

THE STIFF - MAN SYNDROME: A CASE REPORT

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SUMMARY

A patient with clinical and electrophysiological criteria for the diagnosis of Stiff-man syndrome is reported. The disease is very rare and characterised by progressive stiffness and painful spasm affecting principally the proximal limb musculature with an electromyographic pattern of continuous motor unit activity at rest. The etiological factors of the disease are not known and various therapeutic modalities have been suggested.

Key words: Stiff-man Syndrome, Electromyography, Diazepam.

INTRODUCTION

The stiff-man syndrome is a very rare disorder characterised by painful spasm and rigidity of axial muscles associated with continuous motor unit activity on electromyography. It was first described in 1956 and since then few reports have been presented and discussed about the cause, pathogenesis and treatment (1-4). The exact cause of the disease is not clear but recently some pathological entities have been blamed (4-7). Because the disease shows some atypical features, it generally remains undiagnosed or misdiagnosed.

At diagnosis electromyographic data plays very important role. In the treatment continuous motor unit activity can be overcome by some therapeutic medications but diazepam is the most effective one (1).

CASE REPORT

A 59-year-old woman was admitted to hospital because of difficulty in walking. She had painful spasm of all muscles and stiffness, particularly affecting the abdomen, lower back, neck, jaw and thighs. She was hospitalized for diagnostic investigation in September, 1989. She had a normal

personal medical and social history. The illness began three weeks before the admission and gradually progressed. She was first seen by a general practitioner who referred her to our clinic.

On admission, the patient was in poor condition with anorexia, poor feeding, and fever.

On general examination, mental status was normal. She had restricted jaw rigidity. There was stiffness in the muscles of the neck, abdomen, the low back area and painful spasms of the body and nearly all limb muscles. She was unable to walk and the spasm were aggravated by noxious stimuli such as touching and passive motion.

On neurological examination, cranial and peripheric findings were normal except that all deep tendon reflexes were increased and abdominal reflex was absent.

X-ray examination of the skull and computed tomography of the brain and myelography with a contrast medium were normal but X ray of the spine showed lumbar hyperlordosis.

On laboratory examination, lumbar puncture showed normal cerebro-spinal fluid (CSF) pressure and normal culture. Biochemical and electrophores findings were: Protein: 18 mg/100ml, Glucose: 80 mg/100 ml and Chlorure 125 mmol/L.

Blood biochemical studies showed that complete blood count was normal and other standard biochemical studies, including measurements of blood urea nitrogen, glucose, sodium, potassium, creatinine, phosphorus, calcium levels gave normal results. The erythrocyte sedimentation rate was 92 mm per hour. Thyroid function studies and glucose tolerance tests were normal. Muscle biopsies revealed normal histopathological findings.

Electromyography (EMG) showed continuous motor

unit activity and this pattern disappeared after intravenous administration of 10 mg diazepam. Results of repeat EMG studies did not change and the patient's condition was diagnosed as Stiff-man syndrome. Treatment with diazepam 15 mg daily in three doses was started and her general condition improved slightly so the doses were increased to 30 mg daily in one week and 60 mg in two weeks. Two weeks later patient's general condition improved. There was reduction in muscle stiffness and she was walking without difficulty. The patient was discharged and is being followed for two years. She is still taking 30 mg diazepam daily and is well.

DISCUSSION

The Stiff-man syndrome (SMS) is a rare disorder of motor function characterised by progressive fluctuating painful spasm and rigidity of axial muscles associated with continuous motor unit activity on EMG even when at rest (1). The disease was first described by Moersch and Woltmann in 1956 (2). Since then some reports have been presented about its pathogenesis, diagnostic criteria and treatment (3-7). The exact cause of the disease is not clear and psychiatric, myopathic and metabolic disturbances have been proposed. Recently some reports have suggested that the cause may be autoimmune or paraneoplastic (4,5,7). It was thought that SMS was due to impairment of the GABA-ergic inhibitory system and antibodies to glutamic acid decarboxylase (GAD) were investigated (4). Several metabolic disorders associated with SMS have been reported such as type-1 diabetes mellitus (1,4,8), dementia (9), encephalomyelitis (6), bronchogenic carcinoma (5), and esophageal obstruction (10). The disease shows many typical and atypical features and because of its rarity and lack of the diagnostic tests and widespread physician unawareness of the entity, SMS generally remains undiagnosed or misdiagnosed (11). With an insidious onset and slow progression of symptoms, it tends to affect middle aged males. The main complaints are muscular tightness, stiffness and rigidity especially in proximal limbs, lower back and abdomen which result in difficulty in mobility, spinal hyperlordosis and broadlike abdomen.

Neurological examination is usually normal. No diagnostic test but electromyographic data is important and shows continuous motor unit activity in all cases. This pattern can be overcome by myoneural blocking agents, chemical peripheral nerve block, general anesthesia and intravenous administration of diazepam (1,10,11).

At diagnosis, several conditions must be differentiated from the SMS such as acute or chronic tetanus, a primary myopathic process, progressive encephalomyelitis with rigidity, Isaac's syndrome, Wilson's disease, Mc Ardle myopathy or myotonic dystrophy.

Since the disease was first described, many therapeutic modalities have been proposed. Steroids

(12), baclofen (8), sodium valproate (13), clonazepam (14), clonidine and tizanidine (15), and diazepam (1,10,11,15,16). Diazepam is generally accepted as the most effective medication and symptoms decrease with large daily doses. The patient's symptoms similarly relieved with this medication without excessive sedation.

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