Parkinsonian-Like Syndrome Caused by Cyst in Posterior Fossa

Report of a Case

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It is well known that parkinsonism may be caused by a number of different etiological factors which need not be listed here. The occurrence of extrapyramidal syndromes, frequently associated with pronounced parkinsonian features, produced by supratentorial expanding lesions, particularly neoplasms, also is well documented.3,4. On the other hand, after careful review of the literature, no example of extrapyramidal syndrome caused by a space-occupying lesion in the posterior fossa was found. It seems therefore appropriate to report the following case in which a parkinsonian-like syndrome was caused by an expanding lesion within the posterior fossa.

Case Report

Ng, 61-2147. In September 1958, G.P.E., a 39-year-old white male right-handed truck driver, first began to complain of intermittent spells of vertigo of a few minutes' duration. At about the same time he started to complain of cramps in the left thigh. In September 1959, while working in a garage, kneeling beside a car, he suddenly became "dizzy," fell over, and experienced a severe "shaking" of both legs which lasted about 5–10 minutes. This episode was accompanied by vomiting. Some 3 months later, in December 1959, the patient had difficulty in walking caused by a stiffness of the legs, particularly the left. There occurred a "shaking" of his feet in any attempt to use his legs. The difficulty in walking increased steadily over a period of 3 to 4 months, so that by the summer of 1960 it was quite severe and he was unable to climb stairs. In addition, a very marked tremor and stiffness had also developed in the upper extremities. Fine movements of the hands were markedly impaired and handwriting, which had been very poor since the onset of the illness, became illegible. The patient was admitted to Notre Dame Hospital in Montreal in April 1960 and to the Verdun General Hospital in November 1960, with complaints of vertigo and vomiting. He was discharged from both hospitals with the diagnosis of a psychogenic disorder. During the next few months the patient experienced increasing stiffness of both legs, difficulty in starting to walk and in stopping once he had started. He also complained of vertical blurring of words while reading. He exhibited a sadness and irritability in comparison to his previous cheerful disposition.

1st Admission. The patient was admitted to the Montreal Neurological Institute in March 1961, with the chief complaint of stiffness and weakness of both lower extremities and unsteady gait.

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* Patient referred by Dr. D. Lloyd-Smith.

Examination. He was found to be rather apathetic. The following abnormal neurological signs were detected: mask-like facial expression, some limitation of upward conjugate movement of the eyes and some impairment of convergence. There was generalized muscular rigidity, more marked on the left than on the right side, and gross tremor of the legs especially on starting to walk. The patient seemed to move en bloc. There was a wide-based festinating gait and he had difficulty in starting to walk. Poor coordination was found in the left hand. In addition he had a brisk jaw jerk, marked generalized hyperreflexia particularly in both lower extremities, bilateral ankle clonus, and equivocal plantar reflexes.

Laboratory data including cytology and chemistry of the cerebrospinal fluid were normal. Roentgenograms of the skull and chest were also normal. The electroencephalographic study showed mild slow-wave abnormalities over both temporal regions, more marked on the left side. A pneumoencephalogram was carried out which was unsuccessful because the gas did not enter the ventricular system. The lumbar cerebrospinal-fluid pressure was normal. The callosal and cingulate sulci, although not displaced from side to side, were slightly high in position.

Course. A tentative diagnosis of Parkinson's syndrome was made. However, the presence of signs of involvement of the upper motor neurone was considered unusual. While in the hospital the patient was given a trial of Robaxin and Artane which resulted in marked diminution of muscular cramps and rigidity. He was discharged on this medication.

During the succeeding 9 months the patient was followed in the Outpatient Department. There was a progressive increase of stiffness and weakness in all extremities, especially on the left. Although he was still able to walk, there was an increasing difficulty to initiate walking and to stop, and he would fall frequently. There were episodes of vomiting and later, daily occurrences of paroxysmal pain in the neck.

Readmission to the Montreal Neurological Institute was in December 1961.

Examination. Positive neurological findings were emotional depression, mask-like facial expression, some limitation of upward conjugate deviation of eyes, some impairment of convergence, cog-wheel rigidity in all extremities, coarse tremor in all extremities at rest exaggerated by voluntary movements, extreme slowness in performing voluntary movements, festinating, wide-based gait, generalized hyperreflexia, bilateral sustained ankle clonus, and extensor plantar reflexes.

Laboratory data including cytology and chemistry of cerebrospinal fluid were again within normal limits. Roentgenograms of skull and chest were still normal. Electroencephalographic studies showed slight increase of slow-wave abnormalities over both temporal regions,
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more marked on the left, and a slightly less well regulated background activity in comparison with the previous record. The pneumoecephalogram was repeated (cerebrospinal-fluid pressure was still normal): the ventricular system did not fill with lumbar injection of gas. The elevated position of either the callosal or cingulate sulcus suggested some hydrocephalus; the pontine cisterna was small and the cisternae ambiens were spread apart.

Ventriculography was then carried out via biparietal twist-drill holes. Cerebrospinal-fluid pressure was elevated moderately with the head at about 30° above the horizontal plane. There was good communication between the lateral ventricles. The films showed moderate hydrocephalus of the lateral and 3rd ventricles. The aqueduct of Sylvius was somewhat widened and kinked with the upper narrow part of the 4th ventricle displaced forwards. The body of the 4th ventricle was displaced slightly upwards and forwards. The right lateral recess of the 4th ventricle did not fill. Traces of gas, after considerable manipulation, filled only the upper and posterior part of the cisterna magna (Fig. 1).

Operation. A bilateral suboccipital myoplastic craniectomy was carried out on Jan. 4, 1962. Intracranial pressure was exceedingly high. Cerebellar convolutions appeared normal. There was some herniation of the cerebellar tonsils. A cyst, about 1 inch in diameter, covered the vermis. The cyst was adherent to the pia-arachnoid but did not communicate visibly with the subarachnoid space and the fluid was under increased pressure. The cystic fluid was quite clear. The cyst was evacuated and its wall was removed as completely as possible. The upper surface of the vermis, corresponding to the floor of the cyst, was covered with somewhat pinkish arachnoid. After removal of the cyst the cerebrospinal fluid passed freely through the foramen of Magendie and there was free communication with the spinal subarachnoid space, the cisterna magna and the spaces about the medulla. Brain pressure remained low for the remainder of the operation.

Microscopic Study. The wall of the cyst revealed a thin membrane consisting of a single layer of scattered fibroblasts. The histological picture was considered compatible with the pathological diagnosis of an arachnoidal cyst. In contact with the arachnoidal membrane there was a small segment of cerebellar tissue which revealed absence of Purkinje cells and moderate degeneration of the granular-cell layer (Fig. 2).

Postoperative Course. A dramatic improvement in the patient’s condition was already present during the first few days after operation. Even his psychological attitude changed immediately after the operation. The patient, who previously had been apathetic and somewhat hostile, became cheerful and cooperative. The facial expression became normal. Limitation of upward deviation of eyes and impairment of convergence disappeared. Rigidity in the extremities decreased progressively and towards the end of the hospitalization, 24 days after operation, it was felt that the rigidity was reduced about 80 per cent in comparison with the findings on admission. Tremor had disappeared in all extremities and the patient was able to perform rapid alternating movements with minimal clumsiness. There was some residual spasticity in the legs, as demonstrated by very brisk deep tendon reflexes, but ankle clonus had disappeared, and plantar reflexes became flexor soon after operation. During the patient’s stay in the hospital his gait improved rapidly. The wide-based festinating gait was no longer present. However, some residual rigidity was present as demonstrated by diminution of swinging movements of the left arm and by the persistent tendency to move en bloc.

The patient has been followed in the Outpatient Department during the last 6 months. A further progres-
sive generalized diminution of rigidity and improvement of gait have been recorded. Fine movements of the hands and fingers have also improved remarkably as well as the handwriting which is now almost normal. On the last visit to the Outpatient Clinic the neurological examination revealed only minimal residual cog-wheel rigidity in the left arm.

A postoperative electroencephalographic study, carried out 2 months after the operation, revealed a marked decrease in the bitemporal slow-wave abnormalities seen preoperatively, and a better regulated background rhythm.

Discussion

It is not the purpose to discuss the pathogenesis of posterior-fossa cysts. It will be merely mentioned that the plausible diagnosis in the present case is that of an arachnoidal cyst, probably related to a developmental malformation.11,17

In reviewing the literature, no case of extrapyramidal syndrome caused by a space-occupying lesion in the posterior fossa was found. However, 1 case of so-called parkinsonism was found, related to an expanding lesion in the posterior fossa. It occurred as a transient phenomenon during the postoperative course following the partial removal of a left cerebellar astrocytoma.18 This syndrome was explained on the basis of compression of the midbrain because of postoperative swelling.

The neuroanatomical and neurophysiological basis of the extrapyramidal syndromes is still uncertain.1,13 Lesions of the substantia nigra, produced by ultrasonic irradiation in man, were very effective in reducing the rigidity and tremor in a small series of patients.14 This fact emphasizes the functional significance of the locus niger in the pathogenesis of parkinsonism, a significance which was deduced previously only from histopathological observations in man. No further reports have been found on the effects of nigral lesions on extrapyramidal syndromes in man, aside from 1 case of dystonia musculorum deformans alleviated by pallidotomy combined with a lesion of the substantia nigra.16 Conversely, at present, the preferred site of surgical lesions for relief of extrapyramidal syndromes is the nucleus ventralis lateralis of the thalamus, since with this procedure undesirable side effects are less frequent. Moreover, lesions in the nucleus ventralis lateralis of the thalamus result in the interruption of pallidothalamic and thalamopallidal fibers as well as cerebellothalamic circuits which seem to control the rigidity and the hyperkinetic phenomena.4,5,10

The cerebelloponto-olivary centers are postulated to represent the most important centers of coordination for the efferent pathways of the extrapyramidal motor system, together with the pyramidal and the cerebellar systems.19 In fact, there have been a few attempts to ameliorate the parkinsonian syndrome by means of cerebellar lesions, particularly lesions involving the dentate nucleus.7 Because of 1 fatality this approach was abandoned even though previous results were not discouraging. More recently, unilateral removal of the dentate nucleus was carried out in 3 cases of parkinsonism by Tótth18 in 1961. In 2 of these patients stereotaxic lesions in the pallidothalamic system had proven unsuccessful. Removal of the dentate nucleus was followed by marked decrease of rigidity in all 3 cases. Tremor was also de-

Fig. 2. Photomicrograph of surgical specimen showing arachnoidal membrane (right upper corner) forming the wall of the cyst, and cerebellar tissue which reveals absence of Purkinje cells and degenerative changes of the granular-cell layer. Hematoxylin-phloxine-saffron stain, X275.
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increased markedly in 2 cases, but increased in 1 case.

There is some resemblance between the case described in this paper and the "chronic progressive degeneration of the superior cerebellar cortex". In this condition ataxia, accentuation of deep tendon reflexes and parkinsonian rigidity may develop. The pathological picture is characterized by severe atrophy of the superior cerebellar cortex. Courville and Friedman suggested that the arachnoidal thickening, which frequently is present in this condition, may cause compression of the superior cerebellar arteries between the tentorium and the superior surface of the cerebellum and may thus interfere with the blood supply, which in turn may result in atrophy of the superior cerebellar folia.

It is suggested that the symptoms and signs of parkinsonism in the presently reported case were produced through pressure of the cyst upon the dentate nuclei bilaterally and their projection systems.

Summary and Conclusions

1. One case of parkinsonian-like syndrome caused by a large subarachnoid cyst lying above the superior vermis of the cerebellum is presented in which removal of the lesion was followed by immediate dramatic improvement which progressed to almost complete recovery in 6 months. It is suggested that the cure was the result of removal of focal pressure exerted upon the dentate nuclei and their projection systems.

2. The existing views on the importance of the cerebellum in the pathogenesis of extrapyramidal syndromes are confirmed, and it is hoped that the present report will stimulate further research into the role of the cerebellar systems in the physiopathology of parkinsonism and extrapyramidal diseases with the possibility of developing new surgical approaches.

References