Hydrocephalus and Headaches in Paget's Disease of the Skull: Complete Relief by Ventriculo-Ätrial Shunt

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SUMMARY: A 76 year old patient with a long history of headaches was found to have Paget's disease and communicating hydrocephalus. There were (otherwise) no neurological or musculo-skeletal manifestations of Paget's disease, but moderate impairment of intellectual function was present. Treatment with disphosphonates did not bring any significant improvement, but three days following a ventriculo-atrial shunting procedure, the patient became headache-free for the first time in several years.

In the literature, patients with hydrocephalus have been shown to respond quite unevenly to atrio-ventricular shunting, but in most instances the descriptions concerned advanced cases with well-established symptoms of dementia, ataxia and incontinence. Our case is reported to stress the importance of early diagnosis and management of hydrocephalus in Paget's disease for the prevention of widespread neurological dysfunction.

RÉSUMÉ: Les auteurs présentent le cas d’un patient de 76 ans souffrant de céphalées sévères de longue date chez qui l’investigation a montré une maladie de Paget associée à une hydrocéphalie. Aucune manifestation neurologique ni musculo-squelettique n’a été en évidence, l’atteinte neuropsychologique était modérée. Un traitement aux disphosphonates n’a amélioré que discrètement les céphalées. Celles-ci étaient tellement importantes qu’un shunt ventriculo-atrial a été effectué. Trois jours après l’intervention, les céphalées ont complètement disparu.

Des cas d’hydrocéphalie dues à la maladie de Paget ont déjà répondu de façon plus ou moins marquée à une dérivation ventriculaire. Toutefois, tous les cas décrits étaient déjà dément, ataxiques et incontinent. Ce cas illustre donc l’importance de faire un diagnostic précoc d’hydrocéphalie chez les patients porteurs d’une maladie de Paget avant que surviennent les complications neurologiques.

CASE REPORT

A 76 year old man was admitted to hospital in February 1982. He had suffered from violent headaches for 35 years. In 1972, he saw a neurologist. At that time the physical examination was normal. An EEG showed some slow activity over the right temporal lobe. On skull X-rays, there were subtle pagetic changes in the parieto-occipital area. A diagnosis of migraine was made. Early in the history, the headaches had been intermittent, generalized and unrelated to any trigger factor, but there was throbbing in the head. The episodes would last about 6 hours and recur every fortnight, heralded by nausea. ASA would relieve the headache. The pattern had continued for about 15 years and then, the attacks had started increasing in duration, frequency and intensity.

Three months before admission to hospital the pattern of headaches had changed completely. They had become constant, localized to the occipital area and were accompanied by nausea and vomiting. Straining did not increase the pain and codeine offered no relief.

On examination, the patient was a pale, thin man of short stature, subject to the Cambridge Core terms of use, available at https://www.cambridge.org/core/terms. https://doi.org/10.1017/S0317167100045352
EEG showed non-specific slow activity with some sharp components, over the right centro-temporal area, and less frequency over the left fronto-temporal. Skull X-rays demonstrated Paget’s disease mainly in the parietal and occipital bones (fig. 1). A slight degree of basilar impression was also present. The audiogram was normal. The CT-scan showed Paget’s changes of the cranial vault and dilatation of cortical sulci (fig. 2). A technicium brain scan showed increased uptake over the vault, but no abnormal intracerebral uptake.

Figure 1 — Lateral view of the skull. Thickening of the vault with wooly appearance of the parietal and occipital bones suggesting Paget’s disease of the bone. Slight basilar impression is seen.

Figure 2 — CT-scan: Dilatation of the lateral ventricles and some degree of cortical atrophy, best shown over the fronto-temporal areas with enlargement of the cortical and inter-hemispheric sulci.

An $^{111}$Indium-DTPA isotope cisternogram revealed abnormal flow of CSF with early penetration of the ventricles, but with some stasis in the cisterna magna. More importantly there was poor resorption along the superior longitudinal sinus (fig. 3). The total body $^{32}$P bone scan showed increasing uptake only over the skull, consistent with active Paget’s disease.

Neuropsychological tests disclosed a visuoperspective and constructive disorganization with constructive apraxia and visuomotor incoordination. There were verbal and visual memory deficits. An overall psychomotor slowing was recorded but intellectual functions were preserved qualitatively: the verbal IQ was 97 and the performance IQ was 84 (table 1A).

Figure 3 — Isotope cisternogram after injection of 500 Ci Indium$^{111}$ fixed on DTPA. Lateral view at $5^{th}$ hours after injection. There is an abnormal and early penetration of the radionuclide in the ventricles. The peripheral circulation is very slow and the CSF is stagnant in the cisterna magna.

Etidronate disodium (Didronal) was given in doses of 300 mg (5mg/kg) from the first day of admission. A few days later, slight improvement was noted by the patient but he still would call for analgesics regularly. Even Meperidine did not provide any relief. A lumbar tap was done with removal of 250 cc of CSF, without any improvement.

A ventriculo-atrial shunt was established at surgery on March 1, 1982. Pressure in the occipital horn was 80 mm H$^2$O. The derivation catheter from the ventricle with a median-pressure Holter valve was connected to a right atrial catheter inserted through the left facial vein. Three days later, the patient was headache-free for the first time in 35 years. His headaches did not recur for 10 months, when he returned to report the recent onset of headaches, similar to what had existed prior to surgery. On examination, it was not possible to depress the Holter valve behind the left ear, and the isotopic flow study confirmed the malfunction. Two days following replacement of the valve, the headaches had again disappeared. Eighteen months following the first shunting operation, the patient is still symptom-free.

Two months after the first operation, neuropsychological tests had shown overall improvement of judgment, abstractive capacity, concentration and mental arithmetic (table 1B). When last seen, 18 months after the initial operation, there was further general improvement of neuropsychological activity and testing (table 1C).

DISCUSSION

This patient not only complained of headaches, but also had psychomotor retardation, impaired memory and constructive apraxia on psychological tests. His EEG was abnormal; the isotope cisternogram and CT-scan showed hydrocephalus of the communicating type.
The relationship between the long-standing headaches and the Paget's disease in this patient is at first difficult to establish, since the basilar impression on X-rays was minimal, the CSF pressure was normal and since simple hydrocephalus "ex vacuo" can occur without any direct relationship to coexisting Paget's disease, the hydrocephalus and the headaches. However, headaches are not usually associated with hydrocephalus "ex vacuo". Admittedly, from age 41 to 56, the headache pattern was typical for migraine, but there was an increase in the severity of the headaches from age 56 into advancing age. This is unusual for migraine.

Our case is the first reported pagetic patient in the literature who had complained of headaches only and who was totally relieved by the ventriculo-atrial shunt. The objective findings represented by the neuropsychologic tests in our patient have also improved since operation.

The surgical approach has been used in cases of hydrocephalus. Culebras (1974) obtained good improvement in two out of three demented and ataxic patients, following ventriculo-atrial shunting, but the improvement was of short duration. Botez in 1977 reported a 65 year old pagetic female with dementia, parkinsonian features, gait ataxia, grasp reflex of the feet, urinary incontinence and drop attacks) may occur, as well as purely mental syndromes (paranoia or manic-depressive psychosis).

Boudin (1975) has proposed a clinical classification of the cerebral complications of Paget's disease: 1) in the latent phase the patient is neurologically normal; 2) in the second phase associated with hydrocephalus, there is psychomotor retardation, impaired memory, ataxia and urinary incontinence; 3) in the phase of late decompensation, there are severe neurological manifestations and a rapid evolution with stupor, akinetic mutism and eventually coma. The third phase is not reversible as it relates to vascular compromise due to kinking in the arterial thalamo-perforantes and choroidae posterior mediales.

Headache is the most frequent neurologic complaint in pagetic patients (52%), followed by hearing loss (46%). The headaches can be of three types: 1) deep pain in the whole head, constant and unalleviated, in relation to involvement with the vault by Paget's but without involvement of the base of the skull; 2) a sharp localized pain associated with malignant degeneration of Paget's bone; 3) headaches due to the associated hydrocephalus are exacerbated by straining and by the recumbent position. With this last type of headache, there is usually a basilar impression on the skull.

Basilar impression is frequently reported in Paget's (Boudin et al., 1975; Taylor and Chakravorty, 1964; Bull et al., 1959) and it appears to be unrelated to the severity of bone involvement by the disease. Mechanical forces acting on softened bone will result in a progressive invagination of the base of the skull and this will impede the flow of CSF through the basilar cisterns, with the production of hydrocephalus.

The basilar impression will also angulate the aqueduct of Sylvius and displace it anteriorly. With further progression of the disease, the displacement of the brain stem will come to occlude the interpeduncular cistern and distort the arteries passing through the perforated substance.

Corrective treatment of the cerebral complications must be initiated before the onset of the phase of decompensation, in order to be effective. On the medical side, disphosphonates (Alexander et al., 1979), Calcitonin (Detilleux et al., 1977) Mitramycin, Actinomycin D, have been tried with mixed results in an attempt to stem the development of the bony lesions of Paget's disease.

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The interest in this case is the impressive result of the corrective procedure, strongly indicating the importance of an early search.
for hydrocephalus in patients with Paget’s disease. Even though experience in the literature to date is anecdotal we propose long-term follow-up of Paget’s disease, using neurological and neuropsychological investigations, for an early corrective treatment of deteriorations by shunting, since widespread nervous dysfunction may be irreversible when it has become clinically obvious.

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REFERENCES