Papillary fibroelastoma, an unexpected cause of stroke

Outcomes after acute reperfusion therapy

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We present 3 cases of cardiac papillary fibroelastoma (PFE) revealed by acute ischemic stroke (table).

Case 1 is a 38-year-old man who presented with sudden expressive language disorder and right hemiparesis. CT angiography showed a left middle cerebral artery (MCA) occlusion. Acute reperfusion therapy with endovascular thrombectomy (EVT) was achieved successfully. He was monitored in a stroke unit. The aspirated tissue was macroscopically congruent with a fibrin clot but the material was insufficient for pathologic analysis. A brain MRI performed a few days after admission showed a subacute ischemic stroke (figure, A). Blood tests, including hypercoagulability and autoimmunity assays, were normal or negative. Serial EKG and EKG Holter showed sinus rhythm. Transthoracic echocardiography (TTE) revealed an intracardiac mobile mass, with no history of fever or general malaise and negative blood cultures, suggestive of a primary cardiac tumor. Transesophageal echocardiography (TEE) (figure, B) confirmed the presence of an 11-mm pediculate mass bulging from the auricular surface of the mitral valve, with no valve or cardiac dysfunction. The suspicion of a primary cardiac tumor was supported.

Case 2 is a 64-year-old man who presented with sudden left hemiparesis and dysarthria. CT angiography showed a right MCA occlusion. Endovascular acute reperfusion was attempted, but when the arteriography was performed, there had been a distal migration of the thrombus. He was monitored in a stroke unit. A brain MRI showed an established stroke (figure, A). Blood tests and EKG Holter were normal. TTE showed a pediculate mobile mass on the mitral valve. TEE (figure, B) confirmed its presence, 14 mm, in absence of valve dysfunction. There was no history of fever, raising the suspicion of a primary cardiac tumor.

Case 3 is a 73-year-old man who presented with sudden expressive language disorder and right hemiparesis. There was no established stroke or arterial occlusion on CT scan/CT angiography. IV alteplase (recombinant tissue plasminogen activator [rtPA]) was administered in a stroke unit, with a good response. The brain MRI demonstrated a left temporoparietal stroke (figure, A). Blood tests were normal. Serial EKG and EKG Holter showed sinus rhythm. TTE revealed an auricular mass. TEE (figure, B) defined a 9-mm pediculate mass with frayed edges bulging from the left atrial appendage, suggestive of a primary cardiac tumor. There was no cardiac dysfunction.

In all 3 cases, a multidisciplinary team decided on surgical treatment of the intracardiac masses by scheduled surgical excision 2 weeks after the ischemic stroke. Simple excision of the tumor was performed in cases 1 and 3. Excision with biologic valve replacement was needed in case 2. No major surgical complications were reported. Pathologic examination of the surgically removed pieces confirmed the diagnosis of PFE (figure, C).

PRACTICAL IMPLICATIONS

Cardiac papillary fibroelastoma is an uncommon cause of cardioembolic stroke with the potential to achieve a good outcome after acute reperfusion therapies (both IV fibrinolysis and endovascular thrombectomy) and surgical removal of the tumor as soon as possible after stroke.

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Table Principal features of cases 1, 2, and 3

	Case 1	Case 2	Case 3
Age, y/sex	38/M	64/M	73/M
Clinical presentation	R hemiparesis and expressive language disorder	L hemiparesis and dysarthria	R hemiparesis and expressive language disorder
Brain CT scan at admission^a	No established stroke	No established stroke	No established stroke
CT angiography at admission^a	Left middle cerebral artery occlusion	Right middle cerebral artery occlusion	No arterial occlusion
NIHSS score at admission	6	4	16
Acute reperfusion therapy	Endovascular thrombectomy	Not possible due to distal migration of the thrombus	IV alteplase (tPA)
NIHSS score after 24 hours	0	0	0
Brain MRI^b	L caudate and putamen nucleus subacute ischemic stroke	R precentral subacute ischemic stroke	L temporoparietal subacute ischemic stroke
Transthoracic echocardiogram	Mitral valve mobile mass	Mitral valve mobile mass	Auricular mass
Transesophageal echocardiogram	11-mm pediculate mass on the auricular surface of the mitral valve	14-mm pediculate mass on the auricular surface of the mitral valve	9-mm pediculate mass with frayed edges on the left atrial appendage
Surgical treatment	Simple excision of the tumor	Excision of the tumor and biologic valve replacement	Simple excision of the tumor
Pathologic examination	Papillary fibroelastoma	Papillary fibroelastoma	Papillary fibroelastoma

Abbreviations: NIHSS = NIH Stroke Scale; rtPA = recombinant tissue plasminogen activator.

^a Brain CT scan and carotid and cerebral artery CT angiography were performed at admission to the emergency department in all 3 cases.

^b Brain MRI was performed during the first week after stroke in all 3 cases.

Discussion

The frequency of primary cardiac tumors is very low (0.02%)^{1,2}; hence, a primary cardiac tumor embolization is a very rare entity. Cardiac PFE are the second most common primary cardiac tumors (10%)^{1,3} after myxomas.^{1,2,4} Most PFE are found incidentally, but when they are symptomatic, stroke or TIA is the most common clinical presentation.^{2,5} Echocardiogram usually demonstrates a small (≤ 1 cm) mobile mass arising from the mitral valve (less frequently from the aortic or tricuspid valve or auricular or ventricular surface), with a frayed pattern near the edges, correlated with the papillary projections of the tumor.^{1,2} Pathologic examination of the surgical piece confirms the diagnosis,² as it did in our cases. The definitive and curative treatment for symptomatic PFE is surgical excision of the tumor, performed as soon as possible to diminish the risk of early recurrences.^{1–6} Prognosis after surgical removal is excellent.² However, patients with known fibroelastoma may still have a stroke while waiting for cardiac surgery. Furthermore, some patients could reject surgery or not be surgical candidates. There are limited data about acute stroke reperfusion therapy outcomes due to PFE embolization.⁶ The underlying pathologic process is difficult to achieve, since emboli could arise either from the papillary fronds of the PFE or from the thrombi formed on the surface of the tumor.^{2,3,5} In our cases, the good clinical outcomes (NIH Stroke Scale score 0 in all cases at discharge), related to the use of IV rtPA in case 3, to EVT in case 1, and the probable spontaneous recanalization of

the thrombus in case 2, suggest that the underlying pathologic process was a thrombus formed on the surface of the PFE. In this setting, acute reperfusion therapies could improve outcomes in patients with acute stroke with known and unknown PFE. No complications derived from IV rtPA or EVT were reported. This case series suggests that IV rtPA or EVT have the potential to be beneficial in this population.

Author contributions

A. Moreno-Estébanez: drafting/revising the manuscript, contribution of vital reagents/tools/patients, acquisition of data. A. Luna Rodríguez: drafting/revising the manuscript, acquisition of data, study supervision. I. Ugarriza Serrano: drafting/revising the manuscript, contribution of vital reagents/tools/patients, E. Blanco Martín: drafting/revising the manuscript, acquisition of data, study supervision. T. Gonzalez-Pinto Gonzalez: drafting/revising the manuscript, study supervision. J. Larrazabal López: drafting/revising the manuscript, contribution of vital reagents/tools/patients. T. Perez Concha: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, acquisition of data, study supervision.

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(A) Fluid-attenuated inversion recovery MRI sequence of the brain shows a subacute ischemic stroke in all cases, involving the left deep structures (caudate, putamen nucleus, and adjacent white matter) in case 1 (left), the right precentral region in case 2 (middle), and the left temporoparietal lobe in case 3 (right). (B) Transesophageal echocardiography reveals an 11-mm pediculate mobile mass (arrows) bulging from the auricular surface of the mitral valve in case 1 (left), 14 mm bulging from the auricular surface of the mitral valve in case 2 (middle), and 9 mm bulging from left atrial appendage in case 3 (right). Frayed edges of the mass can be seen in the image on the right. (C) Pathologic examination demonstrates avascular papillomas with a single layer of endocardial cells covering the papillary surface of a matrix of connective tissue (left, middle, and right).

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