Case Report

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A case of multiple system atrophy

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Abstract

Multiple system atrophy (MSA) is the most rapidly progressive neurodegenerative disorder among the various types of synucleinopathies. The cause of MSA remains unknown, but it can involve the extrapyramidal system, the pyramidal system, the autonomic nerves and the cerebellum. The main clinical manifestations are Parkinson's symptoms, cerebellar ataxia, pyramidal tract signs and autonomic nervous system disorders. Depending on the initial predominant motor deficits, MSA is subclassified into either Parkinsonian type (MSA-P) or cerebellar type (MSA-C). MSA is rare in the Zunyi area of Guizhou Province, so when it is observed for the first time it often results in a convoluted diagnosis and treatment process, which takes a lot of time, money, manpower and material resources, which can also have a psychological impact on the patient. This report describes the case of a 60-year-old woman who presented with syncope for 1 year combined with dizziness for 1 day. She had been diagnosed twice with transient ischaemic attack in the previous 6 months. Cranial magnetic resonance imaging suggested widening of the cerebellar sulcus and mild cerebellar atrophy. Based on the patient's medical history, physical signs and auxiliary examinations, she was diagnosed with MSA-C.

Keywords

Multiple system atrophy, diagnosis, treatment

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Introduction

Multiple system atrophy (MSA) is the most rapidly progressive neurodegenerative disorder among the various types of synucleinopathies.[1] MSA is considered an orphan disease with an annual incidence rate of 0.1–3.0 per 100 000, depending on age and

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geographical region.[2] Based on the predominant clinical phenotype, the disease is subclassified into either Parkinsonian variant (MSA-P) associated with striatonigral degeneration, a cerebellar (MSA-C) variant with olivopontocerebellar atrophy and predominant cerebellar features, or a combination of both forms, referred to as 'mixed' MSA.[3,4] Based on these findings, the second consensus statement regarding the diagnosis of MSA was proposed in 2008.[3] MSA is defined by specific clinical findings such as a Babinski sign with hyperreflexia, stridor, rapid progression rate, poor response to levodopa, and ataxia or characteristic neuroimaging abnormalities based on magnetic resonance imaging.[5] Because of the significant impact of neurodegenerative diseases on humans, physicians need to obtain more comprehensive knowledge regarding its early detection and prevention, so that they can effectively improve the quality of life of patients affected by the disorder.

Case report

A 60-year old woman was admitted to the Department of Neurology, The First People's Hospital of Zunyi and The Third Affiliated Hospital of Zunyi Medical University, Zunyi, Guizhou Province, China in September 2017 due to experiencing syncope for 1 year combined with dizziness for 1 day. Starting 1 year previously, the patient had experienced repeated episodes of sudden syncope after changing her position. She would fall to the ground, exhibit no response to others, and the episodes would last for a few seconds each time. She reported satisfactory consciousness, long-term constipation, occasional urinary incontinence, without limb convulsions, lip or tongue bites, heart palpitations, chest tightness or difficulty breathing, headache or vomiting, limb sensation or movement disorders. Physical therapy recommendations had been given to the patient such as lying in a prone position, ensuring the head and trunk were 15–20 degrees higher than the lower limbs, and wearing tight trousers as well as elastic socks to apply pressure to the legs. In March 2017, after 6 months of recurrent syncope, she spent 8 days in hospital and was diagnosed with transient ischaemic attack. She was discharged after treatment with nutritional neurotherapy, which improved circulation. The above-mentioned symptoms recurred again, exhibiting the same nature as in the previous event, with dizziness, discomfort, walking unsteadily, and no limb convulsions. So, in September 2017, after 1 year of syncope, she was admitted to hospital for 1 day and diagnosed with transient ischaemic attack. She had a healthy status, did not consume tobacco or alcohol and did not report any history of genetic diseases or similar diseases in her family. The physical examination undertaken at the time of hospital admission in September 2017 showed that her vital signs were in the normal range, and the heart, lung and abdomen were normal. With regard to her nervous system, she was conscious, speaking fluently and smoothly, with normal high-level nerve activity. The meningeal irritation sign was negative, bilateral pupils were equal in size at approximately 3.0 mm, which were sensitive to the light reflex. Sometimes, horizontal nystagmus could be induced. There were no abnormalities found in the remaining cranial nerves, muscle strength was at grade IV and muscle tension was normal. Symptoms of the whole body were symmetrical; her quadriplegia reflex was +++ and a Babinski sign on the left was suspected to be positive. The finger-to-nose test was unstable and the heel-to-shin test was positive. Rotation exercise was acceptable, Romberg was positive and a skin scratch test was positive. In September 2017, cranial magnetic resonance imaging (MRI),

diffusion-weighted imaging (DWI) and magnetic resonance angiography (MRA) suggested the following: (i) mild cerebellar atrophy; (ii) no abnormalities on the brain MRA (Figure 1). In March 2017, dynamic electrocardiogram suggested the following: (i) sinus rhythm; (ii) premature atrial contraction, short-term atrial contraction; (iii) heart rate variability within the normal range; and cardiac colour Doppler suggested the following: the mitral, tricuspid and aortic valves all demonstrated mild regurgitation; and there was normal left ventricular systolic function (ejection fraction: 62%). In September 2017, the standing and lying blood pressures differed by 51/35 mmHg. Serum ion, routine blood examinations, liver function, renal function and fasting venous blood glucose were not significantly abnormal. Combined with the

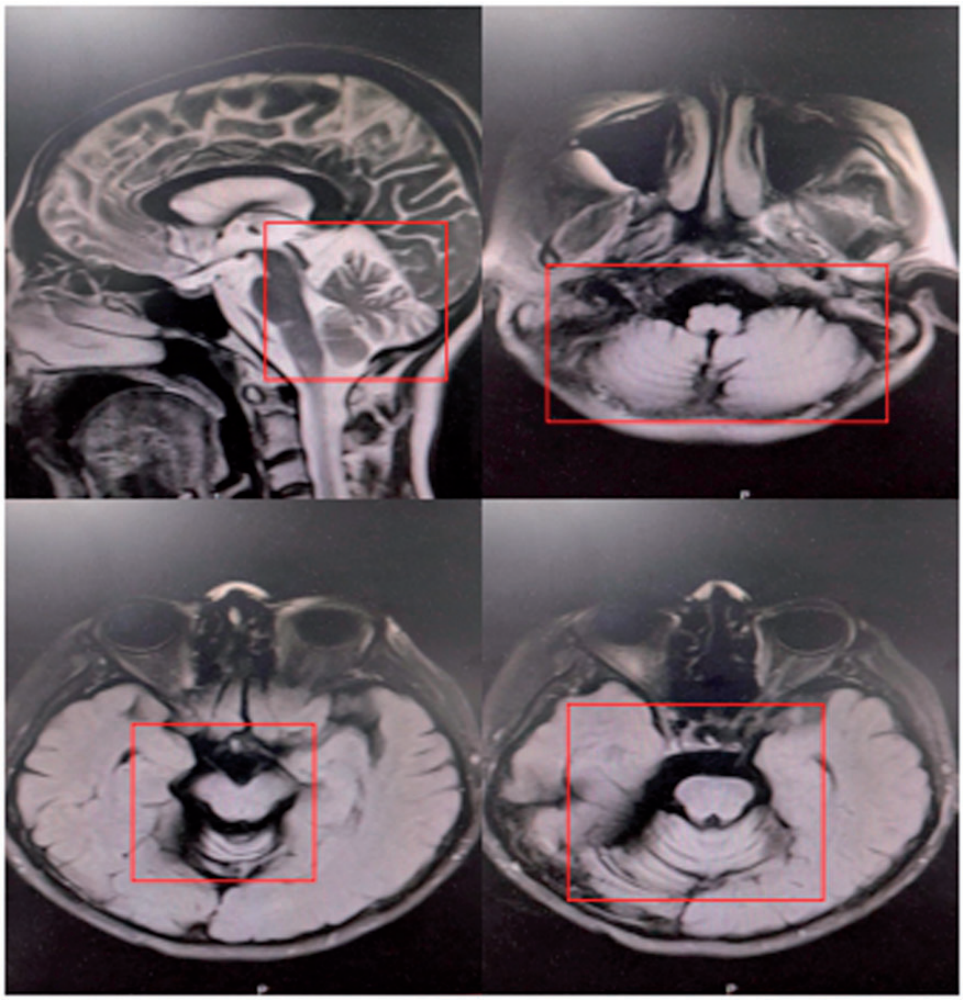

Figure 1. Magnetic resonance images of a 60-year old female patient who presented with syncope for 1 year combined with dizziness for 1 day. The patient had been diagnosed twice with transient ischaemic attack during the previous 6 months. The images show a widening of the cerebellar sulcus and mild brain atrophy. All four figures show cerebral atrophy (box and arrows).

patient's medical history, physical signs and auxiliary examinations, MSA-C type was considered.

Discussion

Multiple system atrophy is a neurodegenerative disease characterized by autonomic nervous system dysfunction, cerebellar ataxia and dopa-low-reaction Parkinson-like symptoms.[6] MSA is mainly divided into two clinical subtypes: MSA-C with cerebellar ataxia as the main disease; and MSA-P with Parkinson-like symptoms.[3,4] In 2008, the revised international consensus regarding the clinical diagnosis of MSA highlighted four groups of clinical features: (i) autonomic dysfunction or dysuria; (ii) Parkinson-like symptoms; (iii) cerebellar ataxia; and (iv) pyramidal signs.[3,7] Autopsies of 134 patients with MSA found that only 83 patients (61.94%) met the MSA pathological diagnostic criteria, while the remaining patients were diagnosed with Lewy body dementia, progressive supranuclear palsy (PSP) and Parkinson's disease.[8] Clinically, only a small number of diseases can be identified via cranial MRI, diffusion tensor imaging, proton magnetic resonance spectroscopy and electromyography.[9] A slit-like hyperintense putaminal rim observed on cranial T2-weighted images suggests MSA-P and the hot-cross-bun sign at the base of the pons on T2-weighted images suggests MSA-C.[1] T1-weighted images of the midbrain show that it is clearly atrophied and decreased in size.[1] The 'hummingbird' sign helps to distinguish it from PSP.[9] Therefore, ongoing data collection from autopsies performed on MSA cases or meta-analysis of existing studies is required.

Patients from European countries have MSA-P,[10] while those from Asian countries mainly have MSA-C.[11] In this current case, the onset of the patient's disease was concealed because her condition worsened gradually. The patient had experienced long-term constipation and urinary incontinence. Her skin scratch test was positive and the difference in her standing and prone blood pressures was 51/35 mmHg. All of these findings suggested autonomic dysfunction. She was unstable with the finger-to-nose test, positive with the heel-to-shin test and positive with Romberg, which suggested cerebellar ataxia symptoms prior to admission. Mild cerebellar atrophy was identified via cranial MRI, DWI and MRA, which also suggested cerebellar ataxia. Therefore, she was diagnosed with MSA-C. In 2011, it was demonstrated that the cognitive function of patients with MSA-C was significantly lower than that of normal controls using the Mini-Mental State Examination (MMSE) scale.[12] In 2015, a study found that patients with MSA-C had lower MMSE and Montreal Cognitive Assessment (MoCA) scores compared with patients with MSA-P and cerebellar cortical atrophy.[13] Neurologists are recommended to use the MMSE and MoCA scales for initial screening at outpatient clinics to determine the presence of mild cognitive impairment or dementia. They should then perform an auxiliary examination to detect and diagnose MSA at an early stage. This will enable neurologists to provide the patient with prompt effective treatment to improve their quality of life and long-term survival.

Declaration of conflicting interest

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