APRAXIA IN CORPUS CALLOSUM LESIONS

REPORT OF THREE CASES

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INTRODUCTION

IN 1884 Bristowe¹ formulated a syndrome of tumours of the corpus callosum emphasizing the occurrence of profound mental disturbance, a tendency to double hemiparesis, difficulty in swallowing, loss of speech and want of control of the bladder, along with negative signs such as the absence of cranial nerve involvement and the mildness of signs due to increased intracranial pressure. Giese,² in 1892, and Ransom,³ in 1895, reported one case each, Giese emphasizing the importance of the double hemiparesis, while Ransom, whose contribution included a review of the cases reported to that time, concluded that the diagnosis could not be made with certainty, but that the occurrence of profound mental disturbance with a double hemiplegia was suggestive of corpus callosum localization. Giese even went further and attempted localization in the different portions of the corpus callosum. In his conclusions, he makes the comment that when in addition to Bristowe's syndrome there are present signs of cerebellar involvement, then it is very likely that the lesion is in the splenium. Eight years later Zingerle⁴ described a case of tumour of the corpus callosum in which the patient early in his illness showed psychic retardation, but on neurological examination manifested an inability to stand or walk even though the muscle power was good and there was no ataxia in the performance of non-equilibratory tests. This striking 'defect in coordination' which manifested itself in an inability to stand or walk in the absence of profound muscle paralyses or ataxia, Zingerle attributed to involvement of the corpus callosum and called it 'callosal ataxia' ('Balken-Ataxie'). In 1905, and again in 1907, Liepmann⁵ made fundamental contributions to the study of the corpus callosum, pointing out that the corpus callosum was the pathway through which cerebral dominance may express itself. He described the occurrence of left-handed apraxia in right-handed individuals as diagnostic of corpus callosum lesions. Hartmann⁶ confirmed Liepmann's findings, and Critchley,⁷ in a comprehensive paper on the distribution of the anterior cerebral artery, again showed that when through a lesion of the callosal branch softening of the corpus callosum follows, left-handed apraxia is demonstrable clinically, providing, of course, the mental and physical state of the patient permits testing.

On the other hand, a good many investigators have attempted to interpret the mental picture encountered in corpus callosum lesions as specific for the corpus callosum. Bristowe had commented on the profound mental disturbance, which simulated that of general paralysis of the insane. In 1906 Raymond⁸ reported two instances of corpus callosum tumour. Although he noted the absence of cranial nerve involvement and commented on the fact that one of his patients showed a disturbance in coordinating movements of his body when walking, Raymond felt that the mental picture was of more significance in localization. He regarded a disturbance in the association of ideas, a memory defect, particularly for recent events, an alteration in character, such as irritability and changing moods, and later a clouding of consciousness or dementia as a characteristic mental picture of corpus callosum lesions. Lippman⁹ collected 50 cases from the literature, to which he added one of his own, and of the 51 cases only two were free of psychic disturbances. Lippmann also quotes Schuster's series of 33 cases, in 31 of which psychic disturbances were found. In 1914 Stern¹⁰ in a detailed paper stated that the syndrome described by Raymond must be rare. His material did not substantiate the high incidence of psychic disturbances as noted by Lippmann and Schuster. Stern felt that mental symptoms may be entirely absent, although frequently enough the mental symptoms appeared early, and were profound. Without regarding any specific psychic disturbances as diagnostic of the corpus callosum, Stern thought that when mental symptoms do occur they are likely to be of an amnesic nature resembling Korsakoff's syndrome and to consist also of defect in spontaneity. In their report of 14 cases of tumours of the corpus callosum, Ironside and Guttmacher¹¹ thought that mental changes such as apathy, drowsiness and defect in memory were often the first symptoms. Recently, Alpers and Grant¹² reported five cases of tumour of the corpus callosum formulating a syndrome which strongly emphasized the mental changes. According to their observation, difficulty in concentration and attention and defective comprehension are the characteristic mental changes in tumours of the corpus callosum. They write: 'A syndrome with the mental symptoms described and with hemiparesis and apraxia is almost characteristic of involvement of the corpus callosum. Less characteristic, but none the less indicative of callosal implication by tumour, is a syndrome consisting of pronounced mental symptoms with drowsiness and hemiparesis.' Alpers and Grant quote Mingazzini and Lévy-Valensi and Milani, all of whom felt that mental changes form the predominant symptomatology of corpus callosum disease.

It would seem, then, that in priority and accuracy of observation the syndrome described by Bristowe forms a sound nucleus for corpus callosum localization. Later investigations have either elaborated upon the mental changes encountered in the attempt to describe a specific psychic syndrome or have consisted of additions, the most significant being Liepmann's apraxia of the left hand. The difference of opinion in the literature as to the relative

significance of the mental changes and the physical findings may be accounted for either by the interval of time from the onset of the disease to the time when the patient comes under observation or more likely by the amount of neighbourhood invasion.

It is well known that growths of the corpus callosum invade the frontal lobes, and it may well be that the profundity of the mental alteration depends upon the extent of the spread into, and destruction of, the frontal lobes. It is also very likely that the severity of the pyramidal tract involvement is again an indication of the amount of pressure imposed upon, or of actual implication of, the internal capsules. If, then, as is likely, the paralysis and the personality changes are essentially neighbourhood signs, what remain of Bristowe's syndrome attributable directly to the corpus callosum are rather the negative signs, such as the lack of increased intracranial pressure and the rarity of cranial nerve involvement. On the other hand, the 'Balken-Ataxie' of Zingerle and the apraxia of Liepmann cannot be explained as neighbourhood signs. They appear to result from a disturbance in the corpus callosum itself.

Strictly speaking, the callosal ataxia of Zingerle is not an ataxia at all. Rather is it a disturbance in the execution of an idea in the presence of competent muscle power and a coordinating system which is intact when the patient is recumbent. As such it is an apraxia of gait. Although Liepmann's dictum that left-handed apraxia is a result of corpus callosum disease is correct, we would like to add that disease of the corpus callosum will produce disturbance in praxis which need not be confined to the arm, but may manifest itself in apraxia of the lower extremities or apraxia of the muscles of mastication or swallowing. In two of the three cases to be reported below, the localization of corpus callosum was made on the basis of an apraxia of gait; in the third, localization was made before the preagonal appearance of an apraxia of mastication and deglutition.

PERSONAL CASES

Case I.—Tumour involving the genu of the corpus callosum; history of 13 months' duration; apathy with drifting attention; mild bilateral pyramidal tract signs and apraxia of gait.

L.D., age 53, was admitted to the service of Dr. Foster Kennedy in August 1926. Her condition was alarming. She was in coma, respirations were stertorous and the pulse rapid and feeble. She was having a series of tonic fits suggestive of incomplete decerebrate rigidity. The head was twisted sharply to the right, the right arm was held rigidly flexed at the elbow, the left arm was extended and rigid, while the lower extremities were in extension spasm.

The history obtained from members of the family was that the patient had been quite well before September 1925. On Labour Day, 1925, she fell down a flight of stairs while under the influence of liquor. There was no loss of consciousness and no bleeding from the orifices. From that time she complained of headaches. In July 1926 she visited a public beach and went bathing in the nude. The family now noticed that her memory was very poor, that she lost interest in her home, and spent most of the time sleeping. When aroused, she complained of severe headache and on

several occasions the headache was accompanied by projectile vomiting. The past history was irrelevant.

Twenty-four hours after her admission the patient recovered from the initial coma and fits. Repeated examinations over a period of two months showed the following: She was apathetic but capable of normal emotional reactions. The two striking things about her mentality were an easy fatigability and an utter absence of any attempt to initiate activity. Her cooperation in the examination could not be kept longer than ten minutes at any one time. She was entirely unconcerned about her environment.

The pupils were equal, regular and reacted to light and upon accommodation. There were no ocular palsies and no nystagmus. The fundi were negative; the fields reconstructed after repeated examinations seemed to be intact. The motor and sensory fifth, including the corneal reflexes, were normal. There was a left central facial weakness, more apparent when the patient laughed. Hearing, phonation, palate, tongue and sternomastoid innervation were intact.

There was no frank paresis. Power of the grips and lower extremities was good. The patient executed movements slowly and deliberately. Finger-to-nose, heel-to-knee, and toe-to-object tests showed no disturbance in coordination. There was no apraxia in the hands.

The deep reflexes in the upper and lower extremities were grossly but equally overactive. The abdominal reflexes were not elicited. There was a bilateral Babinski sign.

Joint position sense was intact. Objects placed in the hands were readily recognized. There was no demonstrable change to pin-prick, touch and temperature.

In testing the gait, it was noted that the patient could not sustain herself. At times she would fall backwards, at other times she would collapse in a heap before the examiner. When supported and asked to take a few steps, the patient made no attempt to walk. The entire weight of her body was thrown upon the examiner and she seemed puzzled as to what was wanted of her.

The laboratory data showed an NPN of 40, a blood sugar of 130. The cerebrospinal fluid was clear, under normal pressure and had 40 lymphocytes per c.cm. The blood and spinal fluid Wassermann was negative.

Her behaviour during almost two months of hospitalization is best described as vegetative. She slept most of the day. She ate if and when she was fed. She was incontinent of faeces and urine and was content to lie in the evacuations until she was attended to. She was never heard to complain and never seen to speak to the patients about her. She died suddenly on the day she was to be operated upon.

The post-mortem examination was limited to the head. Inspection of the brain showed an ironing out of the right frontal convolutions. There was a slight bulging of the right frontal lobe. Upon separation of the two hemispheres, a bulging of the medial aspect of the right frontal region became apparent. Coronal sections of the brain, approximately 1 cm. apart, showed a soft cystic mass in the right frontal lobe extending from the second to, and including, the sixth section. The tumour had invaded and destroyed the genu and anterior portion of the corpus callosum. Microscopically, the tumour was a glioma.

Case II.—Granuloma compressing the genu of the corpus callosum: history of nine months' duration; choked discs, generalized weakness, mild pyramidal tract signs, apraxic gait and urinary incontinence. Suboccipital decompression with exitus.

C.P., coloured, single, 31 years of age, domestic, was admitted to the service of Dr. Foster Kennedy on October 25, 1932. The history was that since January 1932 she had been suffering from severe 'pressure' headaches, localized to the vertex and radiating to the nape of the neck. The headaches were slightly relieved in the recumbent position. At no time were they associated with nausea or vomiting. In October of the same year she became aware of a tendency to drift to the left when walking. Gradual weakness of the left lower and upper extremity set in and at times the left arm would 'go dead.' Just before admission, she began to suffer from urinary incontinence, occurring both day and night, the urine escaping without her knowledge of any urge to urinate. At no time did she have vomiting, visual disturbances, uncinat phenomena, tinnitus or convulsions. She denied venereal infection.

FIG. 1.—Sagittal section of brain in Case II, showing the two granulomatous nodules. The compression of the corpus callosum is not as evident in the photograph as it actually was in the specimen.

On examination the patient was well nourished. She cooperated well in the examination, though she seemed bewildered by the intricacies of the tests. There was some mental retardation, yet considering her endowment the latent period between a command and her response did not seem unduly prolonged. Spatial and temporal orientation was intact. She showed no delusional or hallucinatory formations. She was apprehensive about the nature of her illness.

The neurological status revealed a bilateral papilloedema with recent and old hæmorrhages. There was no anosmia. Visual fields showed general constriction. The pupils were equal, regular and reacted promptly to light and upon accommodation. There was weakness of the right internal rectus muscle upon convergence. There was no nystagmus. The muscles of mastication, sensation over the face, the corneal reflexes, the volitional and mimetic innervation of the facial muscles, hearing and the movements of the palate and tongue were normal. There was no aphasia.

There was a moderate amount of weakness in all the muscles, but no paralysis. The deep reflexes were equally overactive in the upper and lower extremities. The abdominal reflexes were diminished and the plantars were equivocal.

Non-equilibratory coordination tests were executed fairly well with the upper and lower extremities. There was no defect in the sensory status. There was no astereognosis and no apraxia in the upper extremities.

She could not stand without support. When with assistance she attempted to walk, the gait was bizarre. In walking, she would twist the body to one side, then raise the opposite leg much higher than was necessary or usual and bring it down as though she were uncertain as to where the floor was or how to make contact with the floor.

One week after admission ventriculography was performed. Following this procedure, nystagmus on lateral and upward gaze appeared, and in view of the fact that the ventriculogram showed a marked bilateral hydrocephalus without convincing localization of the third ventricle, a suboccipital exploration was performed, though up to the time of the ventriculography it had been postulated that she had a tumour of the corpus callosum. She died several days after the exploration.

Post-mortem examination was limited to the head. The gross appearance of the brain was that of an internal hydrocephalus, with flattening of the convolutions and distended ventricles. A sagittal section of the brain through the corpus callosum showed two granulomatous nodules, one nodule in the thalamus, the other in the gyrus cinguli immediately above the nodule in the thalamus. The two nodules had compressed the corpus callosum between them (fig. 1). Microscopically, the nodules were typical granulomas, probably luetic in origin.

COMMENT ON CASE I AND CASE II

Case I showed the characteristic picture described by Bristowe. Mental signs came early in the disease and the personality deterioration was profound. Signs of increased intracranial pressure were slight, the cranial nerve involvement was limited to a left central facial paresis, and there were bilateral pyramidal tract signs. Noteworthy in the mental picture was the lack of spontaneity and of all interest in her environment. She came under observation, vegetated for two months and died. One sees such personality alteration in frontal lobe tumours, and in this case there is every reason to believe that the profound mental change was due to extensive frontal lobe destruction. The one feature of this case which is not seen in frontal lobe tumours was the disturbance in locomotion.

By way of contrast with the first case, the second patient showed signs of increased intracranial pressure and no mental or emotional defects save a justifiable anxiety. It is worthy of note that in the second case the pathology was discrete, that there was no encroachment upon the frontal lobes and that there was very real compression of the corpus callosum. The unusual disturbance in locomotion permitted an approximately accurate localization of the pathological process.

In both patients locomotion was affected; in Case I it was impossible even with aid, and in Case II most bizarre with support and impossible

without aid. Both patients had forgotten how to walk even though muscle power was adequate, and there was no incoordination in executing tests in the recumbent position.

Reference to disturbance in station and locomotion in tumours of the corpus callosum is frequent in the literature. Bristowe's fourth patient 'could only walk with assistance and had a tendency to walk on her heels and fall over backwards.' Giese's patient had a tendency to veer to the right when walking. Zingerle's patient was unable to stand or walk. One of Raymond's patients walked as though intoxicated. One of Stern's patients fell slowly backward when attempting to stand. Lippmann's patient could not walk without support. Twelve of the 51 patients reviewed by Lippmann showed an unsteadiness in gait, and significant is his comment that in 12 the tumour was almost confined to the corpus callosum with very little neighbourhood invasion.

As has been indicated, Zingerle was the first to call attention to this disturbance in station and locomotion as a callosal ataxia. But since this disturbance in station and gait, which may vary from inability to walk without assistance to an absolute inability to walk even with aid, as in the first case reported here, is out of all proportion to the mildness of the motor disability and is present in the absence of any demonstrable incoordination when the patient is tested in the recumbent position, it becomes apparent that in callosal lesions one is not dealing with an ataxia at all. Rather it is more accurate to look upon this disturbance in gait as a loss of a specialized function: that is, that individuals with this disturbance do not know how to use their lower limbs for walking. As such, this disturbance in gait is an ideomotor apraxia, comparable to the apraxia in the left upper extremity which is at times demonstrable in lesions of the corpus callosum.

The two patients reported above, when tested in bed, showed good muscle power, recognized changes in the position of the joints, and executed the heel-to-knee, toe-to-object, and finger-to-nose tests without any difficulty, in spite of which they showed a gross disorder in station and locomotion. This is an apraxia of gait which results according to Liepmann from interruption in the interhemispherical conduction of impulses. It may be mentioned that an apraxia of the lower extremities could manifest itself only by such a disturbance in locomotion and gait as found in the patients reported.

Case III.—Degenerative lesion of the middle third of the corpus callosum; history of eleven months' duration; pseudobulbar syndrome with emotional explosiveness; quadriplegia and bilateral pyramidal tract signs; apraxic phenomena.

M.L., age 46, secretary, was admitted to the service of Dr. Foster Kennedy on January 2, 1933. In February 1932 the patient was in an automobile accident. There was no loss of consciousness and no bleeding from the orifices. In March of the same year she began to have difficulty with the left upper extremity. At first, objects which she held under the left arm would fall to the ground; then weakness of the left arm appeared. Towards the end of March she had a left-sided convulsion without loss of consciousness. After a few days in bed she was able to continue at work. Her

employer now noticed that she had become unreliable and that her memory was not as good as it had been. In May she consulted physicians at Clifton Springs for the weakness of the left upper extremity. A small bone flap was turned down over the right parietal cortex, following which there was some recovery in the arm. Four days after returning home she had another left-sided convulsion. The weakness now involved the entire left side and she returned to Clifton Springs. In August a ventriculogram was done. Both the radiograms and the spinal fluid were negative at this time. In November of the same year, weakness of the right side started, first in the leg and then in the arm. Her speech became slurring; she could not control her evacuations; and she became subject to outbursts of tears and laughter. During the day she was very drowsy, though she could not sleep at night.

FIG. 2.—Sagittal section of brain in Case III, showing the degeneration of the middle two-thirds of the corpus callosum.

The past history was irrelevant, except that from the ages of 10 to 30 she had had attacks of migraine. The patient's mother, a sister and brother had similar attacks.

Examination upon admission showed a moderately obese middle-aged woman who was utterly helpless. She cooperated well in the examination. The fundi were negative and the visual fields seemed to be intact. The right pupil was larger than the left. Conjugate movement of the eyes to the right was not as good as to the left. There was a left central facial weakness. The tongue deviated to the left. Speech was muffled and indistinct, typically pseudobulbar. There was no aphasia.

There was a double hemiplegia with greater incidence on the left. She could do nothing for herself, the weakness being such that she had to be turned in bed and had to be fed. The deep reflexes were grossly overactive in the upper and lower extremities

and there was a bilateral Babinski sign, and bilateral Hoffmann sign. Praxis in the upper extremities could not be tested. There were no sensory changes demonstrable. She was incontinent of faeces and urine.

A lumbar puncture showed an initial pressure of 260 mm. of water. There was no block. The fluid was clear, had 4 lymphocytes to the c.cm., a total protein of 90 mgm., and the serology was negative.

She was a difficult patient for a ward. Periodically she became confused and disoriented. At such times she would be noisy and restless. Explosive outbursts of laughter alternated with sudden unpredictable outbursts of weeping. *A week after admission it was noted that the patient did not open her mouth or protrude her tongue, or deviate the eyes when asked to do so. She held food in the mouth for a long time before swallowing. Spontaneous movements of the eyes and jaws were, however, intact.* Her condition remained stationary and she died of intercurrent infection six weeks after admission.

Post-mortem examination of the brain showed the right frontal lobe to be smaller than the left, particularly in the neighbourhood of the longitudinal fissure about the motor region and in front of it. This region felt soft to the touch. There was, in addition, marked softening of the corpus callosum, particularly in the middle third. Sections of the softening showed a tremendous number of large compound granular corpuscles filled with fat. There were also a great many mononuclear cells resembling small and large lymphocytes. Many of the smaller cells contained fat. There was complete destruction of the myelin in the softened areas and no evidence of neuroglia proliferation.

COMMENT ON CASE III

This patient presented a picture of a progressive lesion of the brain with acute episodes resulting in a pseudobulbar syndrome. The double hemiplegia and the emotional lability were profound. Late in the disease the patient did not protrude the tongue, open the mouth or move the eyes when asked to do so. In this connexion it is interesting to point out that in 1909 Mills¹³ reported a case of tumour of the frontal subcortex and callosum, where the man 'could not put out his tongue' when asked to do so, and that Bristowe's¹ third patient 'did not fully open his mouth or protrude his tongue when told to.' In the patient here reported, the failure to put out the tongue, open the mouth or deviate the eyes at a command appeared late in the disease, but at a time when the power of the jaw muscles and the muscles of the tongue was good. It is permissible to look upon these signs as apraxic phenomena resulting from a lesion in the corpus callosum.

CONCLUSION

The syndrome of the corpus callosum described by Bristowe is essentially a syndrome resulting from neighbouring involvement of the frontal subcortex. In all probability mental symptoms, when they do occur in callosal disease, follow the structural disturbance in the frontal lobes. However, the attempt to formulate a mental syndrome specific for the corpus callosum has not

been successful. This is not at all surprising, since the mental reaction of an individual to internal or external trauma is conditioned by the total personality equation of the individual. That being so, mental symptoms in organic disease of the brain depend less upon the location of the pathology than upon the personality 'make-up' of the individual.

Disturbances in praxis when not due to disease of the supramarginal gyrus are specific for corpus callosum pathology. The left-handed apraxia described by Liepmann is well known. In this communication other apraxic phenomena referable to lesions in the corpus callosum are described, such as apraxia of gait and apraxia of mastication and swallowing.

What in this paper has been called an apraxia of gait was described by Zingerle in 1900 as a 'Balken-Ataxie.' This apraxia of gait is an inability to use the lower extremities for walking in the presence of good muscle power and in the absence of any ataxia when the patient is tested in the recumbent position.

Three case histories are presented. In each a diagnosis of corpus callosum disease was made before exitus. The first two showed an apraxia of gait, the last an apraxia of the tongue and jaw muscles, in the course of an intracranial focal lesion. It is the purpose of this paper to emphasize the importance of apraxic phenomena in corpus callosum localization.

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