Case Report

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A rare case of short-lasting unilateral neuralgiform headache with conjunctival injection and tearing with progression to neuromyelitis optica spectrum disorder

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Abstract

Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) is a rare primary headache syndrome. However, some cases of secondary SUNCT are attributed to underlying diseases such as demyelination. We herein report a case of SUNCT with progression to neuromyelitis optica spectrum disorder (NMOSD). A 43-year-old woman developed headaches; 6 weeks later, she developed bilateral visual loss and numbness on the left side of her body. She was ultimately diagnosed with NMOSD.

Keywords

Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing, neuromyelitis optica spectrum disorder, aquaporin-4, magnetic resonance imaging, longitudinally extensive transverse myelitis, steroid therapy

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Introduction

Short-lasting unilateral neuralgiform headache with conjunctival injection and tearing (SUNCT) is a rare type of headache that was first described in 1978.[1] The headache is typically characterized by the occurrence of moderate or severe short-lasting orbital, periorbital, or temporal stabbing or pulsating pain lasting 1 to 600 s with lacrimation, conjunctival injection, and rhinorrhea.[2]

Neuromyelitis optica spectrum disorder (NMOSD) is an astrocytopathy of the central nervous system that is associated with aquaporin-4 immunoglobulin G antibodies (AQP4-IgG).[3] The core characteristics include clinical symptoms or magnetic resonance imaging (MRI) findings related to the optic nerve, spinal cord, area postrema, or other brain stem regions and diencephalic or cerebral presentations.

SUNCT is a primary headache syndrome. However, cases of secondary SUNCT attributed to neoplasms, vascular disease, trauma, infection, inflammation, congenital malformation, or demyelination have also been reported. In this report, we describe a patient who was initially diagnosed with SUNCT but was later diagnosed with NMOSD.

Case presentation

A previously healthy 43-year-old woman without a significant familial history of headache presented to the pain clinic at our hospital with a 2-month history of severe pain in the right temporal area. The pain was abrupt at onset, stabbing or burning in quality, and accompanied by lacrimation and conjunctival injection. The frequency varied from 10 to 20 times a day, with each attack lasting 20 to 40 s. Initial brain MRI revealed bilateral supratentorial white matter lesions (Figure 1) without spinal cord or optic nerve involvement. She was diagnosed with SUNCT. Treatment with carbamazepine and lamotrigine had little effect. In addition, supplemental oxygen administration in the hospital had no significant effects.

Six weeks after the onset of SUNCT, the patient developed bilateral visual loss and numbness on the left side of her body.

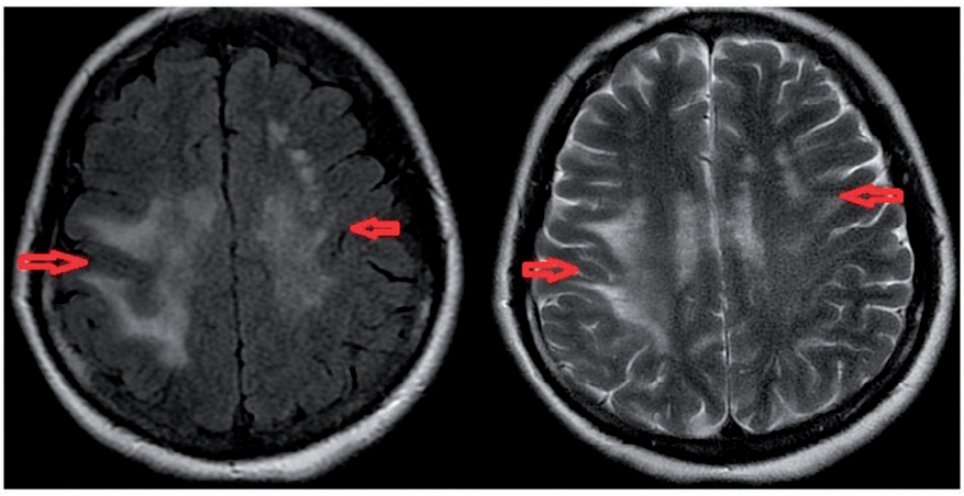

Figure 1. Brain magnetic resonance imaging revealed bilateral white matter lesions.

A few days before hospital admission, these symptoms progressed to limb weakness and urinary retention. Neurological examination showed asymmetric tetraparesis with grade 4 muscular strength in the right limb and grade 3 in the left limb. All sensory modalities were compromised in the limbs and trunk at the T4 level. Lhermitte's sign was positive. MRI revealed longitudinally extensive transverse myelitis (Figure 2). Axial T2-weighted MRI of the cervical spine showed hyperintense intramedullary lesions (Figure 3). The patient's right eye acuity was 0.3 and left eye acuity was 0.4. All laboratory test results were normal, including a blood chemistry panel, complete blood count, erythrocyte sedimentation rate, electrolyte measurements, magnesium measurement, thyroid hormone determination, and liver and coagulation function tests. The anti-double-stranded DNA, endonuclear antigen, and antiphospholipid antibody test results were negative, but the antinuclear antibody result was positive. The results of a paraneoplastic panel (anti-Hu, anti-Yo, and anti-Ri antibodies) were unremarkable. Sjögren's syndrome, myasthenia gravis, vasculitis, rheumatoid arthritis, diabetes mellitus, hypothyroidism, systemic lupus erythematosus, autoimmune encephalitis, and underlying malignancies were excluded. A lumbar puncture was performed, and the cerebrospinal fluid (CSF) pressure was 140 mmH_2O. The IgG level was normal, and no oligoclonal band was detected. Antibodies to myelin oligodendrocyte glycoprotein were negative in the serum and CSF. However, antibodies to AQP-4 were positive in both the serum and CSF. The patient was ultimately diagnosed with SUNCT with progression to NMOSD. Steroid pulse therapy was performed for 5 days (1 g/day of methylprednisolone), and 60 mg of oral prednisolone was administered thereafter.

Approximately 3 weeks later, the patient's neurologic symptoms gradually improved. Prednisone was continued at 40 mg/day. Her headache disappeared, and her visual acuity improved to 0.6 in the right eye and 0.7 in the left eye. She was able to walk without assistance. In addition, her urinary retention, eyelid swelling, and tearing were alleviated.[4] The patient was eventually discharged with continuance of prednisone at 30 mg/day.

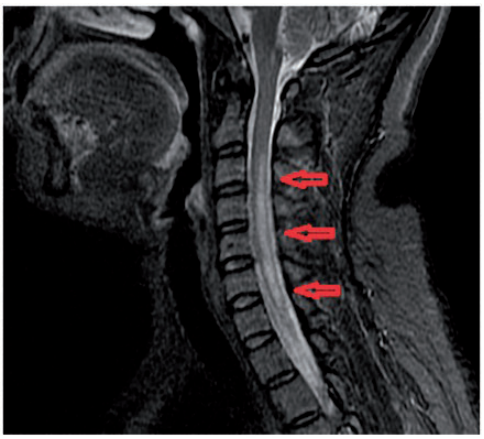

Figure 2. T2-weighted brain magnetic resonance imaging revealed longitudinally extensive transverse myelitis lesions.

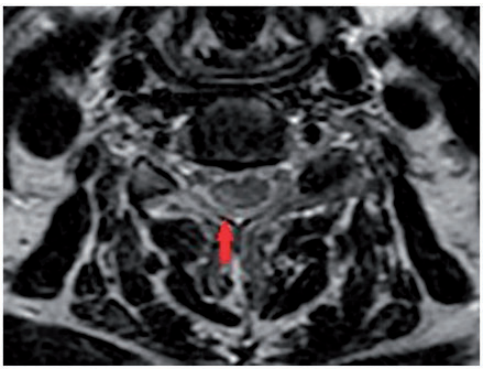

Figure 3. Axial T2-weighted magnetic resonance imaging of the cervical spine showed hyperintense intramedullary lesions.

Discussion

Headaches can be divided into primary headaches and secondary headaches caused by other diseases. SUNCT is a rare type of headache, and most cases are primary headaches. SUNCT is an acronym; the "S" signifies "short-lasting," "U" signifies "unilateral," "N" signifies "neuralgiform," "C" signifies "conjunctival injection," and "T" signifies "tearing."[5] The headache attacks associated with SUNCT are usually unilateral, stabbing, burning, or electrical and are associated with ipsilateral conjunctival injection, tearing, and swelling of the eyelids.[4] The condition is characterized by the occurrence of at least 20 attacks lasting for 1 to 600 s.[6] However, there are descriptions of bizarre clinical cases involving peculiar aspects that can be misleading in clinical practice. Antonaci et al.[7] described a patient with SUNCT syndrome who exhibited paroxysmal mydriasis, and they assumed that the involvement of the ocular sympathetic innervation was responsible for the mydriasis. SUNCT is more common in men than women. Most cases of secondary SUNCT are associated with neoplasms, vascular disease, infection, trauma, inflammation, congenital malformation, or demyelination, suggesting that secondary SUNCT can be a secondary symptom. The response to the lamotrigine therapy can provide useful information. Our case fulfilled the diagnostic criteria for SUNCT. However, our patient was a woman, and treatment with carbamazepine and lamotrigine had little effect.

Several weeks after the onset of SUNCT, the patient developed visual loss, numbness, and limb weakness. AQP-4 was positive, and she was diagnosed with NMOSD. After treatment with corticosteroids, her headache disappeared. The diagnosis of secondary SUNCT with progression to NMOSD was later confirmed. Increasing evidence shows that secondary SUNCT is attributable to several known diseases, such as neoplasms, vascular disease, trauma, infection, inflammation, congenital malformation, and demyelination. The pathogenesis of secondary SUNCT is not well established. The pathogenesis of SUNCT is likely related to the trigeminal nerve conduction.[8] Cao et al.[9] assumed that the dorsolateral medulla, upper cervical spinal cord, pons, and preganglionic fibers of the trigeminal nerve were mainly responsible for the induction of secondary SUNCT. Because of the involvement of the trigeminal nerve branch, trigeminal nucleus, spinal thalamic tract, and trigeminal thalamic tract, pathological changes occur in the pons, dorsolateral medulla oblongata, and cervical spinal cord, which may induce headache.[10,11] Patients with SUNCT present with conjunctival injection and tearing because of neurovascular compression in the trigeminal root entry zone and the first division of the trigeminal nerve. In addition, the distribution of the parasympathetic nerve fibers in the mucous membrane of the lacrimal gland may be invaded.[12,13]

NMOSD is an astrocytopathy of the central nervous system that is associated with AQP4-IgG. It has been traditionally thought to only involve the spinal cord and optic nerves, but several brain lesions are highly suggestive of NMOSD based on the 2015 Wingerchuk criteria.[3] Periependymal lesions in the diencephalon, long corticospinal tract, dorsal brain stem, and hemispheric cerebral white matter are often involved in NMOSD. The initial presentation of NMOSD can include vomiting, weight loss, anorexia, and headache. Brain symptoms can be involved in the initial presentation of younger patients (in their 30s or 40s) with NMOSD.[14,15] Our patient's initial symptom was headache, and her first brain MRI scan revealed hemispheric cerebral white matter lesions. The brain MRI lesions were not adjacent to the lateral ventricular body and did not conform to the

imaging features of multiple sclerosis. Cerebral white matter lesions are likely related to vasogenic edema,[16] and these lesions are often tumefactive, radial-shaped, or long spindle-like signal changes. Similar to the optic nerve and spinal cord, relatively long axons may be particularly vulnerable and are susceptible to inducing NMOSD.[17] Lee et al.[18] described a patient with anti-AQP4-positive long-standing NMOSD who exhibited leukodystrophy-like white matter involvement.

Treatment of NMOSD starts with intravenous methylprednisolone, plasma exchange, or intravenous immunoglobulin. After corticosteroid treatment, our patient's neurologic symptoms improved.

Conclusion

We have presented a rare case of secondary SUNCT attributed to NMOSD. Doctors should pay more attention to secondary headaches and atypical presentations of NMOSD. This is a rare disease, and more research in this area is warranted. A correct early diagnosis and proper treatment are essential to prevent serious symptoms.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethical approval

The authors affirm that the patient depicted in the images or other personal information gave written informed consent for all procedures and for dissemination of her data for educational and research purposes. The study protocol was approved by the ethics review committee of Tianjin Baodi Hospital.

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