Hamartomatous Malformation of the Left Atrium: A Rare Cause of Recurrent Transient Ischemic Attack

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Primary cardiac tumors constitute an infrequent heart pathology with a generally good prognosis. We present the case of a 45-year-old woman without a relevant medical history and with recurrent transient ischemic attacks of unknown origin. After follow-up with neurologic examination and repeated echocardiograms, an elongated mobile mass image was seen in the left atrial appendage. Surgical treatment consisted of resecting the mass by left atrial appendage excision through left atriotomy, showing an elongated image measuring 7 × 3 mm anchored in the pectineus muscle in the left atrial appendage. In the last one, the image was seen more clearly with an elongated, mobile mass of 7 × 3 mm anchored in the distal zone and of echogenicity similar to the surrounding tissues (Fig 1). The mitral valve was somewhat thickened with slight mitral insufficiency. The biochemical test results were all normal, and the tumor markers were negative. A chest CT scan was performed, but it did not provide any relevant information because of the small size of the mass and probably because the CT scan was not multislice. A prior cerebral magnetic resonance imaging (MRI) and CT scan, in the patient study, had revealed findings compatible with an earlier cerebral attack in the area of the right posterior cerebral artery, with no signs of cerebral vascular pathology.

The suggested diagnosis was a benign cardiac tumor provoking thromboembolic phenomena with recurrent TIAs. Lacking any other apparent cause, we began surgery. Using median sternotomy and left atriotomy, we observed a tumor in the left atrial appendage. There was no clear plane of differentiation, and it appeared to affect the entire wall (Fig 2). The tumor was resected by removal of the atrial appendage using left atriotomy. The base of the atrial appendage was sutured with 4-0 monofilament polypropylene in a circular fashion and then with a running suture to face the endocardium. There were no complications in the postoperative period; the patient was transferred from the intensive care unit the day after the operation and discharged from the hospital on the fifth postoperative day.

Anatomopathologic examination revealed bundles of striated muscle mixed with mature adipose tissue and a foliate pattern showing fissures covered with a flattened-

Fig 1. Transesophageal echocardiogram. View of the auricle of the atrial appendage, showing an elongated image measuring 7 × 3 mm.

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cubic mesothelial epithelium positive for cytokeratin AE1/AE3. One of the fissures and most of the surface of the sample showed an endothelial lining (Fig 3). The diagnosis was hamartomatous-like malformation. Several years after the operation, the patient has not had any events, is asymptomatic, and does not require any drugs.

Comment

Cardiac hamartomas are truly rare lesions. In fact, references to this lesion in the literature are anecdotal. They are defined as a benign overload of mature differentiated cells that originate in the organ where they develop, but are placed in a disorganized manner. They are the result of an anomaly in embryonic cell development. The morphologic characteristics, location, and clinical signs and symptoms make it possible to divide them into five types: (1) rhabdomyoma, (2) hibernoma, (3) lipomatous hamartoma of the interatrial septum, (4) lipomatous hamartoma of the cardiac valves, and (5) hamartoma of mature cardiac myocytes [1–3].

One of the main problems derived from cardiac tumors is their potential for forming emboli, which has been estimated to be 12% to 45% [4]. In our case, the patient experienced six TIA episodes in 3 years. El Bardissi [5] reported that, in a series of 323 patients with primary cardiac tumor, the risk of an embolic event was related to location in the left atrium, small tumor size, and states without many symptoms, such as those that did not exhibit mitral insufficiency [5]. All these factors were present in our patient. The American Heart Association guidelines propose that during evaluation of an embolic event, intracardiac masses should be suspected and appropriately pursued with echocardiography based on a suggestive clinical presentation [6]. It should be pointed out that in the case of hamartomas, the diagnostic performance of the echocardiogram can be low depending on the location; this makes it necessary to complete the study with other imaging tests.

Treatment is based on the clinical signs and symptoms and the imaging tests. Such treatment, depending on the suspicion of malignancy and on the clinical data, is generally surgical. In tumor pathology, complete excision is essential to guarantee a good therapeutic result, which is curative in the case of benign pathology. In our patient, other options (e.g., percutaneous closure of the left atrial appendage) were not considered because of the tumor. The prognosis, generally good, depends on the nature of the tumor, its size and its location, which are in turn factors that condition the clinical signs and symptoms. Atrial locations are normally associated with supraventricular arrhythmias and Wolff-Parkinson-White syndrome, whereas ventricular lesions are associated with malignant arrhythmias. Symptoms can be completely absent in both cases.

Turning to imaging tests, echocardiography is the technique of choice in the initial study of cardiac masses because of its availability, low cost, and harmlessness. It makes it possible to detect, locate, and define the shape, extension, mobility, and anchor point, as well as to ascertain the solid or cystic nature of the lesion [6]. The information provided by this technique should be completed with other tests such as CT scan or MRI, which allow an assessment of the structure and hemodynamic component, obtaining static and dynamic sequences. Finally, an angiograph may be indicated to demonstrate a tumor blush in the malignant pathology study or in the study of coronary compression by the tumor. Consequently, the combination of various imaging tests, while emphasizing the role of the echocardiogram because of
its characteristics [6], has a decisive role in the diagnosis and treatment decisions of this pathology.

References

Infant Repair of Massive Aortic Aneurysm With Prosthetic Valved Conduit

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A 4-month-old child with severe infantile Marfan syndrome underwent successful repair of an extremely dilated aortic root and severe aortic valve insufficiency using a prosthetic valved conduit.


Marfan syndrome (MFS) is a progressive multiorgan connective tissue disease that typically manifests in adolescence or early adulthood. Rarely, MSF may be diagnosed in neonates, and these patients frequently suffer from a more severe form of the disease than those diagnosed later in life [1]. One of the most common cardiovascular manifestations of this disorder is progressive dilation of the aortic root [2], the consequences of which account for as many as 80% of premature deaths in these patients [3]. Prophylactic surgery is, therefore, indicated for patients with aortic root dilation and has been shown to improve the prognosis in young children with MFS [1]. The Bentall procedure [4] is the procedure of choice when the aortic valve cannot be salvaged [5]. Although this operation has been reported in young children [1,6,7], this case represents the youngest patient reported in the literature.

A 4-month-old boy was diagnosed at birth (weight 3.8 kg) with infantile MFS. His initial echocardiogram demonstrated mild mitral regurgitation, mild aortic regurgitation, and significant aortic root dilation, measuring 1.7 cm in diameter (Z-score 5.64).

At 7 weeks of age (weight 4.61 kg), the patient started to show signs of heart failure. A repeat echocardiogram demonstrated an increased aortic root dimension, now measuring 2.4 cm (Z-score 10.03). It also revealed mildly depressed left ventricular function, moderate aortic insufficiency, and moderate mitral regurgitation due to mitral prolapse. Anticoagulation medical therapy, beta-blockade, and angiotensin-converting enzyme inhibition were initiated. Over the next several weeks, the patient improved symptomatically. However, by 14 weeks of age (5.32 kg), the aortic root dimension had increased to 3.0 cm in diameter (Z-score 13.83), the aortic insufficiency had progressed, and the left ventricular function appeared more depressed (Fig 1). A subsequent magnetic resonance image confirmed significant dilation of the aortic root (Fig 2), with a Z-score of the sinuses of Valsalva between 8 and 12, with associated severe aortic and mitral insufficiency. In addition, the patient was also incidentally discovered to have a vascular ring, in the form of a right aortic arch and aberrant left subclavian artery. Secondary to the rapid progression of his aortic root dilation, the decision was made to surgically intervene.

At the time of surgery, at 15 weeks of age (weight 5.32 kg), the patient’s aortic root measured in excess of 3.2 cm (Z-score 15.30). Intraoperatively, the aortic valve was trileaflet but was very dysplastic and considered not to be repairable. A composite root replacement using a 19-mm St. Jude prosthesis (St. Jude Medical, St. Paul, MN) was performed. A mitral valve repair was completed using a 28-mm Carpentier-Edwards annuloplasty ring (Edwards Lifesciences, Irvine, CA), and the vascular ring was also repaired. Postoperatively, the patient experienced low cardiac output and underwent elective placement on extracorporeal membrane oxygenation in the intensive care unit. He was weaned off extracorporeal membrane oxygenation uneventfully after 4 days, and discharged home on the 18th postoperative day with no further complications.

Three months after surgery, the patient is thriving (weight 7.06 kg). His electrocardiogram is normal, and his echocardiogram demonstrates improving ventricular function, a well-functioning aortic prosthesis (Fig 3), and trivial mitral insufficiency.

Comment

Aortic aneurysms are rarely encountered in infants and, therefore, not much attention has been paid to the