SYMPTOMATIC DYSTONIA AS A COMPLICATION OF ENCEPHALOPATHY IN AFRICANS

BY

T. ADEOYE LAMBO, M.D., D.P.M.

*From Aro Hospital for Nervous and Mental Diseases,
Abeokuta, Nigeria, and the Psychiatric Clinic,
University College Hospital, Ibadan, Nigeria*

This paper describes five cases of symptomatic dystonia complicating encephalopathy with mental defect in indigenous Africans. In the original description of his case Thomalla (1918) used the general term "dystonia lenticularis" to include double athetosis, pseudosclerosis (Westphal-Strümpell), Wilson's disease, and torsion spasm. Under this general term I present two cases of torsion spasm, two with double athetosis, and one with Westphal's pseudosclerotic clinical features, all showing mental defect and a clinical mixture of related syndromes. In these patients dystonia, their predominant feature, was symptomatic of a definite illness such as encephalitis lethargica or other affection of the basal ganglia; unlike Ziehen-Oppenheim disease, dystonia musculorum deformans, which is of unknown aetiology.

Dystonia is not uncommon as the presenting symptom in some cases of Wilson's disease, and C. de Lange in 1945 (quoted by Reese, 1946), stated that torsion spasm has also been encountered in a case of tuberculoma of the fronto-parietal region. But Wilson's disease and torsion dystonia have never been described outside Europe and America, and certainly not in indigenous Africans.

Torsion spasm is a condition of comparative rarity. So far as I can trace, some 62 cases have been recorded, mostly in Germany and America, since Schwalbe (1908) and Ziehen (1911) first drew attention to it. Blandy (1920) described the first case in England, admitted to the National Hospital, Queen Square, London. However, with the exception of Wimmer (1921), who described the mental condition of his patient as "very slightly impaired," and of Taylor (1920), who noted "progressive mental enfeeblement" in his Case 2, all authors have failed to mention the coexistence of mental defect and dystonia lenticularis. Brain (1955) stated that psychological changes are absent, and Wechsler (1947) observed that mentality is normal. Also Purves-Stewart and Worster-Drought (1952) pointed out that there is no mental impairment.

Most of the earlier cases and opinions supported a conception of a nosological entity, consequent upon familial or racial degenerative process, *ab origine* in the corpus striatum. But in this connexion Meyer and Cook's (1936) cases are important. They described two personal cases of status marmoratus (*état marbré*), both of which were said to be the subjects of a severe degree of mental defect and quadriplegia from infancy. Their first patient suffered also from generalized fits, torsion dystonia, and choreo-athetosis. These observations favour the conception of multiple aetiological factors causing similar syndromes rather than the existence of separate nosological entities.

Four of the five cases presented here were post-encephalitic and had a noteworthy degree of mental defect. The existence of an infective process is doubtful

in the last case, but the clinical features were similar to those of myoclonic encephalopathy. The first case is typical and was characterized by bizarre and grotesque contortions of the trunk and limbs, a marked degree of motor agitation, and alternation of hypotonicity and hypertonicity.

Torsion Spasm

Case 1

A girl aged 5 came under observation in April, 1956. She was the youngest of six children, three of whom died in infancy. The father was aged 39 and the mother 29 at the time the patient was born. The family history revealed no familial or hereditary nervous or mental disease. The grandparents are alive and well.

The patient was normal at birth. In 1954 she had a febrile illness which was followed by a localized rash on the

FIG. 1.—Showing the opisthotonos in Case 1.

right side of the abdomen and on the lateral side of the right leg (? herpes zoster). The rash, which eventually coalesced, first contained clear fluid which later became purulent. The child was described as "very ill" at this stage, lying "immobile and unconscious."

When she got better she had lost most of the previously acquired functions: she could not stand or walk, speech was lost, and she could not understand spoken words. After an interval of a year it became apparent that the child's mental development was retarded; at the same time her movements had become automatic. Eighteen months from the onset of her febrile illness she started to exhibit opisthotonos with complex, purposeless, and predominantly tonic movements (Fig. 1).

When lying she made silly, bizarre, and rather grotesque wriggling movements of the body and limbs with spasmodic twistings of the arm. When efforts were made to lift her up or get her to walk (Fig. 2) she showed characteristic hypertonicity and a curious torsion of the body. The whole picture, which fluctuated and varied from minute to minute, presented at irregular intervals an admixture of athetosis, spasmus mobilis, and chorea, and yet there was evidence of differentiation from these conditions. Definite physical signs included irregular pupils with relative dilatation of the right pupil. She had squint of the right eye, and on passive manipulation of the limbs tonicoclonic movements were felt.

She showed a marked degree of emotional instability, had temper tantrums, and made odd unintelligible noises. One feels that her mental development had been arrested. The condition at present seems stationary.

FIG. 2.—Case 1. Showing characteristic hypertonicity and torsion of the body in the effort to walk.

Case 2

This case showed features of paralysis agitans in addition to peculiar spasmodic twisting of the trunk, extension and pronation of the left arm, and copious salivation.

This patient was a boy of 6½ years with a negative family history. His mother was aged 30 and his father 37. His early development was uneventful even though his birth was unduly prolonged and difficult. He walked and talked before he was 16 months.

Three years previously the patient had a febrile illness with a generalized diffuse rash ("ekitigbi," measles). When he seemed to be getting better he suddenly became worse. He was stuporous for two days, during which he could be aroused only with great difficulty, and even then for only

FIG. 3.—Case 2. Showing characteristic posture.

a short while. Shortly afterwards, when he seemed to have recovered, he started to show speech difficulty, grimacing, and intellectual retardation.

When first seen he was like a young child of 3, with peculiar involuntary movements, a facile smile, and a spasmodic grin. His posture was characteristic, with a moderate degree of lordosis and irregular twistings of the spine, yet his occasional posture of flexion and his rigidity resembled those of paralysis agitans. The left arm was frequently though transiently extended and hyperpronated at one time and almost normal at another (see Fig. 3), and the involuntary movements were tic-like, with great motor restlessness, resembling choreo-athetoid movements of Oppenheim and Vogt, and of Freund and Vogt. Hypertonicity seemed to be the predominant tone.

Although he had lost his speech after the illness, by the time he was seen he was able to utter some words occasionally with comparative ease but indistinctly. Tendon reflexes were difficult to elicit but seemed to be normal. Both pupils showed miosis and there was squint in both eyes. His head was a peculiar shape.

Both this case and Case 1 were still under observation at the time of writing.

Double Athetosis

In the following cases double athetosis, with poor mentality associated with pyramidal lesions, was the most predominant aspect of the clinical picture. Distal muscles of the extremities showed constant involuntary movements. The combination of the athetoid movements with associated movements, and the preponderance of severe motor agitation over paralytic symptoms, were noticeable features in Case 3. Both Cases 3 and 4 showed variable tonic muscular spasms.

Case 3

A Yoruba boy aged 14 was seen at Aro Hospital Clinic in November, 1955. Four years previously he had complained of vague rheumatic pains, fever, and sore throat; these were followed by a short period of delirium. A state of profound prostration ensued, finally giving place to a brief period of insomnia. Soon afterwards he was said to have had what seemed to be myoclonic convulsions, especially of the abdomino-diaphragmatic region. He appeared to have recovered from this condition, but after a lapse of three months he complained of slight fever and was noticed to be drowsy for about two days. Later he was dragging his leg, and the left arm was described as "weak."

On his admission to hospital there were definite physical signs of a positive Babinski reflex on the left, slight tremor, and athetoid movements of both hands, more marked in the left hand. His articulation was a bit impaired, and though he was alert and amiable he showed definite dullness intellectually. He was simple and euphoric. The liver was slightly palpable.

The most prominent features here were the severe tonic muscular spasms, hyperaesthesia of the soles of both feet, moderate twisting of the pelvis, and lordosis. For a long time the patient was obviously incapacitated by the extreme spasticity in both legs and had to be carried in and out of the clinic. Later, his condition improved and he could walk about but with some difficulty. The athetoid hands also showed marked incoordination and extension-hyperpronation of the distal portion of both arms. The face, head, and neck escaped. There was no opisthotonos and no head retraction. A marked degree of athetosis predominated. His condition seems to be improving and the pelvic contortions are now less marked.

Case 4

The next case is one of progressive dementia with behaviour of a low-grade mental defective. Perseveration is present. Myostatic disturbance is a prominent feature. She is untestable.

A girl aged 9 was referred for "nervousness and fidgeting." She was a second child of the mother and had a normal early history. Her father is aged 43 and her mother 27. Another child of the family (by another wife), a half-brother to the patient, has been admitted to Aro suffering from organic psychosis. The father is a nervous, unstable person. The mother, who is mentally dull, has a young baby of 9 months.

Nothing noteworthy happened to the patient until a year ago. She was then supposed to have a mild attack of smallpox, and a week after the onset of the illness she became very excited for about three days, during which she was visually hallucinated. She was taken to the native medicine-man in Ijebu, after whose treatment the child "became normal again." A few months later she had a severe convulsion, just "out of the blue," and from that time, according to her mother, she began to act like an imbecile. She had to be dressed, washed, and her toilet habits had become lax.

On admission she was dull, mute, apathetic, and had no interest in anything around her. She was very dependent on her mother. There was bilateral athetosis, which was more marked in the left than in the right hand (Fig. 4) and which her mother said was the latest symptom to appear and the most troublesome. There were residual signs of hemiparesis on the right side.

Careful analysis of the child's behaviour—for example, withdrawn regressed condition and complete lack of emotional rapport—has occasionally aroused one's suspicion with regard to the presence of psychosis coexisting with bilateral athetosis. Her present state is one of deterioration.

FIG. 4.—Case 3: bilateral athetosis.

Pseudosclerosis

Pseudosclerosis of Strümpell (1898) as a nosological entity has been greatly disputed. The better-known authors on the subject have failed to agree and the present trend is to consider this condition as a subgroup of the syndrome of hepato-lenticular degeneration with all its clinical variants.

The question remains whether the same aetiological factors apply.

The following case was referred to us first diagnosed as neurotic and later as schizophrenic. At this stage the clinical picture, which in some ways reminded one of the early phase of disseminated sclerosis, was later superseded by definite signs and symptoms of striatal disease and a widespread organic damage to the brain.

Case 5

An illiterate Yoruba adult aged 32 was brought in by his mother in May, 1955. She complained of his generalized tremor, progressive dementia, and inability to walk properly; also that he wandered aimlessly about the room most nights and slept the best part of the day. According to his mother his illness started a year before his admission, "the day he shook hands with a friend with whom there was a long-standing quarrel." The right hand started to shake slightly without any progression for three months, after which it spread to the entire body, including the face and head. At this time he found it difficult to sleep and for several weeks lay awake all night.

The difficulty in speech and the signs of mental deterioration did not come into the picture until five months later. The mental symptoms, which included occasional hallucinatory experiences, memory defect for recent and remote events, disorientation for time, place, and person, appeared insidiously and have since dominated the somatic component.

The mother, aged 55, has not been well for several years. There was "nothing significantly wrong" except for a feeling of being unwell. She had corneal changes in her left eye; otherwise physical examination revealed nothing abnormal. The father died eight years ago of a protracted illness of which the mother gave such a poor description, including her own interpretation of the cause, that nothing important could be adduced from it. The patient is the eldest of three children. The other two siblings—a boy of 12 and a girl of 27 years—are alive and well. No family history of nervous and mental illness.

On admission the patient showed generalized tremor, with choreiform twittings all over the body, and these were exaggerated when he attempted to perform any act like shaking hands—that is, marked intention tremor. He could understand spoken words after a long repetition, but was typically staccato in speech when he made an attempt to answer questions. He was disoriented for time, place, and person, and showed complete memory defect for recent and past events.

He was ataxic in all limbs and generally sluggish. The area of hepatic dullness was greatly diminished. There were no physical signs of pyramidal involvement. Although his mother gave a history of mood fluctuation at the onset of the illness, he was apathetic on admission and has since remained so. His face was expressionless and mask-like, and there was the characteristic extreme slowness of movements and increased muscle tone.

His condition is one of progressive deterioration of physical and mental power. Whilst certain features of the physical picture approximate disseminated sclerosis and the mental picture resembles that of organic psychosis, there are certain qualities to the two components of the clinical picture which resist any attempt to explain one component as typical or true and its association with the other as one of essential concomitance. Clinical evaluation of the dyskinetic phenomena, however, places the total picture in the category of lenticular degeneration of a fairly rapid onset (probably toxic-infective), with a pronounced degree of progressive dementia. In spite of the genetic association of pseudosclerosis with hepato-lenticular degeneration, the history of "a slight febrile illness" is regarded here as being more than casual. It would not be unlikely if this proved to be a slight encephalitic attack, as evidenced by its clinical residuum. Campbell and Morse (1924) described a case (including necropsy findings) of pseudosclerosis following erysipelas. These authors also quoted A. Westphal and

Sioli, who in 1922 produced evidence of the infective origin of the pseudosclerotic process. In their case the syndrome followed what was probably an encephalitic attack (cf. Denny-Brown, 1946).

Discussion

In all five cases clinical and laboratory findings support the exclusion of "functional" and organic psychoses, especially juvenile G.P.I. (slight clinical evidence of organic psychosis in Case 4 as indicated above).

It seems that variations in the sequelae of encephalitic process are possibly due to individual variations in the relative susceptibility of different levels of the cerebrospinal axis, and it can be suggested that racial or familial vulnerability and/or previous disease may be an important factor in the focal predominance of the underlying pathogenetic factor.

The clinical interrelationship of these syndromes is of great theoretical interest, especially when one considers the clinical admixtures, in most cases manifesting features of pseudosclerosis, spastic pseudobulbar palsy with contractures and choreo-athetoid movements of Oppenheim and Vogt, and Freund and Vogt, Oppenheim's dystonia musculorum deformans, double athetosis, von Bechterew's hemitonia apoplectica, status marmoratus, Hunt's striatal syndrome, and other ill-defined clinical states which Strümpell (1913) thinks belong to his amyostatic syndrome—for example, the paramyoclonic type of lethargic encephalitis. The relatedness of the clinical syndromes is also reflected in the histopathology of the lesions of the basal ganglia, and earlier investigators placed emphasis upon the manifold pathological findings which may be encountered in the study of the lesions of the basal ganglia.

All the five cases presented here show the "amyostatic symptom-complex" of Strümpell as a sequel to encephalopathy. The maintenance of posture was impaired in all our cases. Involuntary activities varied, showing features of alternating tremor or of choreoid, athetoid, or dystonic movements. Critical analysis of the symptomatology and repeated examinations revealed the fact that although these clinical states are variable, mixed, and could be better understood as a conglomeration of syndromes of the basal ganglia rather than clear-cut disease entities, acquired illness could give rise to clinical states which are indistinguishable in certain respects from those whose aetiological factors are associated with familial and/or racial predilection. There is no doubt that this condition of dystonia musculorum deformans, originally described by Ziehen and Oppenheim, is very rare when compared with the number of patients showing dystonia as a symptom of brain disease.

In considering the constitutional aspect of the clinical syndromes, this material is not large enough, nor could it be sufficiently carefully studied in this respect for one to come to definite conclusions. Kehrer (1928) and Guttmann (1936) have, however, produced evidence of constitutional factors in chorea and athetosis. Kehrer coined the name "choreopathic personalities" for the abnormal individuals in the family of these patients.

It may also be cautiously assumed that there is in these patients a certain *somatic vulnerability* which may be, in part, an expression of genetic-constitutional predisposition to dystonic disease. Also, the same factor may be of aetiological significance in the true dystonia musculorum deformans of unknown aetiology.

Summary

Five cases of symptomatic dystonia, complicating encephalopathy with mental defect, have been described in native Africans and the clinical similarity between these related syndromes has been emphasized. The youngest patient (aged 5) with dystonia musculorum deformans of acquired origin has also been added to the literature on the subject.

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TREATMENT OF A CASE OF COMPULSIVE SWEARING

BY

RICHARD P. MICHAEL,* M.B., B.S., D.P.M.

Registrar, the Maudsley Hospital, London

Coprolalia as a symptom occurs in the major psychoses leading to personality deterioration and disintegration. Compulsive utterances of an obscene nature are, however, an unusual occurrence where the personality remains intact, and generally form part of a wider clinical syndrome originally described by Itard (1825), mentioned by Trousseau (1867), and delineated by Gilles de la Tourette in 1885. The essential features of the syndrome now bearing the latter's name include a childhood onset with multiple motor tics, a vocal tic developing later into vulgar utterances, and sometimes also a compulsion to repeat both the words and the actions of others. Frequently the motor component is impulsive and very bizarre, bearing a relationship to the latah which used to be seen in the Malay peninsula (O'Brien, 1883). But it is usually the uncontrolled audible obscenities that force the sufferer into an increasing social isolation, providing, some writers feel, the basis for an eventual psychotic withdrawal.

These uncommon cases are acknowledged to be extremely difficult to treat; the lack of success may be judged from the multiplicity of remedies tried. The physical restraint, isolation, skimmed-milk baths, and static-electricity treatments of an earlier era have given place to modern psychotherapeutic procedures, yet there are few well-authenticated cases in the literature that have lost their tics. None of the patients of Gilles de la Tourette, five of whom were followed over some years, showed any sustained improvement. Itard's old woman, who was seen by Charcot, began ticking as a child of 7 and died at 85, still pouring forth the grossest obscenities. Guinon (1886) and the other older writers

(Prince, 1906; Gilles de la Tourette, 1899) were uniformly gloomy upon the likelihood of recovery. Kinnier Wilson (1955) gives the syndrome a sinister outlook, characteristically "ending in insanity," and though accurate information on prognosis is sparse most authorities describe a progressive worsening of symptoms. One of the three cases collected by Ascher (1949) from the Phipps Clinic developed schizophrenia; while his two personal cases did not improve with psychotherapy. Of Heuscher's (1953) three cases one also became schizophrenic. Mahler and Rangell (1943) had a boy of 11 under continuous psychotherapy for 30 months, and agreed with the previous authorities that the prognosis was unfavourable.

The following case is described because of the satisfactory response to therapy.

Case Report

A salesman aged 28 was admitted to hospital on June 11, 1954, and discharged on November 26, 1954.

His father, aged 61, was healthy, a printer in charge of a composing department. He was a quiet man, even-tempered, and somewhat ineffectual, who made few demands on the family. The mother, aged 56, was also healthy, but house-proud, hot-tempered, strong-willed, and close to the patient. There was one healthy married sister aged 24. There were no familial illnesses and no history of chorea or tics.

Personal History

The patient's birth on August 2, 1926, was normal, but he was over one month premature and under weight. There were initial feeding difficulties, and the frenulum of the tongue required freeing at 2 weeks. He was concussed at 2, had measles and chicken-pox at 2½, and pneumonia at 4. (These incidents are mentioned because of the possible aetiological importance of brain damage.) Though his speech was retarded the other milestones were normal. A timid shy child with temper tantrums, night terrors, and many minor compulsive rituals, he attended elementary school at 5, secondary school at 12, leaving at 14½ to work in the father's print-shop for two years before becoming a hat salesman. At 18 he was called up for the Army (B1) and served for two years in Egypt before being discharged with the rank of sergeant at the age of 22. He then joined his present firm and has since risen progressively to the position of a senior salesman who covers an important territory.

Play with his sister and other children continued normally until the age of 9, but from that time on he made attachments with boys only. Masturbation began before 15 and continues. At 18, when in the Army, he became the active partner in anal intercourse, and his relationships have remained exclusively homosexual. There is a well-marked madonna-prostitute split in his love objects. There are those men whom he admires and feels love for and also, as he says, the casual pick-ups with whom he has intercourse. Always a good worker, he is money-conscious, ambitious, and successful, yet anxious and tied to the home. Both selfish and sensitive, his manner is ingratiating with superiors, for he is afraid of "the big man in business."

Present Illness

At the age of 7 he developed tics originally thought to be St. Vitus's dance but later diagnosed at Great Ormond Street as habit spasms. The wriggling of his body, facial contortions, and sensitivity to noise subsided gradually over the next two years. But at 9 he began making noises, and these soon developed into the swearing which persisted with such distress to his family until he was 13. Reading *Tom Sawyer* made a deep impression on him and he began to say "huckleberry fuckleberry fuck fuck." He was reported to his teacher for using "dirty words" and scolded severely at home, but the pronouncements had developed an

*Holder of a Medical Research Council Fellowship in Clinical Research.