

OLIVOPONTOCEREBELLAR ATROPHY

Chart

Operation	All combined abdomino-perineal resection of rectum (100 cases)	
Sex	Female 49	Male 52
Time postoperative	6 months to 10 years	Average 28.7 months
Age	29 to 78 years	Average 52.2 years
Plastic operation secondary to colostomy	6 for stricture	1 for prolapse
Protection cover for colostomy	Elastic belt36 Futurd colostomy belt.44 Home made device ... 8 Colostomy bag 2	
Results from irrigations	Good76 Fair12 Poor..... 4 No irrigations..... 8	
Frequency of irrigations	Every other day84 Every day 6 Sporadic 2 No irrigations 8	
Leakage between irrigations	Seldom12 Occasionally66 Regularly 4 None18	

OLIVOPONTOCEREBELLAR ATROPHY (HEREDOCEREBELLAR ATAXIA OF MARIE) WITH ENCEPHALOGRAPHIC FINDINGS

Report of a Case

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Hassin and Harris¹ have stated that heredocerebellar ataxia and olivopontocerebellar atrophy is one disease process. This observation was based upon their review of Keiller's² work, wherein cerebellar atrophy was observed clinically in 10 members of the same family. The

neuropathologic changes were alike in the cases studied and were typical of olivopontocerebellar atrophy. The outstanding changes noted were: diminished size of the cerebellar folia, atrophy of the olivary bodies and of the pons.

Thus, while olivopontocerebellar atrophy is a pathologic entity, clinically it resembles heredocerebellar ataxia of Marie so closely that it seems to have no definite differentiating features. The presence of a hereditary or familial history of this condition was regarded as the differentiating point between Marie's heredocerebellar ataxia and olivopontocerebellar atrophy. However, these authors feel that olivopontocerebellar atrophy may be both hereditary and familial, having demonstrated that, in the patients with the typical clinical picture of heredocerebellar ataxia, there were also typical pathologic findings of olivopontocerebellar atrophy.

With encephalography it is often possible to determine *in vivo* gross pathologic changes in the brain. At least one of the pathologic changes present in cerebellar atrophy is often gross enough to be detected by encephalographic study. Accordingly, encephalographic study was made of a patient who had the typical clinical picture of heredocerebellar ataxia of Marie (olivopontocerebellar atrophy), and this pathologic change was noted. A lateral view of the encephalogram is presented (figure), while the case report follows.

Case Report

A white man, aged 35, rather stocky and obese, complained of progressive difficulty in walking, talking, using his hands, and in balancing and walking up steps. There had been loss of strength in his legs. He felt that the weakness, uncertainty in walking, and the lack of control was now extending into his fingers and hands. The patient stated that he walked on smooth floors as though he were "walking on slippery ice." He recalled that many years ago he had had to look at the ground to keep his balance while running. An intention tremor had been present for two years. The patient stated that in the last year his speech had changed. "I can't talk fast." There was no dysphagia or bulbar sign. This patient had attacks in which he was unable to move when he awakened in the morning. He recalled a "concussion" which occurred when he was kicked in the head while playing football in high school. Herniorrhaphy was done in 1944. Family history revealed that his maternal grandmother had developed "creeping paralysis" at 65 and died at 67. No details were known.

General physical examination of the patient showed the heart, lungs, and abdomen to be negative, blood pressure 145/100. Examination of the cranial nerves revealed no abnormality. No nystagmus was present. Superficial reflexes were present and equal throughout. Deep reflexes showed marked hyperactivity bilaterally in the lower extremities. No clonus was present. The Babinski response was negative. There was marked ataxic disturbance, slightly greater on the left side. The patient walked with an ataxic gait, throwing his feet forward. There was a questionable loss of pain and temperature sense of the lower extremities to the knees. There was a bilateral loss of position sense and marked pastpointing. Romberg test was mildly positive.

OLIVOPONTOCEREBELLAR ATROPHY

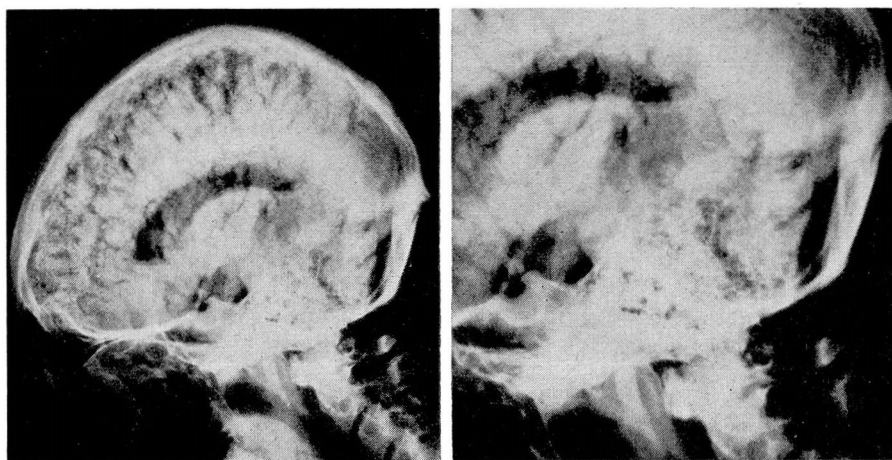


FIGURE a. Encephalogram, lateral view: Cortical air increased, particularly in frontal and parietal regions. Ventricular system shows moderate dilatation with no evidence of obstruction. Fourth ventricle is enlarged. Air overlying the cerebellum is considerably increased, giving the appearance of a projection of the occipital lobe over the cerebellar lobe. Cerebellum is markedly decreased in size. Pontine cistern seems enlarged. The distance from floor of the fourth ventricle to anterior surface of the pons suggests some loss of pontine substance. The olives cannot be visualized.

b. Enlargement of a. in the cerebellopontine area, giving greater detail of the described changes.

Blood study showed 13.0 hemoglobin (84 per cent); 9700 leukocytes; blood sugar 105 mg. per cent four and a half hours postprandially; blood Wassermann and Kahn tests negative. Urinalysis was negative. Spinal puncture showed initial pressure 190 mm. of water. Dynamic responses were normal. Five cubic centimeters of clear, colorless cerebrospinal fluid was removed, and final pressure was 110 mm. of water. Examination of the fluid revealed no cells, no globulin, total protein 33 mg., with negative Wassermann, Kahn, and colloidal gold reactions. X-ray examination of the cervical region of spine, normal; skull, normal. There was no evidence of platybasia and nothing suggesting brain tumor. Basal metabolic rate was minus 4.

Encephalogram was performed March 6, 1946, under sodium pentothal anesthesia. Pressure relations were normal. A total of 265 cc. of air was injected, while 255 cc. of fluid was withdrawn. Examination of this fluid was identical to that of the first fluid examined. It was felt that the patient had olivopontocerebellar atrophy (Marie's hereditary cerebellar ataxia).

Comment

There is presented here the encephalographic picture of a case of olivopontocerebellar atrophy, or Marie's hereditary cerebellar ataxia. While the history of similar involvement in other members of this family is admittedly weak, the clinical picture is that of Marie's disease. It is believed that the gross changes evident in this encephalographic picture represent in part at least some of the gross changes which are

found in olivopontocerebellar atrophy. There is present in these films evidence of marked cerebral cortical atrophy and a moderate diffuse enlargement of the ventricular shadows.

The presence of convolucional atrophy affecting the frontal and superior temporal lobe was emphasized by Keiller² in his studies. That a decrease in cerebral substance and an increase of fluid is present is evident in the amount of fluid removed (255 cc.). The most significant finding, the large collection of air overlying the cerebellum, is apparent. The occipital lobes overlapped the cerebellum, a finding which has been observed rather consistently in postmortem studies. The fourth ventricle is well visualized and appears to be enlarged, thus further emphasizing the marked decrease in size of the cerebellum. While it is in no wise as definite as the cerebellar atrophy, there appears to be an increase in the space representing the pontine cistern area and is suggestive of a diminution in size of the pons. However, this cannot be accurately determined. It is, of course, impossible to identify the olives.

As we stated before, gross changes typical of olivopontocerebellar atrophy are: diminished size of the cerebellum due to atrophy of the white substance of some cerebellar folia, atrophy of the olives and of the pons. In these films it is evident that there is diminished size of the cerebellum, a suggestive decrease in the size of the pons, and an increase in the size of the fourth ventricle; the olives cannot be identified. Thus it is felt that olivopontocerebellar atrophy is the pathologic condition present.

In the original description of this condition by Dejerine and Thomas³ in 1900 it was stated to be neither a familial nor a hereditary condition. It is now generally accepted that Marie's hereditary cerebellar ataxia and olivopontocerebellar atrophy are one and the same disease. Accordingly, it appears that this case fits into such classification. Roentgenographic studies of the less common neurologic conditions are of interest to those concerned with correlations between the clinical findings and the encephalographic changes found. It is with this thought in mind that this case has been presented.

Summary

1. A case of olivopontocerebellar atrophy (heredocerebellar ataxia of Marie) is presented with encephalographic findings which are believed to be consistent with the gross pathologic changes generally found at postmortem examination.

2. The changes observed were a marked cerebellar and cerebral cortical atrophy, moderate ventricular dilatation, and probable decrease in the size of the pons.

REFERENCES

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SPASTICITY: PHYSIOLOGIC AND NEUROSURGICAL CONSIDERATIONS WITH PRELIMINARY REPORT OF TWO CASES

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Spasticity is a condition of the musculature identified by partial or complete paralysis and increased resistance to movement associated with hyperactivity of the reflexes of the tendon and periosteum. The control of discharges going out over "the final common pathway" to produce normal muscle tone is a fine adjustment of many descending impulses arriving at the anterior horn cell, a summation of those from the cortex, basal ganglia, reticular substance, tegmentum, and cerebellum. In the surgical treatment of spasticity four distinct methods have been utilized: excision of the cerebral cortex, resections at lenticular level, cordotomy, and resection of the spinal nerve roots. This paper will discuss these procedures and present the results in two cases treated by cordotomy.

Knowledge of the pathologic process that brings about spasticity is far from complete. It has been shown that changes in muscle tonus can be produced by lesions in cortical areas 4, 6, and 4s, as well as in certain portions of the basal ganglia, reticular substance, tegmentum, cerebellum, and spinal cord.

Physiologically there is a wide diversity of opinion as to the mechanism producing spasticity. In the opinion of the authors it appears advisable to consider that increase in tonus is not always due to a release phenomenon but often to a predominance of one group of descending pyramidal and extrapyramidal impulses over another, or as Magoun has phrased it, "spasticity results from combined disturbances of extrapyramidal innervation." Hence, when attempting surgically to alter the tonus of muscle groups, we must consider all the descending impulses impinging on the anterior horn cell, the inhibitory as well as the excitatory (Bodian, Magoun, Hines, Murphy and Gellhorn).