A STIFF MAN.
DOES HE NEED A NEUROLOGIST OR AN ORTHOPAEDIST?

Maddalena Sparaco[1], Luigi Lavorgna[1], Antonio Gallo[1], Gioacchino Tedeschi[1], Simona Bonavita[1]

[1] Ist Clinic of Neurology, Second University of Naples, Piazza L. Miraglia 2, 80138 Naples, Italy

In March 2016 a 35-year-old man was admitted to our Department for walking and sitting up difficulties and a sensation of trunk and lower limbs (LL) stiffness in the last two years. In anamnesis there was a potus history in the last 15 years, with a progressive moving away from alcohol in the last two years. When he came to our Department, he drinked 150 cc of alcohol/day.

Neurological examination showed gait with enlargement of the crandle and external rotation of foot, impossible in tandem and difficult on toes and heels, difficulty in bending the thighs on the pelvis with contralateral limb lifting, normal tone and strength in the arms but symmetrical increased tone of the proximal muscles of the LL, normal all tendon reflexia and superficial and deep sensitivity, no pyramidal and extrapyramidal signs. The differential diagnosis among stiff person syndrome, toxic motor neuropathy, axial and proximal dystonia was performed. The clinical presentation was suggestive of stiff person syndrome.

Dalakas criteria

for Stiff Person Syndrome

- Episodic stiffness of the muscles, mostly involving the axial muscles (particularly in the abdomen and thoracolumbar paraspinal), leading fixed deformity
- Superimposed painful spasms precipitated by triggers such as noises, emotional stress and tactile stimuli
- Absence of neurological or cognitive impairment that could explain the stiffness
- Confirmation of continuous motor unit activity in agonist and antagonist muscles by electromyographic findings (EMG). Positive serology for GAD65 or amphiphysin autoantibodies confirmed by immunocytochemistry, Western blot, or radioimmunoassay.

Response to diazepam is not a part of original Dalakas criteria, but commonly used for diagnosis of stiff person syndrome.

Brain magnetic resonance imaging (MRI) was normal, spinal cord MRI showed multiple cervical and lumbar hernias. Motor evoked potentials were normal. Electroneuromyography at the LL and paraspinal muscles was normal, as it was at the level of the recti abdominis muscles. Serum anti glutamic acid decarboxylase and anti-amphiphysin antibodies were negative. Diazepam i.v. did not improve the symptoms.
At this point an orthopedic etiology was suspected and a hips and pelvis X-ray, showed a reduction of joint line with marginal sclerosis of the acetabular cavity and roof of the femoral heads, in the absence of traumatic injuries, as a bilateral necrosis of the femoral heads.

A diagnosis of bilateral aseptic necrosis of the femoral heads (ANFH) was made and the patient went to an orthopedic surgeon for prothesis.

The bilateral necrosis of the femoral heads is common between the 3rd and 5th decade of life, particularly affecting men. Osteonecrosis can be due to traumatic or non- traumatic causes. Among a variety, alcoholism is one of the most common cause of ANFH. One of the major feature of ANFH is, groin pain irradiating to the gluteal, anterior and medial aspect of the thigh and some times to the knee. In our patient the absence of pain and of the secondary limp were confounding factors. In conclusion, ANFH may be considered in the differential diagnosis of some neurological diseases (such as Stiff person syndrome) even when the clinical presentation is atypical.

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