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Neuromyelitis Optica with Painful Tonic Spasms

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A thirty-year-old female with cervical cord myelitis and recurrent optic neuritis was diagnosed with neuromyelitis optica (an uncommon demyelinating disorder of central nervous system). Few days after successful treatment of recurrent optic neuritis, she developed painful tonic spasms. Clinical examination and repeated investigations did not reveal any relapse of the disease. The neurologist prescribed oral antiepileptic drugs, which controlled her spasms.

Painful Tonic Spasm (PTS) is known to be associated with Neuromyelitis Optica (NMO) as well as Multiple Sclerosis (MS). However, it is misunderstood and an under diagnosed condition. The fact that PTS could be readily ameliorated with antiepileptic drugs, underscores the importance of its prompt detection.

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Neuromyelitis Optica (NMO) or Devic's disease is a rare autoimmune disorder, characterized by demyelination of Central Nervous System (CNS), mainly the optic nerves and spinal cord. NMO is clinically distinct and separate from Multiple Sclerosis; the latter is more common demyelinating disease of CNS. In addition to the well-known presentations of such disorders, patients with MS and more likely with NMO, may have sensory symptoms that could be severely disabling. Painful Tonic Spasm (PTS) is rather a misunderstood and an under diagnosed condition.

The aim of this case is to highlight the diagnostic criteria for Neuromyelitis Optica and Painful Tonic Spasm which responded well to antiepileptic drug therapy.

THE CASE

A thirty-year-old female presented with episodic, severe pain felt like sparks of fire predominantly in the right upper and lower limbs, but at times also involving the neck and the left side, along with stiffening of the affected limbs, which lasted for 7 days. Each episode had duration of few seconds and would recur after every minute or two.

The symptoms were mild and infrequent but within days they became more frequent and unbearable.

Four months before her presentation, she had acute vision loss of the left eye. She was diagnosed as optic neuritis, and was treated with steroids, which resulted into partial recovery. Two months after, she developed gait difficulty. MRI revealed lesion in cervical spinal cord, hypointense at T1 and hyperintense at T2 weighted sequences extending from 3^{rd} to 6^{th} cervical spinal cord segments, with minimal contrast uptake suggestive of cervical cord myelitis, see figure 1. MRI brain revealed normal result. She recovered partially and was able to walk unsupported with minimal difficulty.

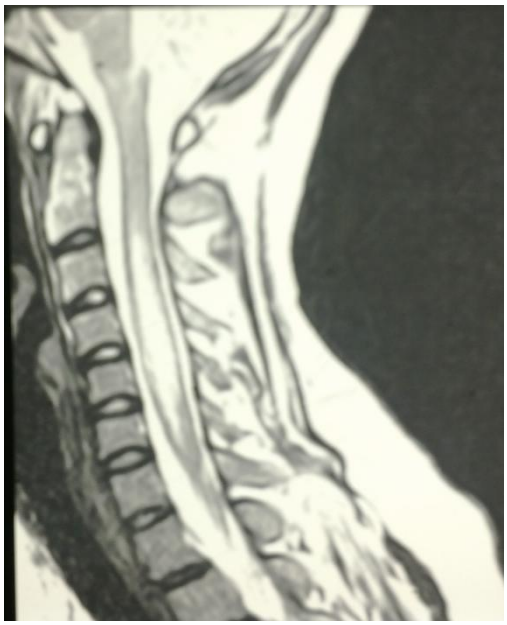

Figure 1: T2W Sagittal View of Initial MRI Scan at Onset of Myelitis, Showing Hyperintense Lesion in Upper Cervical Spinal Cord

Six weeks later, she developed an acute blurring of vision in the right eye associated with periorbital pain. A repeat MRI brain with contrast was normal. The patient refused a lumber puncture. Visual Evoked Potentials showed significantly delayed latency of P 100 at 275ms in right optic nerve with an amplitude of 2.54 uV and non-recordable potentials on left side, suggestive of demyelinating injury of the optic nerves. Her serum ANA, anti-ds DNA, AMA, ASMA were negative. Her diagnostic work-up was consistent with Neuromyelitis Optica (Devic's disease). She received pulse therapy and intravenous methyl prednisolone which resulted in significant improvement.

Subsequently, she was advised oral prednisolone taper and azathioprine (after informed consent); the vision in right eye had improved. A day after her pulse therapy, she reported mild pain in the right leg which she described as stiffening. She was prescribed Baclofen and Pregabalin.

Her condition worsened and she had episodes of tightening or stiffening of the limbs with severe pain. Escitalopram was also prescribed by a physician who considered it as a conversion disorder.

On examination, the patient was an anxious young female of average height and built, conscious, alert and oriented.

The patient was observed as having repeated posturing of her limbs mostly on the right side, in which her limbs would become very rigid attain a position in mid flexion and she would complain of severe pain in the affected limbs. Such episodes would last for 1 to 2 minutes followed by complete recovery and would recur on the same side or on both sides simultaneously after 3 to 4 minutes.

Her visual acuity in right eye was 20/40, while in the left eye was finger counting from 12 inches. Fundoscopy showed bilateral optic disc pallor. Other cranial nerves were intact. Upper limbs had normal motor power but depressed deep tendon reflexes. Lower limbs had increased tone bilaterally. Power at hips was 4/5 on both sides, normal at knees and ankles.

Deep tendon reflexes were brisk at the knees and ankles with bilateral ankle clonus, and equivocal planters. No cerebellar or consistent sensory signs.

A diagnosis of painful tonic spasm was made. She was started on carbamazepine and a repeat MRI scan of cervical spine was requested.

Her pain subsided completely; however, she developed skin rashes due to carbamazepine, which she had to be discontinued, and was replaced with phenytoin.

Repeated MRI scan showed resolution of cervical spinal cord lesion, see figure 2.

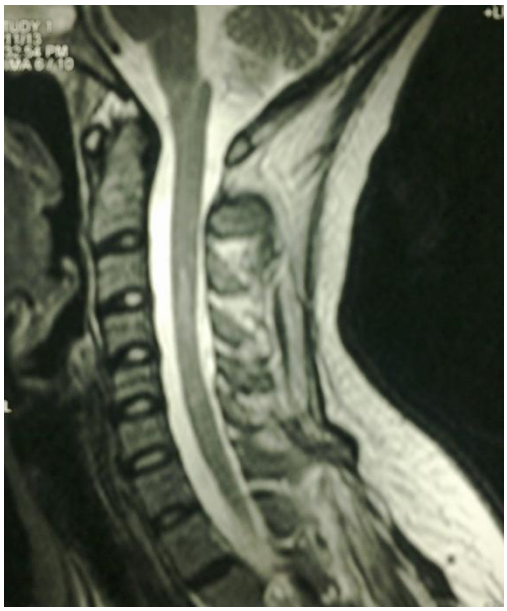

Figure 2: T2W Sagittal Sequences of Cervical Spine. A Follow-up Scan Showed Marked Resolution of Signal Abnormality

Unfortunately, the patient developed phenytoin induced hepatitis, which was replaced with Levetiracetam, which proved to be effective in controlling the spasms.

DISCUSSION

Neuromyelitis Optica, although rare, should be considered whenever dealing with suspected CNS demyelinating disorder. It may have a relapsing course and thus may mimic multiple sclerosis. As far as long term management, it is different from MS; therefore, it is important to recognize this disorder. Two different diagnostic criteria are proposed and are very helpful in diagnosing NMO[1].

Patients with multiple sclerosis and neuromyelitis optica can have a myriad of symptoms and signs. Most notably, they have very unusual pain syndromes which could be disabling as any other motor deficit; one of these is painful tonic spasm[2]. It is an under-diagnosed condition, although it could be easily treated with antiepileptic drugs[3].

Painful tonic spasms are brief episodes of severe pain in half of the body or a limb, along with flexion spasms of the involved limb, at times occurring on both sides of the body without loss of consciousness[4]. Pain is a very prominent part of symptomatology; it is subjective in nature and has spasm of unusual appearance. At times this condition is labeled as a conversion disorder or psychic overlay, as was the case with our patient. Other erroneous labels are flexor spasms, spinal myoclonus and Lhermitte's sign. However, painful tonic spasm is different from all these because of the fact that (i) it is extremely painful, which is not the case with flexor spasms or spinal myoclonus and (ii) it is not triggered by neck flexion nor does it involve the spine predominantly which is found in Lhermitte's sign/phenomena[4,5].

Other possibilities of stereotyped unilateral body movements without alteration of consciousness include epileptic seizures, carpopedal spasm (tetany), and some rare disorders, such as paroxysmal kinesogenic choreoathetosis, and paroxysmal dystonic choreoathetosis[5]. However, none of these disorders are associated with abnormalities of CNS white matter.

Simple partial epileptic seizures with motor manifestations often show abnormality on ictal scalp electroencephalography; however, an EEG may not be sensitive enough. Compared with tonic spasms, partial seizures usually tend to be less frequent and are usually not painful. Carpopedal spasm (tetany) occurs with hypocalcemia or severe metabolic alkalosis; it is usually painless, bilateral, and longer lasting than a tonic spasm. Paroxysmal kinesigenic choreoathetosis and paroxysmal dystonia are rare conditions that begin at a younger age and have characteristic appearance which is very much different from simple tonic postures[5].

Recently, it was found that the appearance of painful tonic spasms are more common in patients with NMO and most of the patients develop such spasms during resolution of the symptoms, as in our case; therefore, any patient with NMO or MS developing PTS does not necessarily has a relapse and hence may not require a pulse therapy with steroids[6].

Antiepileptic drugs are known to relieve PTS symptoms, carbamazepine followed by phenytoin. Tiagabine and levetiracetam have been also effective[4,5,7]. Unfortunately, our patient developed severe adverse reactions with first and second line treatment and therefore other options were considered. Due to its availability and better side-effect, Levetiracetam was chosen and the patient responded favorably.

CONCLUSION

This case highlights the importance of identifying the condition of painful tonic spasm because it is easily treatable with low dose antiepileptic drugs. In addition, it is important that NMO should be considered while dealing with CNS demyelinating diseases as it might be confused with multiple scelerosis[8].

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