Parkinsonism — Dementia Complex, Hydrocephalus and Paget's Disease

M. I. BOTEZ, GILLES BERTRAND, JEAN LÉVEILLÉ AND LUC MARCHAND

SUMMARY: *A 65-year-old female patient with Paget's disease displayed dementia, a bilateral Parkinsonian syndrome, gait ataxia, bilateral grasp reflex of the feet, urinary incontinence and a left hemicerebellar syndrome. She had a marked basilar impression; the radionuclide cisternogram was suggestive of a communicating hydrocephalus with ventricular dilatation. After a shunting procedure there was improvement in both neurological and mental function.*

RÉSUMÉ: *Les auteurs présentent l'observation clinique d'une patiente âgée de 65 ans qui avait une maladie de Paget avec démence, syndrome Parkinsonien bilatéral, ataxie de la marche, réflexe de préhension des pieds, incontinence urinaire et un hémisyndrome cérébelleux gauche. La radiographie du crâne a montré une invagination basilaire; la scintycisternographie avec 111. In a mis en évidence une hydrocéphalie communicante. On a pratiqué une dérivation ventriculo-atriale qui a amené une amélioration autant du point de vue neurologique que du point de vue psychique.*

From Hôtel-Dieu Hospital, Neurosurgical Service of Montreal Neurological Hospital and Clinical Research Institute of Montreal, Montreal, Canada.

Reprint requests to: Dr. M. I. Botez, Clinical Research Institute of Montreal, 110 Pine Avenue West, Montreal, Que., H2W 1R7 Canada.

Although shunting procedures have been recommended as treatment for the occult hydrocephalus syndrome secondary to Paget's disease of the skull (Boudin et al., 1975), there are only three reports of patients who have undergone this procedure (Culebras et al., 1974). The complex neurological and mental symptoms as well as the benefits of the procedure are the reasons for this report.

Case Report

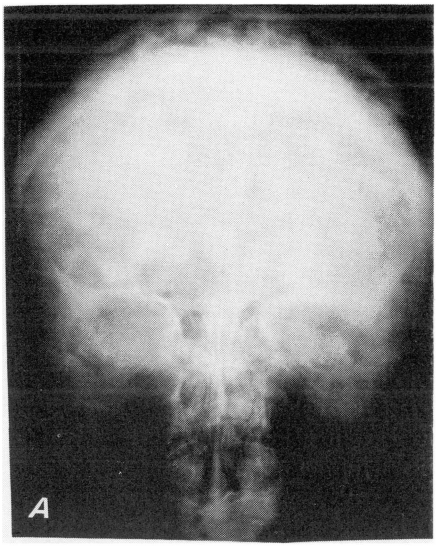
A 65-year-old right-handed woman was admitted to Hôtel-Dieu Hospital on January 15, 1975, because of a three-year history of difficulty in walking and spastic legs plus a six-month history of progressive dementia. She was incontinent and had difficulty in maintaining her balance. There was tremor, akinesia and rigidity, more marked in the upper extremities, and she had been given antiparkinsonian medication (Phenoxene [chlorphenoxamine hydrochloride]) 50 mg three times daily, (Benadryl [diphenhydramine hydrochloride]) 50 mg twice daily and (Symmetrel [amantadine hydrochloride]) 100 mg twice daily, with no improvement. She had been known to have Paget's disease for the last ten years.

General examination showed an enlarged head with frontal prominence and limited motion of the cervical spine. Blood pressure and routine laboratory tests were normal. The patient was alert, but was oriented to person only and was unable to give a coherent history. Her powers of calculation and judgement were poor She usually was silent, speaking only when spoken to after a pause. There was a definite lack of drive. The neuropsychologist tried to apply a battery of neuropsychological tests (Botez et al., 1974, 1975), but only the WAIS and Kohs Blocks Design tests could be completed because the patient was only partially cooperative and because her tremor and rigidity impaired some testing procedures. She had a verbal I.Q. of 88 and a performance I.Q. of 62 (full scale, 72). The total score on Kohs Block Design (Kohs, 1960) was 2 (maximum score 133, mean for her age 95). The neuropsychological examination was consistent with mild dementia.

There was a moderately severe spastic weakness of the lower extremities with a broad-based, ataxic and spastic shuffling gait. Tandem walking was impossible. There was generalized poverty of movement and bradykinesia with some difficulty in initiating movements. She had a 4 c.p.s. tremor in both upper limbs, more marked on the left. All extremities showed a marked "plastic" rigidity with a definite "cogwheeling" in the right arm. She had the Noica and Draganesco (1935) sign of latent muscular Parkinsonian rigidity, tested as follows: When she was in a relaxed supine position, the examiner took the patient's forearm and held it upright; he then performed complete and continuous passive flexion and extension movements of the wrist. After a few seconds the patient was asked to perform a voluntary slow movement, i.e. to raise the lower limb in the air. During such a manoeuver, the articulation of the wrist became gradually blocked: the passive movements became impossible because of increasing tonus which prohibited all other passive movements. This inability ceased after the patient returned to her initial relaxed position. The sign was present bilaterally, but was more marked on the right.

There was cerebellar dyssynergia, with dysmetria and dysdiadocokinesia on the left side of the body She had bilateral palmomental reflexes. Deep tendon reflexes were hyperactive. There was left Babinski sign, while the right plantar reflex was flexor. Hoffman and Rossolimo signs were bilaterally present. She had no grasping of the hands, but had a tonic foot response to stimulation of the sole (Goldstein, 1938) and a grasp reflex of both feet (Schuster and Pineas, 1926).

When she was sitting on a chair high enough to have the feet hanging in the



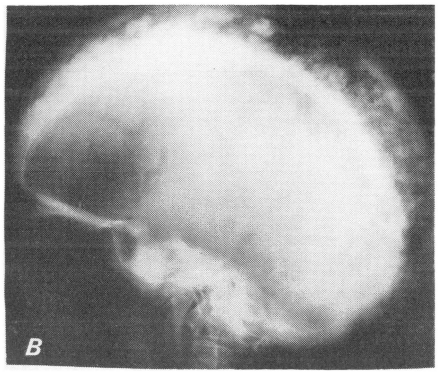


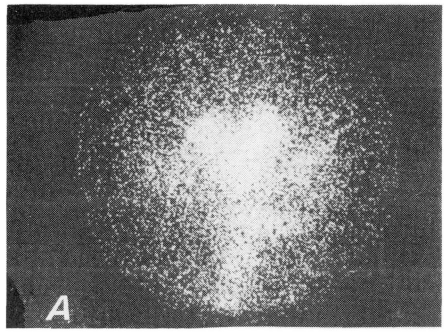
Figure 1—Roentgenogram of the skull. Both frontal (A) and lateral (B) views show severe Paget's disease with basilar invagination.

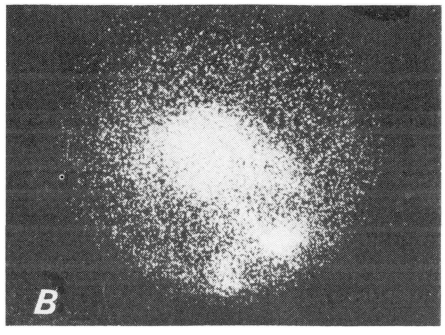
air, a forward and a lateral groping (Fradis and Botez, 1958; Botez 1974, 1975) were elicited on the right foot, but only a slight forward groping on the left was noticed. A sole-placing reaction (Botez, 1976) was elicited on the right foot only

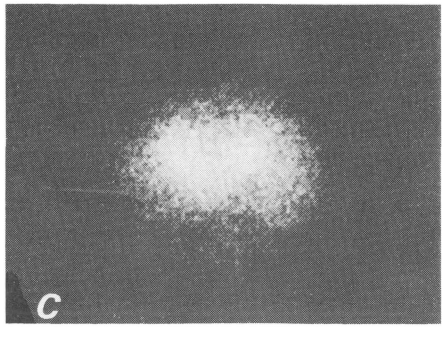
No sensory abnormalities were found. The cranial nerves were normal except for a diminution of hearing on the right.

X-rays of the cervical spine showed a short C_4 vertebral body, but the anteroposterior diameter was larger than normal. X-rays of the skull showed severe Paget's disease involving the cranial vault as well as the base with basilar invagination (Fig. 1).

The radionuclide cisternogram was performed with 111 Indium, fixed on D.T.P.A. (diethylene triamine pentacetic acid) following the technique described by Botez et al., 1975. It showed an abnormal ventricular reflux, ventricular dilatation and absence of peripheral







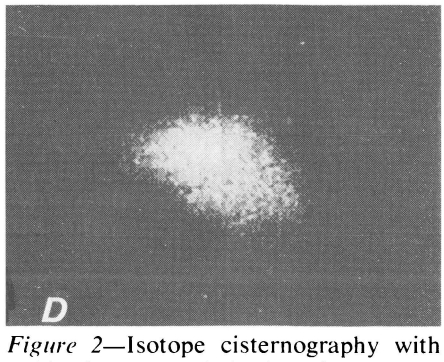


Figure 2—Isotope cisternography with 111 In — DPTA A, Anterior, 8 h. B, Left lateral, 8 h. C, Anterior, 48 h. D. Left lateral, 48 h. There is an important ventricular reflux, absence of peripheral progression and absence of peripheral reabsorption.

progression and of peripheral reabsorption (Fig. 2). These findings were compatible with a communicating hydrocephalus with severe ventricular dilatation. The patient had a normal cerebrospinal fluid content; CSF pressure was not determined.

Besides Phenoxene (chlorphenoxamine hydrochloride), Benadryl (diphenhydramine hydrochloride), and Symmetrel (amantadine hydrochloride) the patient received L-dopa 1.5 g daily. Her condition progressively worsened during the first month of hospitalization; she became bedridden, incontinent, and unable to feed herself. She developed bilateral Babinski signs. No other neuropsychological examination could be performed because of the marked dementia. She was transferred to the Neurosurgical Service of the Montreal Neurological Hospital for a shunting procedure. A pneumoencephalogram was not performed. Computerized transaxial tomography was not performed because the circumference of the head was too large for the scanner. One of us (G.B.) performed a right ventriculoatrial shunt using a Pudenz medium pressure valve. Intracranial pressure was found to be abnormally high.

During the post-operative period, she began to walk and to feed herself. Examination at the time of discharge (April 9, 1975) showed that mental status and overall functions were improved. She retained her tremor and rigidity, but was no longer incontinent. She was discharged on diphenylhydantoin 300 mg daily and L-dopa 750 mg daily

In May 1975 memory for recent events had improved but, tandem walking was still not possible. She had a bilateral grasp reflex of the foot, but no groping of the feet.

L-dopa treatment ceased in July, 1975. At that time, except for difficulty in tandem walking, her neurological examination was normal. Another neuropsychological examination was performed at the end of July 1975. The verbal I.Q. was 102, the performance I.Q. was 84 (full-scale score, 92); the score on the Kohs Block Design was 12 points.

In April 1976, the verbal I.Q. was 106, the performance I.Q. 94 and full-scale score, 101; the score on Kohs Block Design had risen to 36 points.

DISCUSSION

In summary, this 65-year-old patient with severe Paget's disease displayed a progressive neurological

deterioration with the following symptoms: (i) dementia, (ii) gait ataxia with spasticity and bilateral groping and grasp reflexes of the feet followed by the appearance of a bilateral Babinski sign, (iii) urinary incontinence, (iv) a left hemicerebellar syndrome, (v) a Parkinsonian-like symdrome that had been obvious from the early onset of the disease and (vi) a lack of drive.

I — Hydrocephalus has been mentioned as a possible complication of Paget's disease of the skull (Stauder, 1933; Dimitri and Aranovitch, 1936; Clegg, 1937; Riser et al., 1948; Goodbody and Roberts, 1950). The mechanism of hydrocephalus in Paget's disease is quite clear. An advanced basilar impression reduces the capacity of the posterior fossa and obstructs the flow of cerebrospinal fluid through the posterior fossa cisterns (Culebras et al., 1974).

The dominant clinical features in most reported cases of hydrocephalus with Paget's disease (Hann, 1910; Moynan, 1928; Stauder, 1933; Goodbody and Roberts, 1950; Katzenstein-Sutro and Bosch-Gwalter, 1960; Grossiord et al., 1967; Culebras et al., 1974) were gait disturbancés, sphincter incontinence and progressive mental deterioration.

The pneumoencephalographic studies in the three cases reported by Culebras et al. (1974) demonstrated little or no air entering the cisterna magna and lack or paucity of air flow into the subarachnoid spaces. The hydrocephalus was of the obstructive communicating variety with normal CSF pressure in two of their cases.

Our patient met all the clinical criteria of the occult hydrocephalus syndrome, i.e. mild dementia, gait ataxia and urinary incontinence (Adams et al., 1965; Messert and Wannamacker, 1974). The radionuclide cisternogram was suggestive of obstructive hydrocephalus without cerebral atrophy.

II — The pathophysiology of the grasp reflex of the foot and of the spasticity of the legs in the occult hydrocephalus syndrome have been discussed elsewhere (Botez, 1976; Botez et al., 1975; 1976).

III — The cerebellar syndrome is probably the consequence of some degree of cerebellar compression, which is frequently encountered in hydrocephalus secondary to Paget's disease (Culebras et al., 1974).

IV — Regarding the pathophysiology of the Parkinsonian-like syndrome, two points should be made. (i) The association between a Parkinsonian syndrome and occult, normal-pressure hydrocephalus has been reported in two cases by Sypert, Leffman and Ojemann (1973), who considered that mechanical distortion of the basal ganglia secondary to the hydrocephalus was a likely mechanism for the Parkinsonism. They suggested that mechanical distortion could also lead to circulatory insufficiency in the nigrostriatal system, which is known to lead to clinical manifestations similar to Parkinsonism. (ii) In one of the autopsied cases of hydrocephalus secondary to Paget's disease without Parkinsonian syndrome, microscopic examination suggested chronic compromise of blood flow in the territory of the arteriae thalamoperforantes and anteria choroideae posterior mediales (Culebras et al., 1974). A Parkinsonian-like syndrome secondary to hydrocephalus from Paget's disease has not been reported. The syndrome reported here might be due to vascular insufficiency in the nigrostriatal system.

V — The last problem is the indication for a shunting procedure. Three other shunted patients have been described (Culebras et al., 1974). Two of them died without improvement. The third had temporary improvement followed by a relapse, but remained demented even though the catheter was patent. On the basis of clinical and pathological findings, Culebras et al. (1974) postulated a two-stage evolution in the development of dementia in Paget's disease of the skull. Phase 1 is characterized by simple dilatation of the ventricular system associated clinically with mild dementia, gait disturbance and urinary incontinence. In this phase, patients should be shunted. Phase 2 comprises in addition, vascular thalamic lesions and is clinically characterized by advanced dementia. In this phase, patients considered to be irreversible should not be shunted (Culebras et al., 1974). The clinical course in our patient does not confirm this hypothesis. If the Parkinsonism syndrome in our patient was due to vascular lesions of the striatonigral system, she was in phase 2 and should not have benefited from the shunting operation.

We believe that patients with dementia and hydrocephalus from basilar impression secondary to Paget's disease should be shunted because this represents their only chance of improvement.

We have reservations about the safety and applicability of pneumoencephalography in this syndrome. Taking into account the pathophysiology of hydrocephalus in Paget's disease and data from the literature (Adams et al., 1965) plus our observations that a pneumoencephalogram can aggravate and decompensate a normal-pressure hydrocephalus syndrome, we are not in favor of this procedure in these patients. A radionuclide cisternogram and computerized transaxial tomography are the only safe investigations before the shunting procedure. We consider that the benneficial effect of the shunting procedure in our patient can be explained, at least partially, by the fact that no pneumoencephalogram was performed.

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