A CASE OF DECEREBRATE RIGIDITY WITH AUTOPSY.*

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DECEREBRATE rigidity was first described by Sherrington¹ in 1896. His work was concerned solely with laboratory animals, and it was not until 1919–1920 that this syndrome was described in man. At about the same time, Walshe², Wilson³, and Cobb⁴ reported clinical cases illustrating the phenomenon of decerebration. Since then numerous publications have appeared, and the more recent reports giving references are those of Weisenburg and Alpers⁵ and Zand⁶. The case herein reported was discussed with Professor Stanley Cobb, who supervised the histological work and aided in preparing this case for publication.

CLINICAL REPORT.

The patient was a male child, the youngest of nine children, born February 15, 1919, at eight months' pregnancy and after a long difficult labour. His paternal uncle was feeble-minded. Otherwise the family history was negative. The child was blue at birth and remained blue for six weeks. A few hours after birth the first convulsion occurred. Thereafter he had convulsions almost every day. These were generalized, beginning with retraction of the head and protrusion of the eyeballs and followed by rigidity of the arms, clonic movements of the extremities, frothing at the mouth and cyanosis. The patient was said to be exceedingly nervous. "If someone would knock at the door or a dog would bark he would stiffen all over." Any noise would start a spasm. He had no illnesses in childhood, but he did not develop normally; he never sat up, never walked or talked; he had no control of his sphincters.

The patient was admitted to the Monson State Hospital on September 22, 1921, at the age of two years and seven months. He died there at the age of eight years and three months, on May 20, 1927. On admission a diagnosis of epilepsy with mental deficiency was made. At that time physical examination showed a well-nourished boy, mentally below par. His height was 31 inches; weight 21 pounds. Head measurements were: circumference 16 inches; transverse arch 9 inches; glabella toinion 10 inches. The skin was clear and very dry. On the sacral region there was a pigmented spot with a growth of long hair. There were no skeletal deformities. Examination of eyes, nose, ears, mouth and throat was negative. Heart, lungs and abdomen were normal. Patient could not sit, stand or walk.

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He came under the observation of one of us on December 4, 1926, at the age of seven years and ten months. At this time neurological examination was made and revealed the following condition.

Patient lay in bed in a rigid state with the head strongly retracted. The lower limbs were extended, adducted and crossed in scissors-like fashion; knees were slightly flexed, ankles extended, and toes in plantar flexion. The upper limbs were flexed and internally rotated; wrists were pronated, fists closed with the thumbs usually between the first and second fingers. There was very slight spontaneous movement of the legs, and only slight flexion and extension of the knees. The spontaneous movements of the lower limbs were more varied, viz., flexion and extension of the elbows and abduction of the arms. There were also frequent spontaneous movements of the head. Movements of the upper limbs usually occurred simultaneously with the movements of the head. If the head was rotated to one or the other side, the extremity towards which it was rotated (maxillary extremity) would become extended, slightly abducted and internally rotated, and the opposite extremity on the side of the occiput (cephalic extremity) would become flexed. His favourite position of head and arms was, head turned to the right, left arm flexed upon the chest so that the hand came under the chin, and right arm semi-flexed and reposing on the epigastrium. If the patient was lifted into the air on one hand he remained rigidly extended like a block of wood.

A peculiar general motor reaction could be produced by sudden external stimuli. Among these, auditory stimuli such as a blow on a table nearby, sudden clapping of hands, wrinkling of paper, were especially prone to produce this reaction. It was absolutely constant and uniform, and consisted of the following motor responses: Muscular rigidity, retraction of the head and opisthotonos became accentuated; the arms became extended, internally rotated and abducted in a spasmodic manner; the legs were also abducted and extended, sometimes drawn up on the torso, and sometimes there was only extension and flexion of the knees.

Along with these phenomena there was a spasm-like movement of the face, the resulting expression resembling that of an angry or frightened chimpanzee. The lower jaw was protruded forward, the mouth closed, the lower lip everted and the corners of the mouth retracted downward and outward; the eyebrows were raised, eyes opened wide and pupils were dilated. All this was accompanied by deep inspiration and a short period of apnoea, sometimes with flushing of the face.

In addition to this general motor reaction caused by external stimuli, especially auditory, certain local motor reactions could be elicited by local stimuli applied to the skin. If the skin of the ankle were pinched, a local motor reaction would follow, consisting of slight dorsiflexion of the ankle, slight external rotation of the thigh, sometimes a drawing up of the whole leg and occasionally extension of the contralateral leg. These local motor reactions were moderate in intensity and were not quite constant.

The tendon reflexes were all present, equal, but small in range. The response to plantar stimulation consisted of a slight extension of the big toe with occasionally spreading of the other toes. Stimulation of the abdomen produced contraction of the muscles of the stimulated side, giving the impression that the abdominal reflexes were present. The pupils were equal, regular and reacted to light; reaction to accommodation could not be tested. There was a slight divergent strabismus, but ocular movements were free in all directions. Fundi were normal in appearance.

There was generalized muscular rigidity. On palpation the muscles were hard and apparently in a state of semicontraction. Since the patient was emaciated, every muscle could be seen under the skin. On passive movement the rigidity could be readily appreciated. Resistance to passive movement was pronounced and increased with the force applied. The rigidity resembled the plastic, waxy type. If a leg was flexed at

the knee, it would maintain this position for a few minutes before returning to the original position of extension. There were no contractures, excepting in the flexor tendons of calves and flexors of toes.

Another feature of this patient's condition was the occurrence of 'crying spells.' They consisted of a deep inspiration, then a period of apnoea, when the face became very congested, then a cry with a long expiratory phase; simultaneously the whole body would stiffen out; and a few short respirations would end the spell. These fits were often induced by any unpleasant manipulations connected with various tests used in the examination.

COMMENT.

On the basis of the history and the physical findings it is apparent that we are here dealing with a case of cerebral infantile tetraplegia either of congenital origin or due to a lesion occurring during or immediately after birth. The neurological picture is obviously not of recent development but was present since infancy. The main clinical features may be discussed under four headings:

1. Attitude and postural reactions.
2. Muscle tonus.
3. Fits.
4. General and local motor reactions.

1. The patient's attitude was one of permanent extension with retraction of the head, which was maintained even if he was lifted up into the air on one hand. In addition he presented the definite postural reaction known as the tonic neck-reflex of Magnus and de Kleijn. These postural reactions exhibited remarkable constancy; they could be readily induced by passive movements of the head and were often reproduced by spontaneous movements of the head.

2. Muscle tonus was increased. The state of hypertonus was considered as rigidity, to be differentiated from spasticity or pyramidal hypertonus. In fact this case clearly illustrated the main intrinsic characteristics of rigidity. It was equally distributed in the flexor and the extensor muscles. The reaction of the antagonists was exaggerated. Finally, shortening and lengthening reactions were present. The rigidity was waxy, not elastic. Except for the scissors-like crossed legs, which it must be said developed at a later time, and for the faint plantar extensor response, the patient did not exhibit gross pyramidal signs.

3. Since birth the patient had had fits. These were apparently not all alike, but two types may be distinguished. In one type the patient would become rigid and cyanotic and apparently unconscious, then he would have a generalized convulsion with frothing at the mouth. In the second type, which was designated as 'crying spells,' his face would become congested, he would take a deep inspiration and then stiffen all over, but he did not shake and apparently did not lose consciousness. The first type of fit consisted of a tonic and clonic phase with loss of consciousness. The second type of fit consisted simply of the tonic phase without any obvious loss of consciousness. These were purely tonic fits and closely resembled those general motor reactions induced by external stimuli.

4. The patient exhibited motor reactions both general and local, when various external stimuli were applied. The general reactions resembled very closely the tonic fits, and could be produced by any sudden, intense external stimulus. These occurred with the greatest constancy and least provocation following auditory stimuli. In the local motor reactions the patient exhibited moderately exaggerated reflexes of defence, which were not so constant as the general reactions.

FIG. 1.—Superior surface of brain, showing microgyria and absence of corpus callosum. Arrow indicates level of coronal section shown in Fig. 3. Numbers show where specimens were removed for microscopic study: 1, frontal; 2, frontal; 3, motor; 4, low motor; 5, post-central; 6, area striata.

PATHOLOGICAL REPORT.

Gross Examination: The dura is thickened and moderately adherent to the cranium. The subarachnoid space is wide in many places, containing much fluid; in other places the pia-arachnoid is thickened and adherent to the brain surface, especially over the frontoparietal region and along the longitudinal fissure. The cerebral hemispheres are small with marked microgyria,

especially in the precentral region (Fig. 1). The frontal lobes are greatly reduced in size, making it difficult to distinguish landmarks and even the boundaries of the parietal lobes (Fig. 2). The corpus callosum is absent, allowing a direct communication between the third ventricle and the space between the occipital lobes and the cerebellum. On coronal section, the ventricles show marked dilatation (Fig. 3).

FIG. 2.—Inferior surface showing small asymmetrical frontal lobes, relatively large temporal lobes and cerebellum, and marked thickening of the pia-arachnoid.

Microscopic Examination: Sections (taken from the frontal, precentral, postcentral, temporal and occipital lobes, the cerebellum, basal ganglia, pons, medulla and spinal cord) were studied microscopically with the following stains: cresyl-echt violet, phosphotungstic acid hæmatoxylin (Mallory), hæmatoxylin and eosin, and Kulschitsky-Pal modification of Weigert's iron hæmatoxylin stain.

FIG. 3.—Coronal section (at level indicated in Fig. 1), showing the aplasia of the parietal lobes and the absence of corpus callosum. There is also a marked dilatation of the ventricles. Microgyria is conspicuous at the vertex.

FIG. 4.—Section of motor cortex (see Fig. 1, 3) showing narrow cortex, poor lamination and paucity of nerve cells. (Cresyl-echt violet stain).

In all sections of the cortex the cellular architecture is greatly disturbed, and the laminae are difficult to distinguish. The cortical cells are reduced in number and replaced by glia. In the frontal cortex, laminae 1, 2 and 5 are fairly distinct in some places, although they are very narrow. The nerve-cells are normal in appearance, but are arranged in a very disorderly fashion. In lamina 5 of the motor cortex there are a few large, elongated, darkly stained cells, but there are no normal Betz cells (Fig. 4). The streak of Gennari in the area striata is broken up into short pieces. Better architecture is seen in the

FIG. 5.—Section of motor cortex (see Fig. 1, 3), showing neuroglial proliferation and marginal gliosis. (Phosphotungstic acid hæmatoxylin stain).

postcentral and temporal cortex, but the great paucity of nerve-cells is striking. Ammon's horn cells are pale, and the layers are not clearly defined in all places.

Neuroglial proliferation is very prominent (Fig. 5). In some sections it is chiefly marginal, forming a felted network on the periphery; in others, the glial fibres spread out irregularly into all the cortical layers (Fig. 6). The blood-vessels also are very conspicuous. There is great congestion and apparent increase in number of capillaries (Fig. 7).

The cerebellum shows considerable abnormality. The Purkinje cells are reduced in number and are arranged in an irregular fashion, large clumps of

cells in one place and a few isolated cells in another. The granular layer is smaller than normal and the outer molecular layer is relatively larger. There is moderate gliosis.

The basal ganglia also show lesions. Throughout the thalamus there is a fine network of fibres and an increase of glia nuclei. Around the small blood-vessels are areas of granular lilac-staining degeneration. In about the middle of the thalamus there is an area of degeneration, indicated by numerous scavenger cells loaded with myelin in the perivascular spaces. The caudate and lenticular nuclei show scarcity of cells, increase of glia nuclei and excess of capillaries.

FIG. 6.—Section of postcentral cortex (see Fig. 1, 5) showing severe gliosis obliterating cell structure and lamination. (Phosphotungstic acid hæmatoxylin stain).

The exact location of the red nucleus is difficult to make out. Cells are scarce. There is a moderate increase of glia nuclei. Around the small blood-vessels are areas of granular degeneration which are even more conspicuous here than in the thalamus. The substantia nigra shows a slight increase of melanin.

In the pons and medulla there is pronounced diffuse gliosis and a conspicuous excess of capillaries. Numerous granular areas are interspersed among the pyramidal fibres. The medullary nuclei are deficient in cells.

Sections of the spinal cord show a most striking picture. The grey matter appears normal, except for congestion of capillaries and many petechial hæmorrhages which are probably agonal in origin. The ventral horn cells are normal.

FIG. 7.—Section of frontal cortex (see Fig. 1, 1), showing great excess of capillaries. (Phosphotungstic acid hæmatoxylin stain).

FIG. 8.—Section of spinal cord showing extensive areas of unmyelinated fibres, especially in ventral and lateral columns. (Kulschitsky Pal-Weigert stain).

In the white matter are diffuse areas of demyelination in the peripheral and the lateral and ventral columns, sparing the posterior columns and the ground-bundles, which appear normal (Fig.8). This lack of myelin is more obvious

in the cervical than in the thoracic region. In the latter the process appears to be limited to the lateral columns, chiefly the corticospinal tracts. In the cervical region all the tracts in the periphery of the cord show a great dearth of myelinated fibres. There is conspicuous fibrillary gliosis in the lateral columns in the region of the corticospinal tracts (Fig. 9).

Anatomical Diagnoses: Microcephaly and microgyria. Cerebral aplasia, with absence of corpus callosum. Aplasia of cerebellum. Aplasia of medulla. Aplasia of basal ganglia. Degeneration of red nucleus. Degeneration of thalamus. Diffuse gliosis. Corticospinal tract degeneration.

FIG. 9—Section from lateral column of spinal cord showing great fibrillary gliosis. (Phosphotungstic acid hæmatoxylin stain).

SUMMARY.

This case represents the clinical condition of decerebrate rigidity with tonic neck-reflexes of Magnus and de Kleijn, extensor rigidity, pseudo-affective reactions (hyperacusis), and tonic fits. Pathological study of the central nervous system revealed microcephaly and microgyria, agenesis and aplasia with secondary degeneration of the spinal tracts, diffuse gliosis, and degenerative lesions in the red nucleus and thalamus.

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