

CLINICAL CORRESPONDENCE

Stiff-person syndrome presenting with headache

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Stiff-person (or stiff-man) syndrome (SPS) is a rare neuromuscular disorder characterized by axial and limb muscle rigidity and spasm. Painful spasms of muscle can be provoked by sudden noise, strong emotional or other startle stimuli. Electrophysiological studies show continuous low-frequency firing of motor units and simultaneous activity in agonist and antagonist muscles of the affected region that can be abolished by central or peripherally acting agents or neuromuscular blockade (1). An autoimmune basis for the disorder is postulated, based on the presence of antibodies against glutamic acid decarboxylase (GAD) found in 90% of cases defined by strict criteria (2). GAD is the rate-limiting enzyme involved in the synthesis of gamma-aminobutyric acid (GABA), an inhibitory neurotransmitter. Many affected patients have other autoimmune disorders such as pernicious anaemia, vitiligo and thyroid malfunctions (3). A reduction in brain levels of GABA has been demonstrated on magnetic resonance (MR) spectroscopy scans (3). The loss of inhibitory input from spinal interneurons is theorized to lead to an impaired balance between inhibitory and excitatory influences on muscle activity (4).

SPS usually begins insidiously with persistent aching and stiffening of axial muscles. Involvement of the face, neck, abdomen and arms is less typical. Headache as a presenting or major feature of the illness has not been described. Examination generally shows global muscle stiffness, with no other neurological abnormalities, and no abnormalities on computed tomography (CT) or MR imaging of the brain. Treatment is based on enhancing inhibitory activity in the spinal cord through the use of drugs such as diazepam, baclofen and vigabatrin that boost GABA neurotransmission. Steroids, plasmapheresis and intravenous immunoglobulin are also helpful (3). There are reports of improvement in severe muscle spasms and dystonia with the use of botulinum toxin (5).

The authors report the case of a 34-year-old male with SPS in whom headache was the presenting feature, and a complicating feature throughout the illness.

Case report

A 34-year-old white male with a 5-year history of SPS incompletely responsive to treatment with intravenous immunoglobulin, diazepam, steroids and other treatments was admitted to a rehabilitation hospital after placement of an intrathecal baclofen pump. The patient experienced dramatic improvement of his back and leg symptoms following placement of the pump, but developed a severe, waxing and waning occipital headache. Consultation was requested from the pain and headache service. The patient was an excellent historian, and reported no personal or family history of headache until 5 years prior to admission.

At that time the patient had developed a severe occipital headache similar in all respects to the current headache. This first headache occurred with no provocation and escalated over a period of minutes to steady occipital pain rated 10 on a numeric rating scale of 0–10. There were no associated symptoms. The patient reported that the pain was steady, 'like an ice pick', and so severe that he fell to his knees and was unable to move. He believed that 'an aneurysm had burst'. He sought treatment at a hospital emergency department, where mild tachycardia and hypertension were noted. A CT scan of the head was normal. He was treated with intramuscular meperidine, with complete resolution of the headache. Following the initial headache, he had 10–12 similar episodes including the current headache. In each, the pain came on suddenly and reached maximal severity within minutes. Often the severity of the pain would gradually decrease, but the headache would not remit until he received parenteral medication,

usually meperidine. Some headaches were associated with nausea or 'dry heaves'. One headache occurred during sexual activity, immediately preceding orgasm. It did not remit with cessation of sexual activity.

Shortly after his initial headache, the patient developed progressive symptoms consisting of diaphoresis, burning muscle pain, especially in the back, muscle twitching, weight gain, fatigue and hypertension. He was eventually diagnosed with SPS. One test for antibodies to GAD was positive, but other tests were negative. The diagnosis of SPS was made based on electromyographic findings of persistent motor unit activity. The patient was treated with intravenous immunoglobulin, gabapentin, diazepam, steroids and other medications with incomplete resolution of his symptoms. He became progressively disabled as a result of muscle spasm and stiffness that impaired ambulation. The patient reported his belief that, in hindsight, his first severe headache was the initial symptom of SPS.

The patient had a history of hyperprolactinaemia treated with bromocriptine (no pituitary adenoma was identified on MR imaging); sleep apnoea treated with pharyngeoplasty; intermittent hypertension; recurrent staphylococcal furunculosis requiring dicloxacillin suppressive therapy; and bilateral carpal tunnel syndrome.

On examination, the patient was alert and appeared to be in moderate pain. He had bulky, hypertrophied muscles and the appearance of a weight lifter. An intrathecal baclofen pump was present under the skin of the abdomen with a catheter tunnelled subcutaneously to the spinal region; incisions were healing well. The muscles of the arms, back, neck and legs were tight, hard and large, with minimal tenderness on palpation. Muscle tone was increased. There was exaggeration of the normal lumbar lordotic and thoracic kyphotic curves. Range of motion of the neck was limited by muscle bulk and pain. The patient reported tenderness on palpation over the occipital area bilaterally, most marked on the left side, with exacerbation of his headache pain. Sensation in the C2-C3 distribution bilaterally was normal. There were no other neurological abnormalities.

Bilateral greater occipital nerve blocks were performed with 2 ml of 2% lidocaine and 6 mg of betamethasone. Good anaesthesia of the C2-C3 distribution bilaterally was obtained, and the patient reported resolution of his headache. When contacted 2 months after discharge, the patient reported that although he had minor occipital pain from time to time, he had not experienced any further episodes of

severe or disabling head or neck pain. His intrathecal baclofen dose had been titrated based on symptoms, with dramatic improvement of axial pain and increased activity level.

Discussion

We describe a man in whom headache was the presenting feature of an otherwise typical case of SPS. It seems likely that the mechanism of headache in this patient was sustained contraction of hypertrophied neck muscles. The onset and evolution of the patient's headaches occurred in close temporal association with the onset and evolution of SPS, and subsided following placement of an intrathecal baclofen pump. No other explanation for this patient's headaches is as plausible as a primary muscle aetiology. The headaches as described by the patient do not meet International Headache Society (IHS) criteria for a primary headache disorder, although one headache was very similar to IHS type 4.6, headache associated with sexual activity. However, it was not eased by ceasing sexual activity, as required for diagnosis (6).

Although this patient's headache improved after occipital nerve block, the steady character of the pain and lack of paraesthesias or dysaesthesias are not typical of occipital neuralgia. The temporary improvement in pain may have been part of the natural waxing and waning character of the headache, or may have been a placebo response based on conditioning and expectation. Sleep apnoea has also been implicated as a cause of headache, but seems an unlikely explanation in this case, since it had been diagnosed and treated long before onset of the headaches. A low-pressure headache syndrome from spinal fluid leakage following pump placement is unlikely, since the patient's headache was similar to headaches that occurred prior to placement of the baclofen pump, and was not positional. The improvement in headache after titration of his intrathecal baclofen is notable, since the administration of intrathecal baclofen in the lumbar region usually provides limited effects in the cervical and cranial region. This may reflect a reduction in the 'triggering' of cranial muscle spasm by other axial muscle spasms, and represent an indirect effect of the intrathecal baclofen. Similar indirect effects have been seen in patients with upper and lower limb spasticity.

Patients and physicians commonly invoke 'muscle contraction' as an explanation for headache, but scientific evidence that it can cause headache is limited. The IHS classification system does not recognize the

entity 'muscle contraction headache', since the majority of patients to whom this term has been applied cannot be demonstrated to have either elevated muscle tension on electrophysiological studies or muscle tenderness on palpation. Most patients who are considered to have 'muscle contraction headache' are classified by the IHS system as having a subtype of tension-type headache 'associated with disorder of pericranial muscles'. However, the IHS comments that this subdivision should be regarded as optional, 'in view of the poor scientific basis' for it, and acknowledges that 'for decades dispute has prevailed concerning the importance of muscle contraction mechanisms, but conclusive studies are still lacking' (6).

This case suggests that pathological levels of sustained muscle spasm and contraction can in fact produce significant levels of headache. Whether less pronounced or less sustained levels of muscle spasm commonly play an important initiating or perpetuating role in other headache syndromes remains to be determined. Although suggestive of an important role for muscle spasm in the production of some headache disorders, our report is of a single case, and the duration of follow-up is not yet sufficient to decide whether the patient has experienced a true remission with baclofen treatment. However, recent demonstrations that injections of botulinum toxin may be useful in the prophylaxis of migraine also imply that muscle factors may be

more important in headache than previously recognized. It is also possible, though, that botulinum toxin exerts its beneficial effects through mechanisms other than muscle relaxation (7). If headache recurs in this patient, the use of botulinum toxin injections into hypertrophied cervical muscles could be considered.

References

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