A Case of Left Atrial Myxoma Whose Initial Symptom Was Finger Ischemic Symptom

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Summary

We experienced a 45-year-old Japanese man who was transferred to our hospital complaining of acute onset of pain and pallor in the right lower limb. Two years earlier, he had complained of repetitive pain at rest and pallor in the left third and fourth fingers. The physical exam and angiography demonstrated occlusion of finger arteries, however we could not reach final diagnosis. Acute arterial occlusive disease in the right lower limb was suspected. Transthoracic echocardiography demonstrated a gross tumor in the left atrium, which suggested left atrial myxoma. An emergency tumorectomy was successfully conducted. Pathologically, the fragile tumor and resultant thrombosis could have caused the patient’s peripheral circulatory failure at least two years prior to this episode. A rigorous systemic survey is important even when the ischemic symptom is localized in peripheral circulation.

Key words: Peripheral circulatory failure, Embolism, Acute arterial occlusion

It has been reported that the triad of left atrial myxomas are cardiac signs, constitutional signs and embolism, with cardiac signs the most common symptom of them all.1 The prevalence of embolism due to cardiac myxoma is reported as 33%,1 which is not a rare complication of this disease. However, the prevalence of peripheral embolism due to left atrial myxoma is extremely rare (0.9%).2 When seeing peripheral circulatory failure, the survey of embolic source is sometimes difficult unless systemic examination is performed. We report a case with left atrial myxoma, presented with peripheral circulatory failure and needed to consider pathophysiological cause of this disease.

Case Report

A 45-year-old Japanese man with a no notable medical history was transferred to our emergency room (ER) complaining of an acute onset of pain, pallor and numbness in his right lower limb. Two years earlier, he had repeatedly visited our hospital complaining of pain at rest and pallor in the left third and fourth fingers. The attending physician suspected acute arterial occlusive disease and collagen disease as differential diagnoses and checked electrocardiogram (ECG), ankle brachial pressure index, and toe brachial pressure index. Echocardiography was not performed at this point because the patient had never presented abnormal cardiac sounds, or abnormal ECG. A definite cause of the symptom was not identified at this point, and we could not reach final diagnosis. The differential diagnosis included diseases such as autoimmune disease, arteriosclerosis obliterans, and thrombosis due to atrial fibrillation which can present peripheral circulatory failure. An upper limb magnetic resonance angiography (MRA) showed a filling defect in peripheral distal interphalangeal joint in the left third and fourth fingers suggesting circulatory failure (Figure 1). The attending physician suspected TAO as tentative diagnosis based on the patient’s smoking history for many years and clinical symptoms, thereby oral and injectable prostaglandin E1 were administrated. After amelioration of the symptoms, he discontinued the therapy and resumed smoking, due to lack of adherence.

In the ER, the patient’s general condition was good, and cardiac murmur was not audible. His right femoral and popliteal arteries were weakly palpable, but the right dorsal artery was not palpable. We conducted transthoracic echocardiography (TTE), which revealed a huge cardiac tumor in the left atrium (Figure 2A). Brain CT showed a right cerebellar infarction (Figure 2B-1), and multiple small infarcts in cortical lesions (Figures 2B-2 and 2B-3). We detected some wedge-shaped defects on abdominal CT, which suspected a splenic infarction (Figure 2C). Lower limb CT showed occlusion of the proximal part of the right deep femoral artery (Figure 2D). The peripheral side was patent by the collateral circulation. We per-

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formed an emergency tumorectomy in order to prevent further embolic episodes. The surgery was successfully performed, and the resected sample was a cardiac tumor measuring 85×20×20 mm in size (Figure 3A).

On histopathological examination, the tumor was a typical cardiac myxoma, showing stellate or short spindle-shaped cells which had a heteromorphic nucleus in a myxoid matrix (Figures 3B, 3C). A portion of the tumor had a papillary structure, and fibrin was attached to the surface of the tumor (Figure 3D).

After the tumorectomy, antiplatelet therapy was resumed because the patient’s series of embolic episodes were caused by peripheral circulation failure due to the tumor and thrombosis, based on the patient’s imaging and clinical course. We performed lower limb CT after antiplatelet therapy was resumed (Figure 4). Compared to the preoperative CT (Figures 4A, 4C), the lower limb contrast CT showed improvement of the obstruction of the proximal part of the right deep femoral artery (Figure 4B), but obstruction of the right deep femoral artery was also found (Figure 4D). The postoperative course went well; the subjective symptoms were improved by postoperative day 3 and the patient was released from the hospital on the 6th postoperative day.

Figure 1. An upper limb magnetic resonance image (MRI) showed a filling defect in peripheral distal interphalangeal joint in the left third and fourth fingers.

Figure 2. Transthoracic echocardiography revealed a huge cardiac tumor in the left atrium (A). Brain CT showed a right cerebellar infarction (B-1), and multiple small infarcts in cortical lesions (B-2 and B-3). Abdominal CT showed some wedge-shaped defects (C). Lower limb CT showed occlusion of the proximal part of the right deep femoral artery (D).
Figure 3. The resected sample was a cardiac tumor measuring 85 × 20 × 20 mm in size (A). The tumor was typical cardiac myxoma, showing stellate or short spindle-shaped cells which had a heteromorphic nucleus in a myxoid matrix (B, C). A portion of the tumor had a papillary structure, and fibrin was attached to the surface of the tumor (D).

Discussion

We presented the case of a left atrial myxoma whose initial symptom was peripheral circulatory failure in the fingers. In this case, we rigorously examined the cause, and initially suspected TAO from medical history and physical examination as one of the differential diagnosis of this patient. However, to our knowledge, patients with TAO whose initial symptom is ischemic symptom of fingers are rarely reported.3) In this case, we should have considered some type of acute occlusive disease, but the priority of myxoma as a differential diagnosis is low, which account for only 1% of all artery occlusive disease.2) On the other hand, the prevalence of embolism due to cardiac myxoma is 33%.11 Embolic cerebral infarction is the most common complication among all embolic episodes in cardiac myxoma, and the incidence of embolic cerebral infarction is 29%.12 By contrast, peripheral embolism as an initial manifestation is not very common at 13%.11 If we meet a case with arterial thrombosis or occlusion, transthoracic echocardiography is a mandatory basic evaluation.

In a case series of 19 cardiac myxomas over 75 years of age, 15% of atrial myxoma were found by chance with echocardiography, but the 16 symptomatic patients (85%) had left ventricular failure (47%), positional symptoms (25%), pyrexia and poor general health (17%) or systemic embolism (17%).61 In this patient, MRA was effective in depicting a filling defect in peripheral distal interphalangeal joint in the left third and fourth fingers. High-resolution MRA has been reported to be effective in the assessment of ischemia of the palmar ulnar artery7) and finger artery flow in the Raynaud’s phenomenon.66 Therefore, high-resolution MRA could be a routine examination tool when arterial occlusive diseases are suspected.

The main mechanisms of embolism induced by cardiac myxoma are tumor embolism and thromboembolism. The pathology of cardiac myxoma can be classified into two types:9) Type 1 myxomas have a villous surface and soft consistency, and type 2 myxomas have a smooth surface and compact consistency.69 The prevalence of each of two types is 50%.11 Type 1 has been reported to cause tumor embolism more easily (Type 1, 29%-75%; type 2, 8%-12.5%).5-9) Thromboembolism is caused by a thrombus adhering to tumor’s surface.12) Tissue factor on a tumor surface and interleukin-611) and calretinin,11) which are secreted by myxoma cells activate the platelet aggregation activity. Our patient’s tumor was a type 1 myxoma morphologically. However, in this case, it is quite possible that the mechanism of peripheral circulatory failure was due to a thromboembolism, because the patient’s ischemic episode was improved by vasodilators before the tumorectomy. In fact, the resected specimen was covered by a thrombus on the tumor surface.

In conclusion, we treated a patient with a cardiac myxoma whose initial presentation was finger ischemic symptom. Although differential diagnoses of this case included TAO, the fragile tumor and resultant thrombosis could have caused the patient’s peripheral circulatory failure at least two years prior to this episode. In such a case, a rigorous systemic survey of the embolic source is important to clarify the pathophysiology.
Disclosures

Conflicts of interest: The authors declares that there is no conflict of interest.

References


Figure 4. Preoperative CT showed the obstruction of the proximal part of the right deep femoral artery (A, arrow), and patent distal part of the right deep femoral artery (C, arrow). Postoperative CT showed improvement of the obstruction of proximal part of the right deep femoral artery (B, arrow), and newly obstruction of the distal femoral artery (D, arrow).