

Multiple deep white matter lesions mimic multiple sclerosis as an unusual complication of left atrial myxoma

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Abstract: We report the case of a patient who developed right hemiparesis with an acute onset 2 weeks before being transferred to our hospital. The brain MRI revealed multiple lesions in deep white matter; some were ovoid and perpendicular to the lateral ventricle, typical of cerebral demyelinating disease such as multiple sclerosis. The patient reported no history of hypertension, diabetes, dyslipidemia or other conventional vascular risk factors. The initial diagnosis was clinically isolated syndrome. However, low fever and petechiae in the extremities developed during hospitalization, which reminded us of cardioembolism. Echocardiography finally revealed a left atrial myxoma. The patient's symptom alleviated after resection of the tumor.

Keywords: Multiple sclerosis (MS), Myxoma, Clinically isolated syndrome (CIS)

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Introduction

Cardiac myxoma is the most common benign cardiac tumor, the vast majority (75–80%) of which are located in the left atrium.¹ There is a 2:1 female preponderance, and the age at onset is usually between 30 and 60 years.² In addition to cardiac symptoms, systemic embolization is the most common complication of myxoma. More than two-thirds of myxomatous emboli migrate to the central nervous system, leading to neurological manifestations including embolic or hemorrhagic strokes, aneurysms or metastases.¹ However, multiple deep white matter lesions mimicking those of demyelinating disease, especially multiple sclerosis (MS), caused by myxomatous embolization are rare and seldom reported.^{3,4}

Case report

A 62-year-old housewife was transferred in March 2013 to our institution after experiencing 2 weeks of right hemiparesis with a sudden onset. The neurological examination revealed decreased muscle strength (MRC grade 4) in her right arm and leg. Cranial magnetic resonance imaging (MRI) showed multiple lesions in the deep white matter. Some of the lesions were ovoid and perpendicular to the ventricles, as

described as 'Dawson finger', typical of MS. The biggest ovoid lesion located close to the left lateral ventricle was considered responsible for her right hemiparesis (Figure 1(A)–(F)). MRI of the spinal cord was normal. A lumbar puncture disclosed mild elevated protein (510 mg/l, normal range 150–450 mg/l) in cerebral spinal fluid (CSF). The CSF IgG-index was normal and oligoclonal bands (OB) were negative. The results of serum ANA, ENA, ANCA were normal. The computed tomography angiography (CTA) of brain and neck revealed no artery stenosis or malformation. The patient reported no history of hypertension, diabetes, dyslipidemia or any possible occupational exposure. Taking the impressive lesions on MRI into account, an initial diagnosis of clinically isolated syndrome (CIS), that is, the first episode of relapsing–remitting MS was considered.

However, the patient developed low fever and petechial rashes in the extremities during hospitalization. These petechiae were dark red, flat to the touch, and did not lose color when pressed. These phenomena could not be explained by MS, but reminded us of cardioembolism. An echocardiography was done immediately, which finally revealed a left atrial myxoma (Figure 1(G), (H)). Therefore, the diagnosis was

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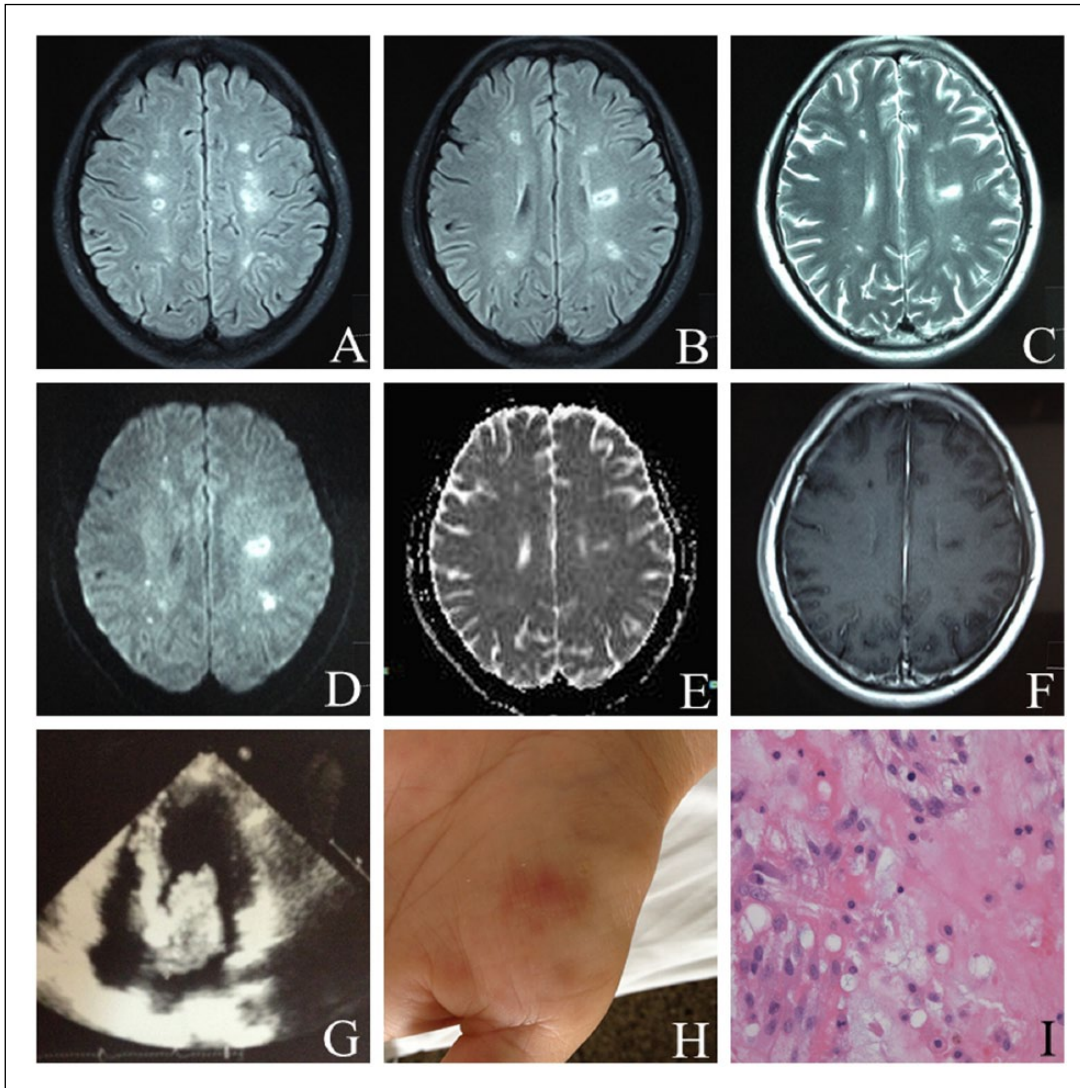


Figure 1. (A, B) MRI-Flair images showed multiple deep white matter lesions close to lateral ventricles. (C) Periventricular lesions on T2-weighted imaging (T2WI). (D) Some lesions were shown as high signals on diffusion-weighted imaging (DWI). (E) The apparent diffusion coefficient (ADC) was elevated in the lesion perpendicular to the left lateral ventricle. (F) No enhancement on Gadolinium-enhanced T1-weighted imaging. (G) Echocardiography showed a left atrial myxoma. (H) Petechial rashes in the patient's palm. (I) Photomicrograph of the histopathologic specimen showed spindled and stellate cells with eosinophilic cytoplasm and round to slender nucleus embedded in myxoid matrix (H&E, $\times 400$).

corrected as embolic stroke due to myxoma. The tumor was subsequently resected and pathologically confirmed (Figure 1(I)). After the surgery, the fever and rash gradually vanished and the patient's neurological disability alleviated gradually after several weeks' rehabilitation.

Discussion

Myxomas are generally thought to originate from multipotential mesenchymal cells of the endocardium. Cardiac myxomas are the most common primary

cardiac tumor in adults, representing as many as 83% of all primary tumors of the heart.⁵ They are found more commonly in young adults with stroke or transient ischemic attack (1 in 250) than in older patients with these problems (1 in 750).⁶ There is a 2:1 female preponderance, and the age at onset is usually between 30 and 60 years.² The sex ratio and onset age is similar to that of relapsing–remitting MS population.

Left atrial myxomas produce symptoms via the following principal mechanisms: obstruction of the mitral valve (dizziness, palpitations, dyspnea, and

congestive heart failure), systemic embolization (peripheral or cerebral), and constitutional symptoms (fever, fatigue, and weight loss). Neurologic symptoms have been reported in 26–45% of patients, with embolic infarct being the most frequently observed event.^{7–9} These may be the result of embolization of fragments of the myxoma itself or due to surface thrombi. Neuroimaging findings of myxomatous cerebral embolism may present as single or multiple vascular territory ischemic events distinctive for a proximal embolic source.⁸ However, as in this case, embolic lesions of deep periventricular white matter mimicking those of MS are rare.^{3,4}

We consider the reasons why the patient was initially diagnosed with CIS as follows: she had no risk factors of cerebral vascular disease; an acute onset could be explained by an autoimmune inflammation event; the deep periventricular lesions, some of which were consistent with the description of ‘Dawson finger’; the age and sex of the patient in accord with the demographic characteristics of MS population.

The key points leading to the suspicion of cerebral embolism were low fever and embolic rashes in the extremities. These nonspecific systemic symptoms and minor embolic phenomena may be easily overlooked, but are of great help to differential diagnosis. Negative CSF OB and no infratentorial and spinal cord lesions were also clues that do not support demyelination. Furthermore, the elevation of apparent diffusion coefficient (ADC) value of the lesion (Figure 1(E)) does not exclude the possibility of ischemic stroke, as there is an increasing trend from reduction to pseudonormalization to elevation of ADC values at subacute to chronic time points (≥ 7 days from stroke onset) during the evolution of stroke.¹⁰ ADC maps might not be so useful in differentiating subacute ischemia and demyelination in this case.

In conclusion, it is recommended that when making the diagnosis of MS or CIS, particular caution should be taken to exclude any other possibilities that can also cause multiple deep white matter lesions. Careful observations of constitutional symptoms out of nervous system must be emphasized.

Conflicting of interest

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