neurological symptoms. The CSF pressure was again raised (250 mm water) and she improved when the pressure was reduced by CSF withdrawal (20 ml on two occasions). She is currently taking prednisolone 10 mg and chlorambucil 5 mg/day and is well.

Discussion

Both patients presented with headache and papilloedema. The CSF pressure of the first patient was not raised at the time of lumbar puncture but this investigation was delayed to exclude a space-occupying lesion, by which time the patient had begun to improve. We feel that intracranial hypertension was the most likely cause of his presenting symptoms, which closely resembled those of his sister in whom raised intracranial pressure was clearly demonstrated. The classical features of Behçet's disease developed after the neurological symptoms of our first patient but preceded those of our second.

Neurological involvement is reported to occur in 10 to 25% of patients with Behçet's disease (Hughes & Lehner, 1979). In a large British series of 71 patients (Hughes & Lehner, 1979), 10 had definite involvement of the central nervous system but none had raised intracranial pressure. However, over the years the number of case reports of patients with Behçet's disease with clear evidence of intracranial hypertension has steadily grown.

In 1959 Masheter reported a 29 year old male with headache, papilloedema and raised CSF pressure in whom Behçet's disease had been diagnosed 1 month earlier. A review of 11 patients with Behçet's disease by Haim & Sherf (1966) includes a 28 year old female who presented with headache and vomiting due to intracranial hypertension; a diagnosis of 'pseudotumour or serous meningitis' was considered but whilst in hospital, she developed oral and genital ulceration and a diagnosis of Behçet's disease was subsequently made. In 1970 in a review of 12 patients by Kalbian & Challis, 5 had involvement of the nervous system and 3

presented with a remarkably similar clinical profile suggestive of benign intracranial hypertension. O'Duffy *et al.* (1971) described a 39 year old male who presented with an encephalopathy, papilloedema, left hemiparetic signs, recurrent oral ulceration and arthralgia, who was subjected to a craniotomy; no tumour was found and his clinical signs were attributed to Behçet's disease. The surgeon described the dura and cortex as 'unusually vascular'. More recently, other case reports have appeared (Pamir *et al.*, 1981; Rosenberger *et al.*, 1982; Bank & Weart, 1984) and have suggested that the underlying mechanism is venous sinus thrombosis. Pamir *et al.* (1981) have described 6 patients with Behçet's disease with elevated CSF pressures and normal size ventricles. 4 had angiography and in 2 of these a superior sagittal sinus thrombosis was demonstrated. Similarly, thrombosis of this sinus was seen in 2 of 5 patients with neurological involvement in a review by Rosenberger *et al.* (1982), and in a 41 year old man described by Bank & Weart (1984).

The use of the word 'benign' to describe the raised CSF pressure in Behçet's disease might be questioned but the mortality in this presentation of the disease (one patient in the series of case reports cited) is low and certainly less than that from raised intracranial pressure from cerebral tumours.

We would suggest that the association between benign intracranial hypertension and Behçet's disease has not been adequately emphasized. Patients with Behçet's disease may present with incomplete symptomatology and the diagnosis delayed or overlooked. A careful history and examination for evidence of Behçet's disease should be conducted in all patients presenting with benign intracranial hypertension as treatment directed at controlling the underlying disease may avoid the need for repeated lumbar puncture. Furthermore, benign intracranial hypertension should be added to the criteria that are considered when making the diagnosis of Behçet's disease.

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