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Cardiac Papillary Fibroelastoma - Source of Cerebral Embolism Treated with Intravenous Thrombolysis

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Study concept and design: Matijević and Habek. Acquisition of data: Matijević, Poljaković, Ilić, Čikeš and Habek. Analysis and interpretation of data: Matijević, Poljaković, Ilić, Čikeš and Habek. Drafting of the manuscript: Matijević. Critical revision of the manuscript for important intellectual content: Matijević, Poljaković, Ilić, Čikeš and Habek. Administrative, technical, and material support: Matijević, Ilić, Čikeš and Habek..

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Abstract

Objectives: We present the case of a 41-year-old man with sudden development of left hemiparesis due to infarction of the right middle cerebral artery, successfully treated with intravenous alteplase.

Methods and Results: Transthoracic echocardiography showed small mass in the left ventricle, the patient underwent surgical operation, and the histological examination confirmed the diagnosis of the papillary fibroelastoma.

Conclusion: It remains to be investigated whether heart ultrasound should be included as an evaluation prior intravenous thrombolysis in selected patients with stroke because of apparently increased risk of bleeding. However intravenous thrombolysis should not be postponed due to lengthy investigation because of its potential in reducing morbidity of stroke patients.

Causes of ischemic stroke in young differ from those in older patients.[1] As intravenous thrombolysis, as the only currently widely available treatment for stroke must be performed within 3 hours from symptoms onset, usually there is no time to exclude all these conditions which could potentially be harmful.

Case report

A 41 year old man, without history of serious illness and known cerebrovascular risk factors was admitted after sudden development of left hemiparesis. Brain multi-sliced computed tomography (MSCT) showed no early stroke signs (Figure 1A). NIHSS was 13, and according to our protocol we started thrombolysis with intravenous tissue plasminogen activator (tPA), 105 minutes after symptoms onset. Twelve hours later follow-up MSCT showed infarction in the territory of the right MCA, without bleeding (Figure 1B), and NIHSS was 2. Further investigation showed hypercholesterolemia, doppler sonography was normal, coagulation tests were normal. Electrocardiogram showed no signs of atrial flutter. Two days after admission transthoracal echo (TTE) visualized a small (1,5 x 0,9 cm) mass on the gracile peduncule connected to posteromedial papillary muscle and tendinous chords (Figure 2). An urgent operation was successfully performed, without neurological deterioration. Histological examination of the removed mass was consistent with papillary fibroelastoma (PFE) (Figure 3). Follow-up cardiac status, including control TTE was normal, and the patient was transferred to a rehabilitation facility.

Discussion

Cardiac PFE is a rare primary cardiac tumor and, unlike myxoma, the embolization is uncommon.[2] Mostly it is described in patients over 50 years old, with no sex preference, has no symptoms or becomes evident during investigation after stroke.[2,3] However, although rare, stroke is possible sequel of cardiac PFE. A fibrin thrombus attached to the circumference of the PFE, as well as the PFE itself, can be a source of embolic material.[2] Similarily, emboli from atrial myxomas may be composed of either thrombus, tumor itself, or both.[4] In either case, tumor emboli without fibrin thrombus are unlikely to lyse with thrombolytic therapy. Despite this, anticoagulant therapy is recommended for stroke patients with PFE up to the day of the surgical resection.[5] Clinical symptoms and physical examination in presented patient did not suggest a diagnosis of tumor emboli. There are 2 reports about intra-arterial thrombolysis in patients with atrial myxoma and PFE, which were successfully recanalized,[3,6] and only one case of myxoma in which an intravenous thrombolytic agents have been used with complete recovery of neurological deficit and without complications.[7] Another report showed a patient treated with intravenous tPA who developed hemorrhage remote from the location of ischemic stroke.[8] These authors suggested that intra-arterial local thrombolysis may be a preferable alternative because of greater risk of hemorrhage from occult tumor emboli or microaneurysms.

Although, in our patient, MSCT in a control interval visualized an infarction in the territory of the right middle cerebral artery, neurological recovery was almost complete. Our patient did not have hypertension, diabetes, had negative family history for cerebrovascular disease and he didn’t smoke. Coagulation tests and repeated electrocardiogram were normal, as well color-doppler ultrasound of carotid and

vertebral arteries. We are routinely performing TTE in all patients with stroke, which in this case revealed a small mass on the gracile peduncule connected to posteromedial papillary muscle and tendinous chords. In a case-control study, the sensitivity of TTE for PFE was 61.9%, while that of transesophageal echocardiography (TEE) was 76.6%.[9] Because the tumor was clearly visualized by TTE, and there were no signs of other cardiac abnormalities we did not perform TTE. Based on above mention tests and absence of all other stroke risk factors we concluded that PFE was source of emboli and cardiac surgery was performed. The effectivnes of intravenous thrombolysis can be explained that a fibrin thrombus was attached to the circumference of the PFE, which led to significant improovement in this case. Different histological properties of myxomas and PFE may, in part, explain the different propensity for bleeding after intravenous thrombolysis.

In conclusion, cardiac tumor emboli are very rare, but the first clinical presentation can be neurological. PFE is a rare but potentially treatable cause of embolic stroke. There are no randomized controlled studies of intravenous thrombolysis in patients with cardiac tumors due to relative small number of patients so risk of bleeding is hard to define. To the best of our knowledge, this case is a first reported case of PFE successfully treated with intravenous thrombolysis. Although one case report showed bleeding after intravenous thrombolysis in patient with cardiac myxoma, the benefit of this therapy far outweighs the risk, so intravenous thrombolysis should not be postponed due to lengthy investigation because of its potential in reducing morbidity of stroke patients.

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Figures

Figure 1. A) Brain CT on admission; B) Brain CT 12 hours after thrombolysis showing infarction in the territory of the right MCA, without bleeding.

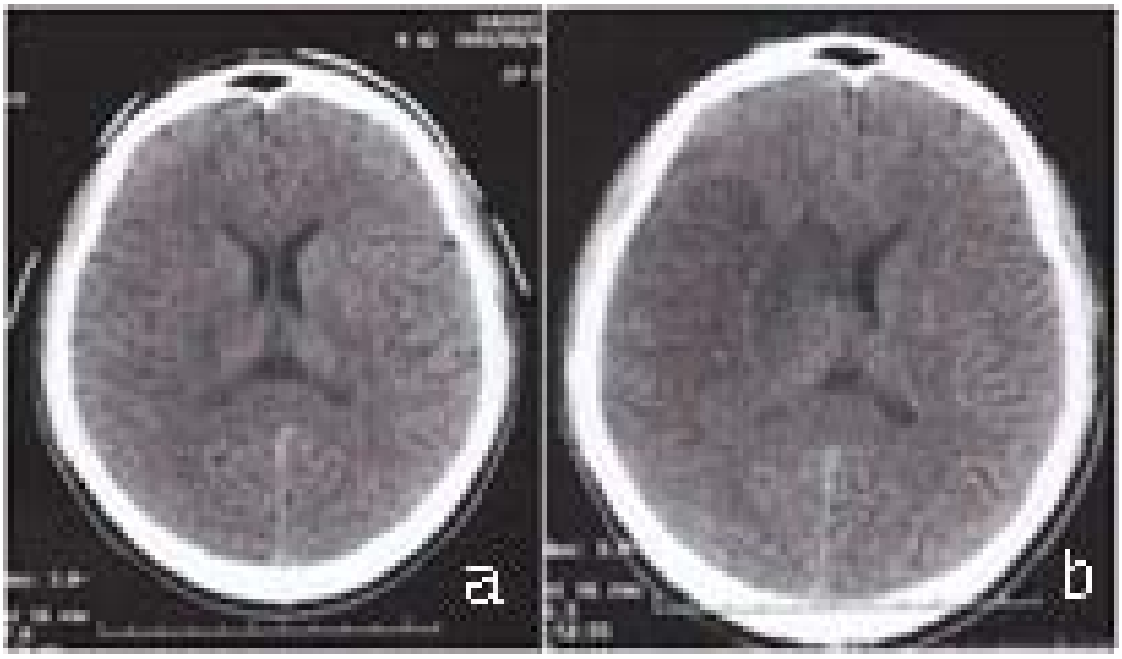

Figure 2. Transthoracal echocardiogram, apical three chamber view showing floating tumor in the left ventricular cavity. FE = tumor (fibroelastoma); LV = left ventricle, LA = left atrium; Ao = aorta; RV = right ventricle.

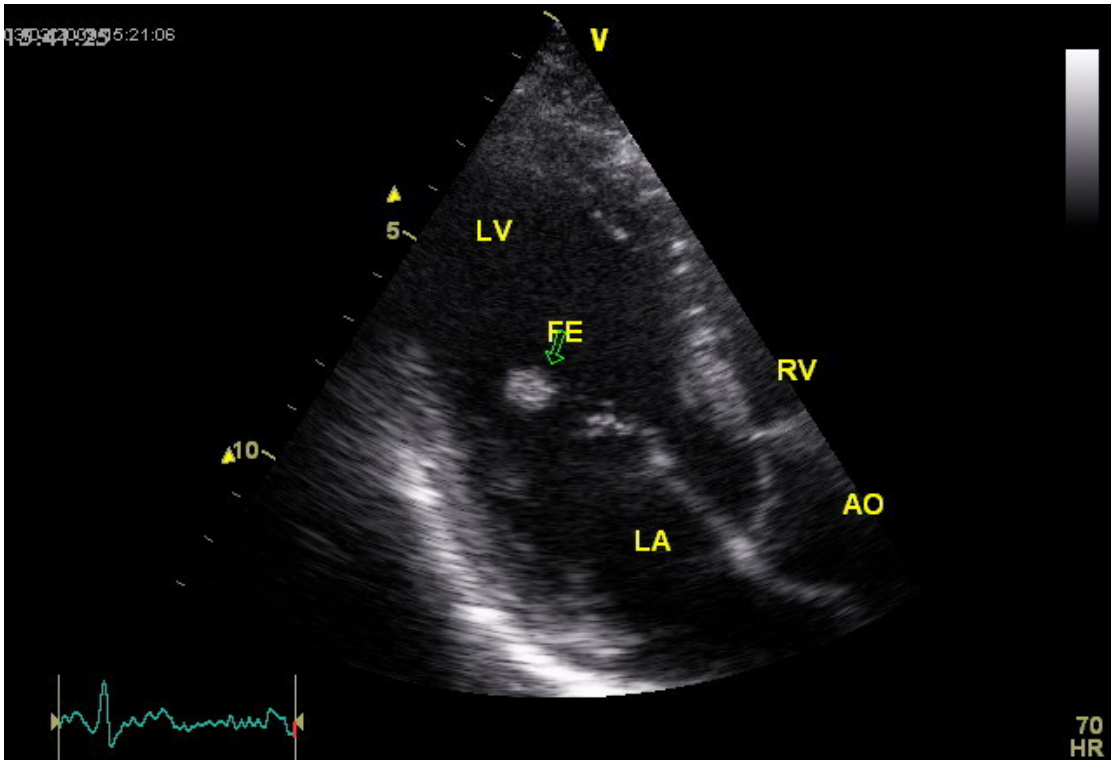

Figure 3. Histological examination shows core of connective connective tissue surrounded by endocardic cells (H&E).