Short report

Spontaneous intracranial hypotension with slit ventricles

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SUMMARY A case of spontaneous intracranial hypotension syndrome is described. Computed tomography showed slit ventricles with tight basal cisterns. Prompt improvement of symptoms was achieved by intramuscular dexamethasone treatment. A follow-up CT scan demonstrated re-expansion of ventricles and basal cisterns.

Headache following lumbar puncture has been well known since the introduction of the procedure, and it has been related to cerebrospinal fluid (CSF) hypotension due to CSF leakage through the needle-hole in the dura. The headache is characterised by accentuation in the erect posture and amelioration by lying flat. An identical type of headache may in rare instances emerge spontaneously. The first cases were published in German literature and the syndrome was called spontaneous/essential aliquorrhea¹⁻⁴ or spontaneous intracranial hypotension.⁵ The cause of this syndrome has remained speculative. In one recently published case examined by computed tomography (CT) the syndrome was associated with extremely small ventricles and tight basal cisterns.⁴ We report a case with slit ventricles and virtually non-existent basal cisterns during the symptomatic period with normalisation of the CT findings when the patient was symptom-free after dexamethasone treatment. The possible pathogenetic mechanisms are discussed.

Case report

On 14 March, after standing up in the morning, a previously healthy 38-year-old woman felt severe headache which was relieved by lying down. On the previous day she had felt quite fit and participated in a 10 kilometre cross-country ski-tour. Three days after onset of the headache she visited the out-patient department, and on the 4th day she was

admitted to the Department of Neurology. The posture-dependent headache which continued unchanged was associated with dizziness, nausea and malaise. Except for slight tenderness of the neck muscles the clinical examination was normal. A lumbar puncture on the 3rd day after onset resulted in a dry tap. This was the result also on the 4th day but aspiration with a syringe gave a colourless and clear sample of CSF with normal cell and protein content. Radiographs of the skull and chest were normal, as was the brain scan. The dominant activity in the EEG was 9.5 Hz alpha mixed with episodic, symmetrical and synchronous theta activity. ECG was normal, as were the blood tests (ESR, complete blood cell count, blood glucose, creatinine, alkaline phosphatase, SGOT, serum Na, K, Cl, Ca, P and Mg). The osmolalities of serum and urine were normal, as were the thyroid function test and serum cortisol. On 16 April CT examination of the head was performed (fig a, b). The lateral, 3rd and 4th ventricles were extremely narrow, slit-shaped, and the basal cisterns were virtually non-discernible. On the cortex only minimal amounts of CSF were present. The headache did not improve and on 19 April fluorocortisone was started 0.3 mg/day by mouth. After five days' unsuccessful treatment with dexamethasone 20 mg/day was instituted. In one day the patient became practically asymptomatic. The dose was gradually tapered and from 5 May the corticosteroid treatment was continued with prednisolone 10 mg/day by mouth. After one week, however, the patient displayed symptoms of a manic psychosis with paranoid features and the treatment was stopped. The psychiatric symptoms disappeared in a few days, the patient remaining free from headache. A follow-up CT on 9 July showed ventricles and basal cisterns of normal size (fig c, d). At one-year follow-up the patient was symptom-free.

Discussion

The clinical picture of spontaneous intracranial hypotension or spontaneous aliquorrhea syndrome has been well known for more than 40 years but the

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Fig: Computed tomography during the symptomatic period, showing slit ventricles and tight basal cisterns (a, b). Three months later CT showed ventricles and cisterns of normal size (c, d).

cause of this syndrome, unlike the clinically identical post-lumbar puncture headache, has been speculative.

Headache following lumbar puncture has been generally assumed to result from leakage of CSF

through the puncture-hole in the dura causing CSF hypotension and distension of the pain sensitive structures at the base of the brain. The leakage of CSF through the puncture hole in dura has been proved by isotope studies and surgical exploration.⁶

Schaltenbrand and Wolff³ have reasoned that the spontaneous aliquorrhea syndrome may result from either decreased CSF production (from an unexplained disturbance of chorioid plexus function) or enhanced CSF resorption. The latter seems improbable. Posture dependent headache following minor trauma such as a fall on the buttocks has been claimed to be due to a dural tear with subsequent leakage of CSF.⁵ A purely spontaneous tear in the dura seems unlikely because of its strong texture.

CSF hypotension has been associated with increase in brain volume⁷ and the CT findings in our case support this view. The slit-shaped ventricles, tight basal cisterns and scant CSF over the cortex fit well with diffuse brain oedema. Dexamethasone treatment resulted in rapid disappearance of headache, and the CT scan three months later was normal. The cause of the apparent brain oedema remains unknown but it might be secondary to dilatation of brain veins and downward brain sagging which have been observed during CSF hypotension.⁷ The extremely tight ventricles during the headache period of spontaneous intracranial hypotension may compromise the production of CSF by the chorioid plexuses and thus lead to a vicious circle. The exact mechanisms remain unresolved but brain swelling having perhaps some role in the pathogenesis.

In earlier reports the most promising therapeutic results were achieved by hypotonic saline iv infusions.^{1 2 4} In headache after lumbar puncture fludrocortisone has been advocated,⁸ probably be-

cause of its sodium retaining property. In our case fludrocortisone had no effect on the symptom perhaps because of increase in brain swelling. The immediate therapeutic response to dexamethasone seems logical because CT indicated the presence of diffuse brain oedema. If post-lumbar puncture headache is also associated with brain oedema, dexamethasone might be worth a therapeutic trial in that condition.

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