Short report

Dementia and hydrocephalus in Paget's disease: a case report

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SUMMARY Dementia and hydrocephalus in an elderly female were related to basilar impression caused by Paget's disease of the skull. Ventricular and lumbar thecal pressures were normal, but isotope cisternography suggested obstructive hydrocephalus. The patient improved after ventricular shunting. The importance of prompt investigation of dementia occurring in patients with Paget's disease is emphasised.

Paget's disease of the skull may lead to basilar impression,¹ and this in turn may produce a variety of neurological features.² Hydrocephalus is an important occasional sequel as it may produce a potentially reversible dementia. However the pathogenesis of hydrocephalus in Paget's disease of the skull is uncertain. We report the case of a patient with Paget's disease of the skull and normal pressure hydrocephalus who improved after ventricular shunting.

Case report

A 69-year-old woman was admitted for investigation of dementia, increasing gait disturbance and urinary incontinence over twelve months. The dementia consisted of marked short-term memory loss, progressive decrease in spontaneous activity and speech, and apathy. Though able to stand and walk unaided, her gait was broad-based and unsteady. The incontinence was mild and not especially disturbing to the patient. The head was markedly enlarged and scaphocephalic. The neck was short. The patient was inattentive and had an obvious memory loss. Her gait was broad-based, and she fell often. There was bilateral sensorineural hearing loss. The optic fundi were normal. Ocular movements were normal, and there was no abnormality of cerebellar or bulbar cranial nerve function. Muscle strength was normal, and all deep tendon reflexes were intact. Sensory examination was normal.

Skull radiographs confirmed gross Paget's disease of the vault and base. There was platybasia, the basal angle being 146°. There was also basilar impression, the tip of the

odontoid process lying 16mm above Chamberlain's line. Computed tomographic (CT) scanning disclosed considerable ventricular enlargement (fig A). Lumbar puncture with the patient in the lateral decubitus position yielded clear colourless cerebrospinal fluid (CSF) at a pressure of 70mm H₂O. Isotope cisternography was highly abnormal, revealing failure of progression of activity over the cerebral convexities, without reflux of isotope into the lateral or third ventricles. Most of the activity at twenty four hours was concentrated in the abnormally displaced basal cisterns.

A ventriculoperitoneal shunt was inserted using a right parietal burr hole. The right lateral ventricle was cannulated and the CSF pressure was recorded. The pressure varied from 30–70mm H₂O with each phase of the ventilatory cycle. A medium pressure Pudenz flushing device and a medium pressure Raimondi peritoneal catheter were inserted. The patient's postoperative course was uncomplicated and she was discharged from hospital fourteen days after operation. There was immediate and obvious improvement in her memory, interest in surroundings, gait and continence. A CT scan five days after operation showed that there had been a marked reduction in the size of the ventricles (fig B). Two months after surgery she had become active and independent, and no longer required any domiciliary supports on which she had previously been entirely dependent. Her gait was normal and she remained continent.

Discussion

The occurrence of mental symptoms and hydrocephalus in Paget's disease of the skull has long been recognised.^{2–5} However, there have been only six cases reported of the concurrence of normal pressure hydrocephalus⁶ and basilar impression due to Paget's disease.^{7–10} It has been suggested that in

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Received 8 February 1982 and in revised form 5 May 1982.
Accepted 19 May 1982

Figure (A): CT scan before operation. The lateral ventricles are enlarged. There is some cortical atrophy. (B): CT scan five days after ventriculoperitoneal shunt. The ventricles are much smaller.

basilar impression due to Paget's disease an obstruction of the basal cisterns prevents the flow of CSF over the cerebral convexities.^{4,7,9} This obstruction can be readily shown by isotope cisternography^{9,11-13} or air encephalography,⁶⁻⁸ though the latter is no longer considered necessary in diagnosis. In addition, Hens and van den Bergh have proposed that in basilar impression there may be an acquired stenosis of the Sylvian aqueduct due to a mechanical displacement of the brain stem in relation to the cerebral hemispheres.¹⁰ They reported a case of normal pressure hydrocephalus in a patient with basilar impression due to Paget's disease in whom positive contrast ventriculography revealed narrowing and deformation of the Sylvian aqueduct.

Our case differs from those so far reported. While there was a clinical triad of dementia, gait disturbance and incontinence, and while the CSF pressures were low, the isotope cisternogram pattern was that of obstructive hydrocephalus, with failure of reflux of isotope into the ventricles.¹⁴ On CT scanning, the fourth ventricle was not enlarged. It is tempting to speculate that our patient may have had an element of obstruction due to aqueduct stenosis, but ventriculography could not be justified to explore this possibility. Rather, the atypical cisternogram may serve only to highlight the fact that isotope cisternography will not always accurately delineate the pathological anatomy in patients with hydrocephalus. Further, it has been found elsewhere that isotope cisternography is not in itself predic-

tive value in selecting patients for ventricular shunting.^{11,13,15}

It is generally agreed that patients with normal pressure hydrocephalus who have an apparent or suspected cause are more likely to benefit from shunting.¹⁵⁻¹⁷ This group includes patients who have had a head injury, subarachnoid haemorrhage or meningitis, and could now be extended to include patients with basilar impression secondary to Paget's disease, as the majority of patients reported to date have improved after shunting. The treatment of choice for patients with the hydrocephalus-dementia complex and basilar impression due to Paget's disease of the skull is ventricular shunting.^{9,10} Whereas suboccipital decompression has been advocated for relieving certain cerebellar and brain stem features due to basilar impression, it is not indicated for hydrocephalus, as it can relieve neither the cisternal block nor anatomical distortion of the Sylvian aqueduct.

Patients with Paget's disease of the skull who present with dementia, gait disturbance or incontinence should be investigated, and skull radiographs and computed tomography can be recommended as a routine. Where hydrocephalus occurs secondary to basilar impression, it appears that worthwhile clinical improvement can be obtained with ventricular shunting.

We thank Dr Jack Wodak for reviewing the manuscript.

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