DOUBLE VENTRICULAR SHIFT IN A CASE OF CEREBRAL HEMIATROPHY

ROBERT M. GEIST, JR., M.D., and C. ROBERT HUGHES, M.D.

Department of Roentgenology

CEREBRAL hemiatrophy has always been an entity involving considerable controversy and confusion. Since its first introduction into the American literature by Dyke, Davidoff, and Masson[1] in 1933, attempts have been made to standardize the terminology. The original 9 cases presented by these authors displayed atrophy of one cerebrum and homolateral skull changes associated with varying degrees of contralateral hemiplegia, Jacksonian seizures, and mental deficiency. In all but one of the cases the onset of the illness began during the first eighteen months of life. The etiology was variable and in several cases was unknown. Roentgenography revealed a homolateral thickening of the vault with over-development of the sinuses. Air studies demonstrated hemicortical atrophy and an enlargement of the ventricular system with a shift to the atrophic side. All of the cases presented had been examined several years after the original illness.

A similar series of 9 cases was presented a few years later by Casamajor and Laidlaw.[2]

Independently at the same time Alpers and Dear,[3] in an effort to clarify the subject, suggested that the classification be broken down into primary and secondary on the basis of the age of the patient when the original illness developed. They considered it to be primary when the patient was under 3 years of age. The signs and symptoms in their 22 cases of primary hemiatrophy were essentially the same as those listed by Dyke, Davidoff, and Masson, and by Casamajor and Laidlaw.

Ross[4] presented a group of 36 patients ranging in age from 11 to 49 years. Included in his series were both primary and secondary types but he did not make any attempt at differentiation. Emphasis was placed on the varied causes which included in order of frequency: central nervous system infection, obscure, congenital, traumatic, and systemic infection.

Considerable objection has been made by Josephy[5] to this procedure of lumping together a group of varied diseases which produced atrophy. It is his contention that the term cerebral hemiatrophy should not be used as a "catch all" but should be reserved for that specific type of true atrophy described by Schob in which there is a chronic parenchymatous degeneration of the cortex. This is in contrast to the lobar sclerosis or ulegyria in which there is a destructive process with interruption of the blood supply and scar formation. Such a classification on a histopathologic basis seems much to be preferred.

Our case of hemiatrophy may be classified as a primary type which histopathologically represents an extensive hemilateral ulegyria. Many of the cases

of Dyke, Davidoff, and Masson, and others also fall in this classification. This case is believed to be unique in that the patient was examined at the time of the original causative illness and the encephalographic changes observed throughout the course of the resulting atrophy. Of particular interest is the reversal in the direction of the ventricular shift.

Case Report

A girl 15 months old had experienced episodes of high fever of short duration for two months which the parents attributed to teething. On April 11, 1947, she became acutely ill with one of these attacks of fever. She failed to respond to the usual treatment and the following day was admitted to a local hospital. At this time the child was comatose and exhibited almost continuous right-sided Jacksonian convulsive seizures lasting twenty to thirty minutes. The temperature was 106 F. There was no significant history prior to the current illness and the child had apparently been normal since birth.

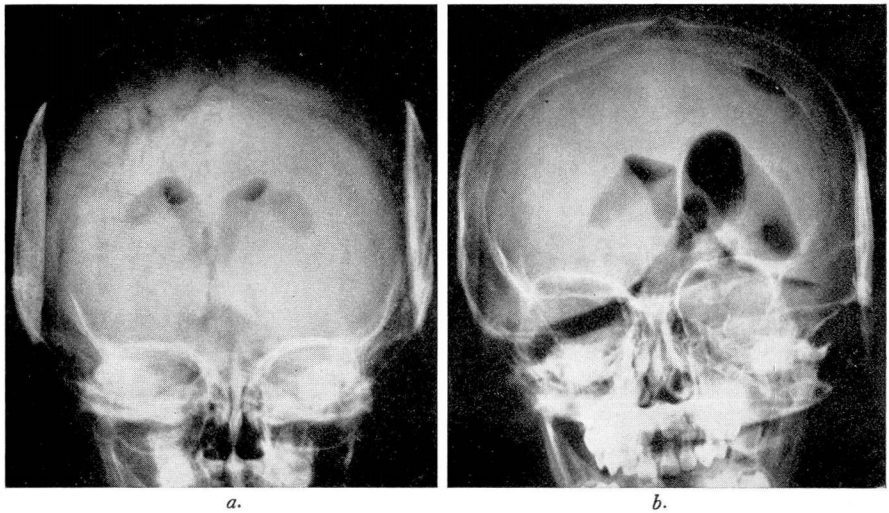
Sponge baths and enemas were instituted in an effort to reduce the fever. In the course of the first day the temperature dropped to 101 F. and the convulsions ceased. Examination showed a right facial paralysis. The pupils were round and equal. The disks were not choked. A spastic right-sided paralysis was present. Spinal tap revealed normal dynamics with a normal cell count, protein and sugar. The urine was negative. The white blood cell count was 9100 with 81 per cent neutrophils. The child was placed on large doses of penicillin and sulfadiazine. Her condition remained unchanged with the temperature ranging from normal to 100 F.

On April 16, she was transferred to the Cleveland Clinic Hospital. On admission the child revealed a right hemiplegia without loss of sensation. There was no choking of the disks and reflexes were normal. Examination of the blood showed 4,120,000 red blood cells with 9.5 Gm. hemoglobin and 5275 white blood cells with a normal differential count. The Kahn test was negative. The urine and stool examinations were negative. Routine skull films were normal.

The child's condition remained unchanged and on April 18, an encephalogram was done. Forty-one cubic centimeters of clear colorless fluid were displaced with 51 cc. of air. The spinal pressure could not be measured. On analysis the spinal fluid was normal. The films showed obliteration of the cortical sulci on the left with slight displacement of the ventricular system to the right (fig. 1a). The possibility of acute cerebral edema from a cerebral infarct was considered most likely although a neoplasm could not be ruled out. The condition of the child changed little with the exception that she became more alert.

On April 30, a second encephalogram was made. Forty-eight cubic centimeters of spinal fluid were displaced with 58 cc. of air. On analysis the fluid was normal. Films showed no change from those taken on April 18. An exploratory trephine of the left parietal region was then carried out. The underlying cortex appeared normal but a cannula introduced encountered a widespread area of softening. A biopsy was made which showed severe tissue degeneration. The patient was discharged from the hospital on April 30, with the diagnosis of probable infarction.

The child was readmitted to the hospital on August 12, 1948 for follow-up study. She had shown a steady improvement throughout the year; mental development seemed normal and she was able to walk fairly well. Speech was good. However, the right arm showed only minimal recovery. On August 13 an encephalogram was again performed. Sixty cubic centimeters of fluid were displaced with 70 cc. of air. The initial pressure was 330 mm. of water. Examination of the films showed a severe left cortical atrophy with dilation of the left lateral ventricle and the third ventricle. There was a shift of the entire ventricular system to the left (fig. 1b). The diagnosis was that of cerebral hemiatrophy following infarction. The child was discharged on August 18.



a. *b.*

FIG. 1. (a) Encephalogram done April 18, 1947 showing obliteration of the cortical sulci with displacement of the ventricular system to the right. (b) A second encephalogram done August 12, 1948 showing hemiatrophy with a reversal in the direction of the ventricular shift.

Comment

We have been particularly interested in the encephalographic changes which this case of cerebral infarction demonstrates. The loss of visualization of the cortical sulci and the shift of the ventricular system to the contralateral side in the acute phase is what one would expect in view of the extensive edema and necrosis. However, the differential diagnosis between such a process and a space-filling lesion presented a problem which could not be solved definitely except by biopsy and progress encephalography. On follow-up encephalography, severe atrophy of the left cerebrum was demonstrated. The left lateral ventricle was considerably dilated and the entire ventricular system was shifted to the ipsilateral side. Even at this point when the diagnosis of cerebral infarction with atrophy was established, the actual cause of the infarction could not be determined. The possibility of an infectious etiology is plausible although purely speculative.

The degree of atrophy present at this time was measured by the method of Evans[6] in which the width of the lateral ventricles is compared to the width of the skull. We obtained a figure of .34 (normal .16 to .29). Despite the atrophy there was no associated homolateral skull thickening. Since this is believed to be a process resulting from decreased internal growth pressure, such changes would be expected to appear eventually.

The actual mechanism of ventricular shift such as was demonstrated in our case is discussed at length by Robertson and Childe,[7] and Crothers and Wyatt.[8] They are of the opinion that the shift as demonstrated in acute edema is on

the basis of expansion alone. In cerebral atrophy the shift towards the atrophic side is attributed to differential growth. They were unable to demonstrate any evidence that the shift was due to scarring or traction from adhesions.

In our opinion, this case supports the contention of Walker[9] that serial encephalograms in all types of retardation and palsies in children are of great diagnostic significance.

Summary

In a case of cerebral hemiatrophy the active and passive process is demonstrated by means of serial encephalography showing a double ventricular shift. The diagnostic significance of serial encephalography in retardations and palsies in children is illustrated.

References

1. Dyke, C. G., Davidoff, L. M., and Masson, C. B.: Cerebral hemiatrophy and homolateral hypertrophy of skull and sinuses. Surg., Gynec. and Obst. **57**:588-600 (Nov.) 1933.
2. Casamajor, L., and Laidlaw, R. W.: Convulsive states with evidence of brain hemiatrophy. Am. J. Psychiat. **96**:165-175 (July) 1939.
3. Alpers, B. J., and Dear, R. B.: Hemiatrophy of brain. J. Nerv. and Ment. Dis. **89**:653-671 (May) 1939.
4. Ross, A. T.: Cerebral hemiatrophy with compensatory homolateral hypertrophy of skull and sinuses, and diminution of cranial volume. Am. J. Roentgenol. **45**:332-341 (March) 1941.
5. Josephy, H.: Cerebral hemiatrophy (diffuse sclerotic type of Schob). J. Neuropath. and Exper. Neurol. **4**:250-261 (July) 1945.
6. Evans, W. A., Jr.: Encephalographic ratio for estimating ventricular enlargement and cerebral atrophy. Arch. Neurol. and Psychiat. **47**:931-937 (June) 1942.
7. Robertson, J. S., and Childe, A. E.: Relation of ventricular asymmetry to contracting intracranial lesions. Arch. Neurol. and Psychiat. **43**:80-89 (Jan.) 1940.
8. Crothers, B., and Wyatt, G. M.: Mechanism of displacement of ventricular system in children with atrophic lesions. Arch. Neurol. and Psychiat. **45**:246-260 (Feb.) 1941.
9. Walker, A. E.: Encephalography in children. Am. J. Roentgenol. **32**:437-456 (Oct.) 1934.