DEMENTIA INFANTILIS WITH CORTICAL DYSRHYTHMIA

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Dementia and the major psychoses are rare in children. Weygandt, in 1907, introduced the term dementia infantilis to describe cases of progressive mental deterioration of obscure causation, occurring in young children whose previous development had been normal. Since then many of the cases to which this description applies have been found to have a definite etiology in inflammatory, degenerative or demyelinating diseases of the nervous system. By common consent the term has come to be applied to the rather diverse clinical group of cases described originally by Heller (1908). Attempts have been made by Zappert (1921) and others to limit it to a circumscribed syndrome, but, as Voigt (1919) has pointed out, Heller's cases differed considerably from one another and may well have had a varying etiology. It is perhaps better, therefore, to regard dementia infantilis in a wider sense as a clinical grouping from which more circumscribed syndromes will continue to be split off as investigation proceeds. It is one of the objects of this paper to describe a case which conforms clinically to this group and in which electroencephalographic study has yielded a clue to the underlying pathological state. The type of abnormal cortical activity found in this case may well have been present in some of the cases previously described.

The clinical similarity between cases of the infantile dementia group in spite of a varied causation is not remarkable when it is considered that in general the kind of psychological disturbance seen in children in whom there is gross psychotic disturbance depends at least as much upon the stage of mental development at the time it occurs as upon the nature of the etiological process. It is probable for this reason that Heller's cases, whilst alike clinically, were

DEMENTIA INFANTILIS WITH CORTICAL DYSRHYTHMIA 123

not of similar origin. The essential features of these cases were normal early development, onset at the age of about four, early interference with speech, stereotypies and stupor alternating with periods of motor restlessness. All terminated in a profound dementia. Heller specifically mentions the absence of convulsions and of gross abnormalities in the nervous system.

Some years later de Sanctis (1925) described a syndrome with many features in common with Heller's cases. To this he gave the name 'phrenasthenia aparetico-aphasica tardiva.' The essential difference was that this group had a slightly later onset and that some of the cases had convulsions. The clinical features, especially the catatonic behaviour, had much in common, not only with Heller's cases, but with the dementia praecocissima previously described by de Sanctis (1906, 1908) and by Constantini (1908, 1911). This latter group showed most of the features of catatonic schizophrenia, including mutism, stereotypies, echolalia and attacks of impulsiveness alternating with stupor. It is of interest that one of Constantini's cases showing this picture also had convulsions.

Clinically, dementia praecocissima occupies an intermediate position between Heller's dementia, as defined by Zappert (1921) and prepubertal schizophrenia, as described by Potter (1933) and by Lutz (1937). De Sanctis's phrenasthenia appears to have clinical features common to all three conditions, and was thought by that author to have an organic origin.

It is evident that clinical states in children in which catatonia and disturbances of psychomotor tempo accompanied by dementia are the main features are not specific and may indicate the presence of either a functional psychosis or of an organic deterioration of the nervous system. Lange (1933) records two cases which accord with Zappert's criteria but which were almost certainly due to chronic epidemic encephalitis. Cases of Schilder's disease starting elsewhere than in the occipital cortex, and of late amaurotic idiocy in previously normal children may show an exactly similar clinical picture. In other conditions of gross disease of the nervous system differential diagnosis is nearly always possible by reference to physical signs. The non-specificity of catatonic behaviour is further illustrated by its frequent occurrence in aments (Weygandt, 1907) and in the psychoses of aments (Earl, 1934).

There is little mention in the literature of the clinical behaviour of cases of epileptic dementia occurring in children of normal early development. This can only be because most cases occur in mentally defective or neurologically disabled children, and because most cases of dementing epilepsy in children are past puberty before the dementia reaches a severe degree. It seems not improbable that catatonic phenomena can occur in children in the later stages of epileptic dementia as they do at times in adults. Disorders of motility with hyperkinesis alternating with psychomotor retardation are common in the earlier stages, and these features are also present in some of the schizophrenia-like psychoses. Differential diagnosis between epileptic dementia and schizophrenic psychosis may not, therefore, be easy in the absence of a history of convulsions. It seems possible that in the instances recorded by de Sanctis and Constantini, in which fits occurred in a setting of infantile dementia, that the dementia may have been due to an epileptic state in which the fits were not frequent.

The application of electroencephalography has shown that much of the abnormal cortical activity characteristic of epilepsy occurs without demonstrable alteration in the individual's behaviour at the time the record is taken. The essential process in epilepsy is now seen to reside not so much in the convulsions but in the dysrhythmic state which is their background. Dementia is known to be at least as rapid in petit mal as in major epilepsy, and the possibility of epileptic dementia occurring in the total absence of demonstrable fits has now to be envisaged. The clinical features of such a dementia, as has been shown, might well be similar to those of the schizophrenia-like psychoses of children.

In the case to be described a previously normal child developed symptoms of the type described by Heller and by de Sanctis, but in the electroencephalogram showed a profound disturbance of the cortical rhythms. The diagnosis of epileptic dementia was confirmed after over a year by the appearance of actual convulsions.

Case report

The patient is a boy of eight years whose parents first sought advice two years ago because he had ceased in the course of four months to make progress at school, where he was formerly regarded as among the brighter pupils. At that time he spent long periods staring and grimacing and had wandered aimlessly away from school on a number of occasions.

FAMILY HISTORY. The parents are both of above average intelligence and show no gross neurotic traits. In the electroencephalogram, however, both gave abnormal records of the kind described by Lennox, Gibbs and Gibbs (1940) as frequently occurring in the relatives of epileptics. The patient is an only child. The father's brother stammers and has violent attacks of temper. The maternal grandmother was 'peculiar' and her brother was an epileptic defective. The maternal great-grandfather was an epileptic dement and was certified at the age of thirty.

EARLY HISTORY. Normal pregnancy and delivery. Good progress in early months and no serious illness in this period. Began school at five and a half and was regarded by teacher as a particularly bright child. At six could print legibly, read simple phrases, add small sums of money and do simple mental arithmetic (e.g. $14+3$, $9+4$). He could do one-line compositions spontaneously, knew the primary colours and numerous shades and spent much time in drawing. He mixed well with other children, though a little timid, and was regarded as physically and mentally normal for his age.

PRESENT ILLNESS. The onset was gradual. Shortly after his sixth birthday he had a mild attack of whooping-cough and was kept away from school in order not to infect other children. On his return he was in excellent health, but his difficulty in learning was at once apparent. He appeared incapable of assimilating any new knowledge and became inaccurate and unreliable in using methods learned previously. He failed with simple sums and was unable often to remember material learned the day before, though able to sing songs learned a year before at home. He lost the ability to name shades of colour with any reliability, though still able to recognize the primary colours. He would stare vacantly before him and when addressed would repeat the speaker's words.

By the time he was six and a half his teachers regarded him as dull and quite ineducable, though they said he often gave inconsistently good answers which showed that he had been paying full attention though appearing to stare

vacantly. On one occasion, after his class-mistress had apparently been unable to explain a lesson to him, she found him explaining it accurately to some other children. Later he was unable to remember where to hang up his clothes, though knowing that he had to do so. He spent much of his time staring, grimacing and smiling to himself.

He walked out of school on several occasions and took buses to distant places where he was found stranded and unable to account for his actions. At home he alternated between stupor and excitement. In the latter phases he would run about tearing up papers, throwing things out of windows and being over-active, noisy and destructive. At night he had to be tied in bed by his parents, as he frequently got out and slept on the floor. He gradually became completely wet and dirty.

At this point, general measures having no effect, he was referred for a psychiatric opinion and diagnosed as schizophrenic. Attempts were made at a psychotherapeutic approach, but he was quite unco-operative. When seven and a half he suddenly fell down and was unconscious for half an hour. There was no tonic or clonic convulsion. He was sent to a neurologist, who referred him to one of us as a probable case of Schilder's disease. No abnormalities in the nervous system had been found.

His deterioration had thus taken place over a period of eighteen months, and he has now been under observation for a further six months. Apart from the attack of unconsciousness described, no fits, major or minor, had been observed up to the time of admission.

Clinical state on admission. The patient was in a somewhat stuporose condition, but took notice if spoken to sharply and obeyed commands if repeated. His vision was evidently quite good. Echolalia was marked, each sentence usually being repeated once. Echopraxia was present in marked degree and he often showed *flexibilitas cerea*. He made grimaces at intervals and smiled and laughed to himself. At intervals he became over-active, climbing up to the windows, dancing on his bed and singing nursery rhymes. He was incontinent of urine and faeces. He sometimes resisted handling and attempted to bite. For the first month no petit mal attacks were observed, and there were no major convulsions.

On examination of the nervous system no abnormality was found and hearing and vision were proved to be within normal limits. An x-ray of the skull showed no abnormalities, the Wasserman reaction was negative in blood and cerebrospinal fluid, and the blood count was normal. The cerebrospinal fluid showed an initial pressure of 170 mm. on two occasions at three-month intervals. Cells were never over 3, and the protein was not increased.

Initial electroencephalogram findings. The apparatus used consisted of two independent channels of balanced input, special type low-frequency amplifiers recording on the smoked paper of a large Palmer kymograph (Golla, Graham and Walter, 1937). Bipolar leads were taken from silver electrodes with saline pads placed on widely separated areas of the cortex (Walter, 1937). On the first occasion when the electroencephalogram (E.E.G.) was taken, few alpha waves were seen from either hemisphere; these had, however, reached the lowest extreme of adult frequency at 8 per second. Low-amplitude 3 to 4 per second waves and bursts of 2 per second 'spike and wave' complexes were seen from all areas, but the delta wave disturbance had its origin chiefly in the frontal lobes, especially in the right side. The effects of hyperventilation could not be observed, through lack of co-operation, but the inhalation of CO₂ materially reduced the number of petit mal complexes.

Progress and response to treatment. The patient's progress was first observed for three weeks without drug therapy of any kind. Frequent E.E.G.'s showed marked changes from day to day, but these did not correlate to any

RECORD I: 9 per second waves (alpha rhythm) from the right hemisphere, and a run of 3-4 per second waves (delta rhythm) from the left hemisphere.

RECORD II: Delta waves of higher amplitude with random spike and wave complexes from both hemispheres.

RECORD III: Runs of spike and wave complexes with a frequency of 2 per second at a time when petit mal was not clinically discernible.

RECORD IV: The same as record III at slightly greater frequency.

RECORD V: Some weeks later: A run of spike and wave complexes with a frequency of 3 per second during an attack of clinical petit mal.

RECORD VI: 9 per second waves (alpha rhythm) from both hemispheres. The record improved greatly for a short time on anti-convulsants and ammonium chloride. This record was taken a few days before the onset of ketosis. There was no constant relationship between clinical and E.E.G. improvement.

In all the records the upper tracing is from electrodes placed on the right frontal and right occipital regions, and the lower tracing from electrodes placed on the left frontal and left occipital regions. The time marking is in seconds. The amplification is approximately the same in all records. To simplify printing the records have been reproduced here in black lines on white. The originals were white on black.

Records I, II, III and IV were taken prior to treatment with anti-convulsants.

DEMENTIA INFANTILIS WITH CORTICAL DYSRHYTHMIA 127

extent with clinical changes, although a decrease in the amount of 'spike and wave' complexes was possibly associated with a decrease of catatonic behaviour and an increase of spontaneity and responsiveness. Twelve hours' starvation induced a continuous series of 'spike and wave' complexes lasting for over half an hour, but these, which were at only 2 per second, were not associated with clinical petit mal. Although the patient was mute and echopraxic, he would follow the investigator with his eyes, and obey forceful commands. Some weeks later, when clinical petit mal could be observed, the 'spike and wave' complexes reached the normal adult frequency of 3 per second.

Benzedrine sulphate, in doses up to 40 mgm. per day, had no effect electrically, but lessened the hyperkinetic behaviour. Sodium diphenyl hydantoinate (epanutin), in doses up to 0.1 gm. t.d.s., produced a marked improvement in the E.E.G. in the sense that fewer delta waves (less than 6 per second) and an increase of alpha waves (8 to 13 per second) were seen. Petit mal complexes were almost abolished if the patient breathed CO₂ while receiving epanutin. The optimum effective dose of the latter was reached at 0.5 gm. t.d.s. and no further improvement in the E.E.G. resulted from higher dosage. In view of the effect of CO₂, the patient was then given ammonium chloride 20 grains t.d.s., and this produced a marked change in the record, and for minutes at a time no delta waves could be seen from any area. The petit mal complexes were also reduced in amount. There was, perhaps, a slight increase in spontaneity and responsiveness during this period, which was unfortunately terminated after ten days by the onset of ketosis.

The patient was under observation for a period of six months, but the investigation was interrupted on one occasion by scarlet fever, which was followed by a spontaneous clinical improvement and on another by the advent of three major epileptic fits, which were followed by a marked deterioration of his condition. Following these fits the patient had signs of a left hemiplegia, and the left pupil was larger than the right. These signs disappeared in three days. The first true clinical petit mal were observed by the nursing staff about one month after admission, and in such attacks he was unresponsive to any stimuli. Later clinical petit mal was observed during E.E.G. examinations, and it was remarkable that at such times the 'spike and wave' complexes were always of greater frequency.

The impression gained was that the electrical record could be improved by drugs, but only at the increased risk of clinical fits. There was no evidence that lasting benefit in the mental condition or arrest in the progress of the dementia could be obtained.

Discussion

The mode of onset of the condition described and the clinical features present during the first year of its course fully justified at the time the diagnosis of dementia infantilis, if this term is used in the wider sense of Weygandt. In the echolalia, grimacing, stupor with *flexibilitas cerea* and outbursts of aimless and disconnected activity and incongruous laughter there was every reason to regard the clinical state as catatonic, and the underlying process as a schizophrenia-like one. Not only were abnormal physical signs in the nervous system completely absent until major epileptic attacks had occurred, but petit mal attacks could not be observed even at times when the electrical record was showing a continuous series of the 'spike and wave' complexes associated with that condition. Without the aid of electroencephalography the diagnosis of dysrhythmic (i.e. epileptic) dementia could not have been made at all until

the appearance of major or minor attacks, neither of which was observed until eighteen months after the onset.

It seems reasonable to suppose that, apart from the aparetic-aphasic group of de Sanctis in which convulsions were usually present, some of the cases of schizophrenia-like psychosis in childhood (Lay, 1938) with catatonic behaviour described in the literature may actually have been cases of dysrhythmic dementia in which the disturbance of cortical activity was insufficient at any one time to cause outward signs in the form of epileptic attacks, and yet was present for a sufficient time to cause a rapid dementia. In the present instance there can be little doubt as to the diagnosis. Quite apart from the absence of raised intracranial pressure and of abnormal physical signs, except in the period following major fits, the variability of the abnormal waves, the changes produced in them by drugs, and the presence of the typical 'spike and wave' complex practically excludes the possibility of a space-filling lesion or a degenerative process to which the dementia and the electrical disturbances might be secondary. It is generally agreed (Walter, 1938) that this complex is not found in such symptomatic convulsive states. Apart from the unusually long sequences of waves of petit mal type, the records obtained were typical of paroxysmal cortical dysrhythmia (Gibbs, Gibbs and Lennox, 1937) as seen in most cases of idiopathic epilepsy. In view of the finding of these workers (Lennox et al., 1940) that in 35 per cent. of cases of epilepsy the electroencephalographic records of both parents showed abnormalities, it is of interest that in this case both parents, although they were without symptoms or abnormal personality traits, showed unequivocal disturbances of the electrical record.

Many cases of schizophrenia-like psychosis in childhood, like the present case, exhibit hyperkinetic behaviour. Such behaviour is common in epileptic children. It is also seen in the type of child described by Jasper, Solomon and Bradley (1938), Lindsley and Cutts (1940) and Walter (1938), who presents a behaviour problem accompanied by an abnormal electroencephalographic record in the absence of convulsions. The type of record, however, bears little relation to the kind of behaviour abnormality. In the case described there was no clearly demonstrable relationship between the clinical and electrical states, and both hyperkinesis and stupor were accompanied by increased or decreased abnormal activity.

This case shows clearly that dysrhythmia of epileptic type can, in children, be accompanied by a typical catatonic as well as by an hyperkinetic state. Neither catatonia nor hyperkinesis can therefore be regarded as specific for any one kind of underlying pathology. The condition here found, to which the name dementia dysrhythmica infantum may be given, must therefore be added to the number of pathological states which may give rise to the clinical picture of dementia infantilis.

The electrical records were remarkable for the long periods of petit mal activity which were found. Series of 'spike and wave' complexes continued for as long as thirty minutes without alteration, though such complexes were of slower frequency than that seen during petit mal fits. During some of these periods the patient showed no clinical abnormality, but later attacks of petit

DEMENTIA INFANTILIS WITH CORTICAL DYSRHYTHMIA 129

mal were seen in relation to them, and then the 'spike and wave' complexes reached the normal frequency.

Although treatment with sodium diphenyl hydantoinate produced both an improvement in the record and some clinical improvement, there seemed after six months no prospect of the patient's becoming educable.

Summary

A case of dementia with catatonic behaviour starting at the age of six is described, in which a marked cortical dysrhythmia was found on electroencephalographic examination.

The clinical similarity between this case and the schizophrenia-like psychoses of childhood is discussed.

No definite relationship was observed between the abnormalities of electrical rhythm and the clinical state of the patient at the time of taking the records.

Abnormal electrical records were obtained from both parents.

Long series of repeated electrical complexes of petit mal type were recorded. Although these were of uniquely long duration, they were not accompanied by clinical attacks until the frequency of the complexes had risen to 3 per second.

It is suggested that some of the recorded cases of the dementia infantilis group may have been cases of epileptic dementia in which the dysrhythmia was present without outward clinical manifestation. The name dementia dysrhythmica infantum might be applied to this group.

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