Atrial myxoma: a rare cause of peripheral neuropathy

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Atrial myxoma is the most prevalent primary heart benign tumor of the mesenchymal origin with an approximate prevalence 0.0017-0.19% [1]. It is often associated with embolism and very rarely presents as peripheral neuropathy. This article reports a case of peripheral neuropathy in a patient with atrial myxoma.

The patient, a 36-year old Chinese male, had a history of six month of weakness in both legs which was associated with numbness in the lower limbs. These symptoms became more severe along with fatigue. They improved after rest, but never disappeared. Two weeks before admission, his symptoms became worse with difficulty in walking. The patient was otherwise well with no significant past history of illness except repeated headache. The patient was always able to control his bladder and bowels. Admission checkup displayed clear consciousness and ability to walk only with the support of others. He had body temperature 37.9 °C, pulse 80 beats/min, and arrhythmia. Cardiac checkup showed III/6 murmur in mitral area, an abnormal point of apex beat in lower left area, and an area of cardiac dullness extended to the lower left area. No tenderness was found in his arms and legs with joint mobility slightly restricted. His cranial nerve examination was normal. Neurological examination found normal muscle strength, muscle tone, sensory, deep and superficial reflexes in his arms. In his lower limbs muscle strength measurements were proximal (4+/5) and distal (3/5). His lower limb reflexes were depressed relative to upper limb reflexes. Laboratory determinations showed normal serum electrolytes. The level of C reactive protein (CRP) was 78 mg/dL, and that of erythrocyte sedimentation rate (ESR) 124 mm/h. Cerebrospinal fluid analysis was normal. CT brain scan showed symmetric white matter ischemia, most prominent in the basal ganglia. His magnetic resonance imaging showed hyperintense signal patches on T_2 WI in deep regions of both hemispheres, without obvious mass or reinforcing effects (Fig. 1). One echocardiogram showed a 36 × 24 mm sized left atrial mass arising from the atrial septum consistent with atrial myxoma. Another echocardiogram showed a 29 × 22 mm sized left atrial mass, left atrial mitral valve incompetence, instant current of regurgitation reaching 28 mL and left atrial enlargement with volume reaching approximately 90 mL. Lower limbs vascular ultrasound showed disordered lower extremity arterial blood flow spectrum, reduced blood flow and slowed bilateral dorsalis pedis artery flow velocity. Resistance index was significantly reduced. His left dorsalis pedis artery maximum flow velocity was reduced to 17.4 cm/s, and his right dorsalis pedis artery maximum flow velocity was reduced to 5.4 cm/s. An electroneurogram displayed prolonged motor nerve conduction velocities of bilateral tibial nerve (left 33.0 m/s, right 38.9 m/s), slowed sensory nerve conduction velocities of sural nerve (left 39.7 m/s, right 41 m/s), abnormal F-waves of lower limbs, disappearing H-reflex from tibial nerves. These parameters were normal in the upper limbs. Cardiac surgery confirmed the presence of a left atrial mass which was removed. Pathological findings showed a left cardiac myxoma with denatured, flat cells lining the surface of anterior mitral valve, and local surface granulation tissue hyperblastosis (Fig. 2). After the operation and treatment with anticoagulants and antibiotics, most of his complaints disappeared gradually: his lower limb weakness recovered rapidly, while hypoesthesia still remained. Three weeks after operation, his CRP returned to normal, and his ESR was reduced to 40 mm/h. A control vascular ultrasound study showed disordered lower limb arterial blood flow spectrum, reduced blood flow, left dorsalis pedis artery maximum flow velocity at 28.8 cm/s and a right dorsalis pedis artery maximum flow velocity of 12.4 cm/s.

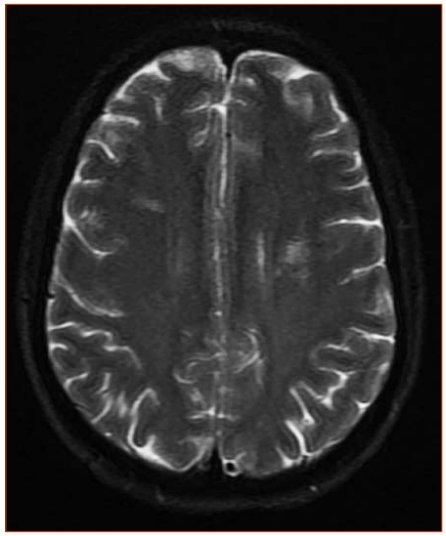

Figure 1. Magnetic resonance scan demonstrated bilateral white matter lesions compatible with ischemia.

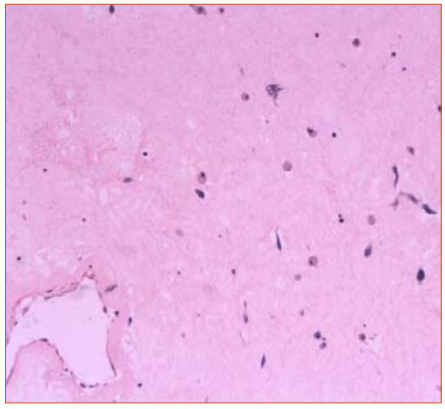

Figure 2. Pathological findings showed left atrial myxoma with denatured, flat cells lining the surface of anterior mitral valve, local surface granulation tissue hyperblastosis.

Atrial myxoma is a rare cause of embolic events and cardiovascular disease, including strokes [2,3]. Cardiac manifestations depend largely on the tumor's location and intracardiac extension, therefore cardiological symptoms and hemodynamic repercussions may not present in some cases, this making its diagnosis very difficult [4]. In this case, we only found a mitral murmurs, without any other cardiac complaints. Other systemic manifestations found in cases of atrial myxoma, such as arthralgia or myalgia, were not seen in this patient. Cardiac evaluation, especially echocardiographic examination in cases with stroke of unknown reason, are cru-

cial for the diagnosis. Noninvasive echocardiography will be the first choice for diagnosis, and has high accuracy in assessing tumor attachment, endocardial site localization, tumor's other descriptions without the risk of tumor fragmentation and embolization. The incidence of primary tumors of the heart is between 0.0017-0.19% in unselected patients at autopsy.

Kuroczynski et al [5] summarized 57 patients operated due to an atrial myxoma during the past 22 years. These authors reviewed the diagnostic methods, incidence of thromboembolic complications, valve degeneration, surgical repair techniques, recurrence and re-operation, and calculated the Kaplan-Meier survival curves. Surgical excision of the tumor appears to be curative, with few recurrences in long-term follow-up. After diagnosis, surgery should be performed urgently, in order to prevent complications such as embolic events or obstruction of the mitral orifice, and pathological findings confirm the diagnosis. Regarding our case, the progressive disappearance of peripheral neurological symptoms supports the idea that cardiac myxomas can be surgically cured [4] and that immediate and complete surgical excision would reduce the risk of recurrence. Follow-up examinations, including echocardiography, should be performed regularly.

Our patient presented as a demyelinating peripheral neuropathy predominating in the lower limbs. Santangeli et al [6] reported that the peripheral demyelinating neuropathy can be the first clinical presentation of cardiac myxoma. Our findings suggest that both systemic embolism and atrial myxoma-induced inflammatory phenomena can contribute to the pathophysiology of peripheral neuropathy. First, the clinical manifestations of asthenia and low temperature in both legs plus lower limbs vascular ultrasound findings suggested that vascular embolization related to embolus was one possible cause of the disease in our patient. Atrial myxomas may easily cause cerebral and peripheral vascular embolism, generating multiple cerebral and peripheral nervous symptoms [7,8] with incidence reaching 40%, even 80% if accompanied by infection [2]. In fact, although there were no definite symptoms, brain CT displayed ischemia, which might be induced by cerebral embolism. Second the high levels of CRP and ESR suggest that pro-inflammatory cytokines may be implicated in the pathophysiology of its peripheral neuropathy. Similarly, Mendoza et al [9] reported that systemic manifestations of cardiac myxoma seem to be caused by pro-inflammatory cytokines release, in particular interleukin 6 and alpha tumor necrosis factor.

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