THE STIFF-MAN SYNDROME

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In 1956 Moersch and Woltman[1] first recorded a state of 'progressive fluctuating rigidity and spasm', for which they coined the term 'stiff-man syndrome', signifying that the aetiology and pathogenesis of the condition were unknown. At the time of writing only 16 cases have been described. It is suggested that the case under consideration falls into this syndrome, although there are a number of features at variance with the published cases.

CASE REPORT

G.H., a Coloured male aged 24 years, was involved in a motor-car accident on 7 December 1957 and sustained an injury to the right leg. There was no head injury or loss of consciousness.

He was taken to the New Somerset Hospital, Cape Town, where the diagnosis of a compound fracture of the right tibia and fibula was made. After the administration of 1,500 i.u. of tetanus antitoxin, and intramuscular penicillin and streptomycin, the patient was taken to the theatre and an open reduction was performed under general anaesthesia. He remained in the ward until 12 March 1958 when he was discharged. During that time his leg was in plaster-of-paris, he felt well, and for the latter part of his stay he was able to use crutches. He then attended as an out-patient and had no complaints.

On 18 April 1958 the patient was readmitted to the surgical ward for a change of plaster and under general anaesthesia the wound was inspected and found to be satisfactory, and the plaster-of-paris was reapplied. He was discharged on 24 April. On the day of discharge he became aware of a dull ache in the mid-thoracic region of the spine. The pain was persistent, but did not radiate and had no specific aggravating or relieving factors. Analgesics were of no help and the patient had difficulty in sleeping at night. The following day he noticed a generalized body stiffness, and one day later he began 'jerking'. This initially involved the right leg, but over a period of 24 hours the whole body suffered agonizing spasms. These spasms were precipitated by movement or a sudden jerk, although they would also occur spontaneously while he was lying still in bed. They were unaffected by any form of sedation. There were no spasms during sleep. After 10 days the patient observed for the first time that his jaw felt stiff; he had difficulty in opening his jaw, and chewing food and swallowing required considerable effort. At this stage he was seen at another hospital by a neurologist, who agreed that trismus was present, but felt that the whole state was unlike tetanus and made a diagnosis of hysteria. The patient was given 'flexin' largactil, phenobarbitone sodium, and phenergan—all with no immediate effect. Nevertheless, after 3½ weeks the spasms lessened in severity and soon ceased completely.

He enjoyed perfect health for the next 2 months, attending the out-patient department at the New Somerset Hospital for periodic changes of the plaster, and eventually he was able to use a weight-bearing plaster. During this period he had no backache, no rigidity and no spasms.

On 25 July 1958 the plaster-of-paris was again changed, no anaesthetic being administered. Seven days later the backache recurred. Although the character of the pain was identical with that experienced initially, the site had altered; he felt pain maximally in the lumbar region. The following day he became 'stiff all over', and one day later generalized spasms started. These were again unbearably painful and persisted unchanged until the time of readmission to the New Somerset Hospital on 14 August 1958 after 10 days of spasms followed by 2 days of trismus. He was unable to swallow because he felt that this induced a spasm. During the 2 days before admission he also had difficulty in breathing. He described this as 'not being able to get air into my lungs—everything feels tight and the harder I breathe the less air I seem to have'. As the depth of respiration increased everything became 'blurred and distant' and he passed into a state of unconsciousness which lasted for 10 minutes. There were no epileptic jerkings. On recovery of consciousness he felt no ill-effects. These 'attacks' were repeated twice on that night, and once again on the next night, the night before admission. For a few days before admission he had had a slight cough, productive of white sputum. His appetite was good, and the bowels regular, while there had been no disturbance of micturition. He did not complain of any paraesthesia. Further interrogation was uninformative.

The past history was not significant. The patient was very reticent about his personal life. After passing standard 9, he had left school after a difference of opinion with his family. He then embarked upon a nursing career, but left after 3 years of training because he felt 'there was no future in it'. He said that he was happy in his occupation as a motor-car driver. He appeared to have many friends and was engaged to be married.

On examination (14 August) the patient, a young, well-nourished Cape Coloured man, was anxious, hyperventilating considerably, and a prominent feature was profuse sweating, which was generalized, but most marked over the forehead and face. There was no rash or clubbing present. The right leg was encased in a below-the-knee plaster-of-paris. Three enlarged discrete, tender lymph glands were palpated in the right groin. He was afebrile. All the peripheral pulses were palpable, and there was no evidence of cardiac failure. He had a mild cough productive of white sputum. Pulse, 110 beats per minute, regular in sinus rhythm. Blood pressure 120/80 mm. Hg (but during a spasm it rose to 145/105 mm. Hg). There was no cardiomegaly, and auscultation did not reveal any abnormality. The respiratory system was normal. Abdominal examination was rendered difficult because of spasms, but during intervals it was possible to make a satisfactory assessment, and no abnormality was detected.

The patient was fully conscious and alert. He was disgruntled and truculent and resented being questioned or examined, frequently remarking that he wished to go home. Every 1-2 minutes he experienced generalized spasms involving the jaw and face—a typical risus sardonicus facies resulting—and the neck, trunk and both upper and lower limbs. At such times he developed opisthotonus and the toes and feet were plantar-flexed, while the big toes were dorsiflexed. These spasms lasted at the most from 3 to 4 seconds and were intensely painful to the patient. During a spasm the patient was unable to talk and breathed with difficulty, but was able to move the eyes about. All the muscles of the body contracted so that they stood out prominently. The spasms were not all of the same intensity.

In between spasms he was able to talk with comfort, and he suffered no pain. There was marked permanent rigidity of the neck, shoulder girdle and spinal musculature, so much so that passive movement of the neck was impossible. Moderate trismus was present. The arms and the abdomen showed some increase of muscle tone, but not to the same extent as the neck, while that of the latissimus dorsi and lower limb muscles was normal. Any attempt to move a limb passively or to palpate the abdomen induced a spasm, but during the course of the examination this tendency became less obvious until eventually the patient achieved moderate relaxation of the abdomen. There was no muscle tenderness. The spasms appeared to occur spontaneously, but were also induced by sudden movement, both voluntary and passive, and by any jar or loud noise. Although trismus was present, no difficulty in taking food or swallowing was present and no spasms were precipitated.

For the rest, neurological examination was normal, motor power being good, reflex activity of normal degree, and sensation intact. The optic fundi were normal. Slight tenderness was present over the lower thoracic and the lumbar spines. The urine was normal in all respects. Haemoglobin 11 g.%; WBC 9,000 per c.mm.; ESR 6 mm. in the first hour (Westergren). A blood smear was normal. The blood WR was negative. All investigations of the serum chemistry were within normal limits: Blood urea 44 mg.%; serum albumin 5·2 g.%; serum globulin 3·3 g.%; serum calcium 9·8 mg.%; serum sodium 137 mEq./litre; serum potassium 3·7 mEq./litre; serum chloride 102 mEq./litre; serum CO_2 53 vol.%; thymol turbidity 1; zinc turbidity 12; serum v. d. Bergh negative; serum bilirubin 0·5 mg.%; serum inorganic phosphorus 3·8 mg.%. X-rays of the chest and the spine were normal. The patient would not allow a lumbar puncture to be performed.

Course

While in the casualty department the patient was given 10 c.c. of calcium gluconate intravenously with no benefit. In the ward 'equinal' and phenobarbitone sodium were administered. The following day the patient was less anxious and more managable and, though the rigidity was unchanged, the impression formed was that the spasms were slightly less frequent. The patient had no spasms while asleep. During the course of the day a fever of 100°F developed, the sputum was noted to be purulent, and the patient was given parenteral penicillin (2 million units per day) and streptomycin (1 g. per day).

At 1 a.m. on 17 August he complained of difficulty in breathing and was observed to be hyperventilating. The spasms appeared to be slightly more marked in severity, but not in duration. Soon loss of consciousness followed, lasting for 15 minutes, during which time the patient was pale, breathing deeply, with dilated pupils. Muscle tone was completely flaccid and he had no spasms. Recovery occurred fairly rapidly—the patient sat up and asked for water, the whole episode lasting about half an hour. During the remainder of the day his condition was unaltered, but in the early hours (2.45 a.m.) of 18 August 1958 the tightness in the chest, hyperventilation and loss of consciousness were repeated. However, this time the patient failed to recover: he remained

pale, flaccid and breathing deeply and after about 5 minutes his heart stopped beating.

Post-mortem Examination (Dr. N. Woolf) did not reveal the cause of death. Both the macroscopic and microscopic examination of the organs showed no abnormality. In particular the histology of the brain and muscles was normal, the latter only showing slight granular degeneration and atrophy in parts.

DISCUSSION

As described by Moersch and Woltman,[1] the stiff-man syndrome consists of a relentlessly progressive stiffness of the muscles. While the trunk muscles are usually affected initially, stiffness may commence in the neck or limbs. Eventually all muscles are involved, the face, hands and feet being minimally affected. Superimposed on the affected groups there are severe and painful spasms. The muscular rigidity and spasms fluctuate in intensity, but over a period of years become progressively more severe. The duration of symptoms varies from 6 to 15 years. The case described by Asher suffered with rigidity and spasms for 15 years before succumbing.[2] The affected muscles are in a state of permanent and excessive contraction, which is painless. Spasms are an exacerbation of this state but are excessively painful.[3] A spasm may occur spontaneously or be precipitated by a sudden jar or movement. Moersch and Woltman note that the degree of stiffness varies from time to time and mention that two patients stated that if they moved gradually and slowly the muscular tightness would wear off for brief periods. During sleep and under thiopentone anaesthesia the affected muscles appear normal.[3] Both Asher[2] and Price and Allott[3] comment on the excessive sweating, and Asher's patient also had bouts of hyperpnoea. Of further interest is the relationship that both Price and Allott's case and this patient show to orthopaedic procedures.

Apart from rigidity and spasms, clinical examination is unrevealing, and side-room and laboratory investigations, including electromyography, have all been normal, with two exceptions, viz: (*a*) 4 of Moersch and Woltman's cases and Asher's case showed a trace of glucose in the urine, and (*b*) Price and Allott demonstrated an abnormality of phosphorus metabolism—when active carbohydrate metabolism took place the level of inorganic phosphate rose (normally it should fall). The significance of these findings is still obscure. The aetiology of the syndrome is unknown, but suggestions have been made that it is a form of myopathy[2] or a progressive metabolic disorder.[3] Only one other necropsy report is available, and the muscle changes found were non-specific, so that there is as yet no sound basis for relating this syndrome to the group of myopathic diseases.

The case here reported differs from the stiff-man syndrome as hitherto described in a number of ways, viz: (*a*) the short course of the illness, (*b*) the presence of trismus, and (*c*) the fact that muscles which were not actually stiffened or rigid were also involved in the spasm. Naturally the question of tetanus arises. There are a number of difficulties in entertaining this diagnosis. The man was given tetanus antitoxin at the time of the injury and in a recent series it has been shown that the incidence of fatal tetanus after prohylaxis is very low.[4] Secondly, this man was having generalized painful spasms at a time when there was no generalized muscular rigidity. Then again, only after 10 days of spasms did he develop trismus. Cases of tetanus invariably start with stiffness of the jaw before the rigidity and spasms develop.[5] His mode of death too, was unlike that of the usual fatal tetanus, in which the patient frequently shows terminal mental clouding, and dies either from pneumonia, asphyxia, or hyperpyrexia. Both recurrent and relapsed tetanus are very rare conditions.[6, 7]

Strychnine poisoning was considered, but in this condition there is complete relaxation between spasms, no trismus is present, and the distal parts of the extremities are involved to a greater degree.[5, 8] Hysteria was one of the diagnoses made in this case, and many of the patients suffering from this syndrome have been labelled as 'functional'. While the condition is clearly not fundamentally a psychiatric one, there may well be a hysterical overlay in many of the patients. Dystonia musculorum deformans and tetany were also considered, but readily excluded; the former because the spasms were not associated with torsion movements, the latter because no increase in excitablity of the myoneurone was demonstrated by the usual tests, and at no time did he show *la main d'accoucheur*.

Every report of this syndrome stresses the uniform failure of all varieties of medication to arrest the course of the disease or to relieve the rigidity and spasms—phenobarbitone, chlorpromazine, morphine, pethidine, methenesin and many other drugs have been tried unsuccessfully. The cause of death in this patient is still obscure; as witnessed and from post-mortem evidence he did not die from asphyxia. There were no abnormal findings at autopsy and the muscles were normal. Possible causes of death include exhaustion, and fatal carotid-sinus syncope following upon compression of the sinus during a spasm.

SUMMARY

A fatal case of fluctuating muscle rigidity and spasms is reported. It is suggested that the case is to be included in the 'stiff-man syndrome', which is a condition of fluctuating progressive stiffness of all the muscles, with superimposed painful muscular spasms and with profuse sweating as an added feature.

There are, however, differences from the syndrome as described, the essential ones being the presence of trismus, and the accelerated course followed in this case. This patient died 4 months after the recorded date of onset; the average duration of symptoms in this syndrome as described is 9 years.

Tetanus, both recurrent and relapsing, has been discussed, and the reasons for not considering this as the diagnosis are stated.

The aetiology of this syndrome is completely unknown, and it is refractive to all forms of therapy. Post-mortem studies failed to reveal either the basis of the disordered muscle function or the cause of death.

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