



A Rare Cause of Pediatric Stroke: Left Atrial Myxoma

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Abstract

We describe a case of sudden-onset left-sided hemiparesis and dysarthria in a five-year-old boy. Acute vascular malformation bleeding or ischemic stroke was suspected. Neurological examination three weeks after the initial event revealed mild residual facial paresis. Brain angiography ruled out a vascular malformation. A work-up echocardiogram revealed a 4-cm left atrial mass compatible with cardiac myxoma. Urgent surgical resection of the mass under cardiopulmonary bypass confirmed the diagnosis. Uneventful recovery followed surgical resection. In this report, we present a partially embolized left atrial myxoma that caused an acute ischemic stroke, which is rarely considered and encountered in the pediatric population.

Keywords

myxoma, pediatric, atrium, stroke, surgery

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Introduction

Primary cardiac tumors are rare in the pediatric population, with a prevalence ranging from 0.0017% up to 0.28% in different autopsy series.¹ Nadas and Ellison reported an incidence of 0.027% in 11,000 pediatric autopsies.² Benign lesions usually predominate, accounting for more than 90% of all pediatric tumors.³ Myxomas are rare (0%-5%) in pediatric patients and are usually diagnosed in older children and adolescents.

Atrial myxomas may present with a variety of symptoms related to the size and location of the tumor, and the majority of the patients would experience one or more of a triad of symptoms: atrioventricular valve obstruction, systemic embolization, and constitutional symptoms. However, early diagnosis may be challenging because of nonspecific symptoms ranging from congestive heart failure to neurological deficits. Therefore, these cardiac tumors are often misdiagnosed.^{1,4,5}

Despite the benign pathological pattern, its growth in a small infant heart can make it clinically very aggressive, requiring emergency treatment.⁶ We report a case of a partially embolized left atrial myxoma causing an acute ischemic stroke that was successfully resected.

Patient and Methods

A retrospective review from the medical record was performed. A permission to publish this case report was granted by the patient's family.

A five-year-old boy weighing 21 kg from a northeast rural area in Argentina, presented at the emergency department of the referring hospital with sudden-onset left-sided hemiparesis and dysarthria while playing at home. His medical history was unremarkable. Acute vascular malformation bleeding or ischemic stroke was suspected. Computed tomography (CT) revealed a right parietal hypodense lesion with mild mass effect. Magnetic resonance imaging (MRI) performed at the referring hospital the week of the event showed an extensive hyperintense right fronto, parietal, and cortico-subcortical lesion on T1-Fast Spin Echo (T1-FSE) and T2-Fluid attenuation inversion recovery (T2-FLAIR) sequences with gyriform pattern of contrast enhancement (Figure 1).

The patient was transferred 750 miles from referring hospital in rural area to our center 23 days after the onset of

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Abbreviations and Acronyms	
AVM	arteriovenous malformation
CT	computed tomography
2D	two-dimensional
3D	three-dimensional
MRI	magnetic resonance imaging

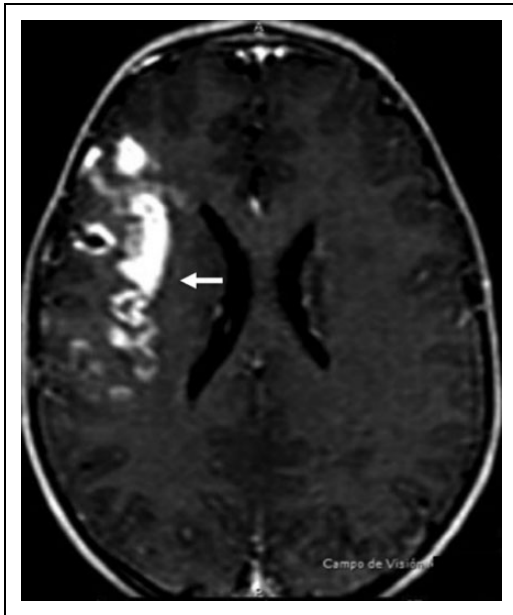


Figure 1. Cerebral magnetic resonance imaging (MRI) axial T1-Fast Spin Echo (T1-FSE) + gadolinium showing extensive subacute ischemic lesion compromising the right frontal cortex (white arrow).

neurological symptoms, due to social reasons and unclear stroke etiology for further neurological assessment and potential percutaneous treatment.

Upon admission, physical examination revealed stable vital signs. Cardiac auscultation included a regular heart rhythm with normal S1S2. There was a 2/6 tele-diastolic inflow murmur at the apex. Neurological findings revealed a mild residual left facial paresis.

Digital subtraction cerebral angiography performed to rule out a vascular malformation indicated the absence of brain arteriovenous malformation (AVM) and no abnormalities in both the middle and anterior cerebral arteries. Vertebrobasilar circulation was normal, and no microaneurysms were observed.

Biochemical hemostasis thrombophilic work-up was unremarkable. A Doppler color and three-dimensional (3D) echocardiogram revealed a pedunculated giant 4 × 4 cm left atrial tumor attached to the free wall of the left atrium (Figure 2). The mass prolapsed into the mitral valve annulus during diastole, resulting in mild to moderate inflow obstruction and mild mitral valve regurgitation.

After an extensive discussion of the differential diagnosis with cardiology, neurology, neurosurgery, and pediatric intensive care teams, we felt that urgent surgical resection was

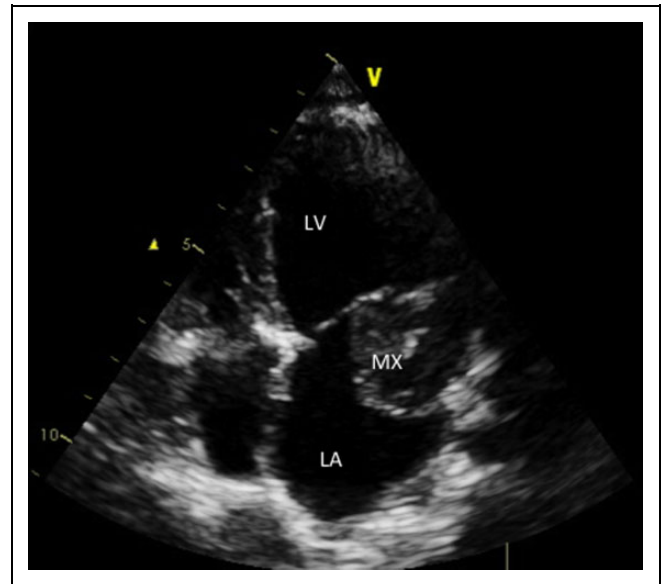


Figure 2. Two-dimensional (2D) echocardiogram long-axis apical view. Preoperative examination shows a round mass (MX) occupying the left atrium (LA) and prolapsing into the left ventricle (LV) during diastole.

indicated with the aim of preventing further potential embolization and devastating brain damage. Through a median sternotomy approach, cardiopulmonary bypass was instituted using aortic and bicaval cannulation. After cardioplegic arrest was achieved, the right atrium was longitudinally opened. The atrial septum was exposed and incised starting at the fossa ovalis, revealing a large mass, which was extremely fragile. The tumor was retracted and the pedicle, attached to the left atrial wall, was exposed. The implantation location was observed between the left atrial appendage and the left pulmonary veins. The friable and gelatinous mass was gently and carefully removed from the left atrial cavity, avoiding any fragmentation. The mass was completely excised including a full-thickness portion of the left atrial wall where the tumor pedicle was attached (Figure 3), and the gap was repaired with a 6-0 PROLENE suture. The mitral valve was examined and found to be competent. The atrial septum incision was closed using a running suture. Finally, the atrial free wall was closed and the patient weaned from cardiopulmonary bypass uneventfully.

Results

The patient was extubated in the operating room. Postoperative color Doppler two-dimensional (2D) and 3D echocardiography revealed a good left ventricular function, no residual intra-atrial mass, and mild mitral regurgitation (Figure 4). Electrocardiogram showed sinus rhythm. Postoperative course in the cardiac intensive care unit was uneventful, and the patient was discharged on day 5 postoperatively in an adequate clinical condition. Neurological postoperative recovery was complete.

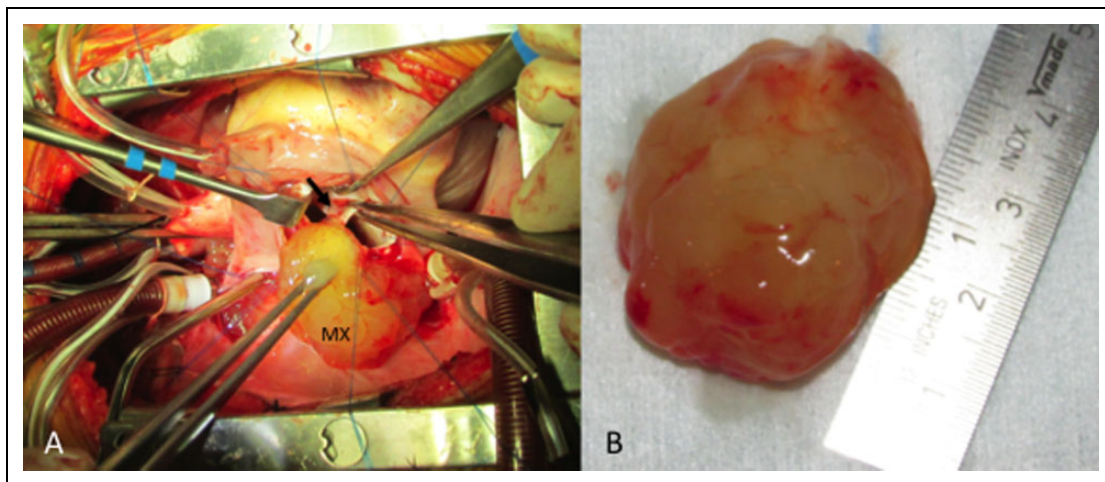


Figure 3. A, Intraoperative picture showing the left atrial myxoma (MX) being resected through the interatrial septum, showing the attachment to the left atrial free wall (black arrow). B, Gross pathological image of the excised mass. Of note, the villous surface and the gelatinous aspect of the tumor.

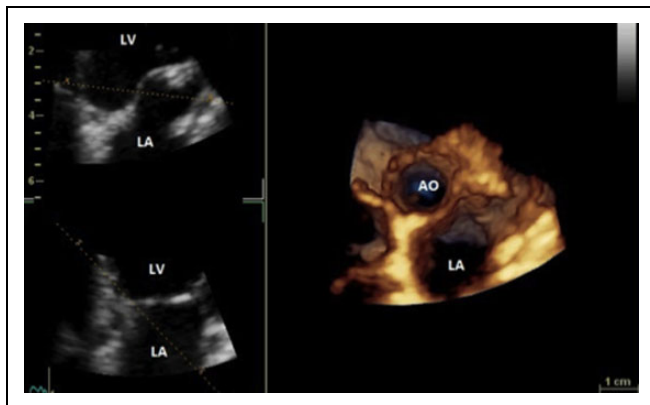


Figure 4. Echocardiogram modified view showing no residual lesion in the LA. Ao indicates aorta; LA, left atrium; LV, left ventricle.

Macroscopic pathologic examination of the mass revealed pediculate atrial myxoma of 4 cm in diameter (Figure 3). It was jelly like, multilobulated, rounded, and appeared as a “white-brownish” mass. At microscopic examination, the mass consisted of abundant myxoid matrix with numerous isolated “stellate” cells scattered in the ground substance.

Comments

Cardiac myxoma is reported to be rare in infancy and childhood.⁶ In 1972, Steinke et al reported a review of 23 cases of left atrial myxomas.⁷ Since then, to the best of our knowledge, 23 cases have been published in 11 papers including patients under 12 years old. This is the 24th patient reported.

About 75% of myxomas originate in the left atrium with the majority rising from the interatrial septum at the border of the fossa ovalis, but they can also originate, less frequently, from the posterior atrial wall, the anterior atrial wall, and the atrial appendage.⁵ In our patient, the tumor was originating from the left atrial free wall, which is less common.

The clinical spectrum can be wide and the majority of the patients present with one or more of a triad of symptoms, known as “Goodwin triad.” These include embolic phenomena, inflow chamber obstruction, and vague constitutional symptoms. Clinical manifestations secondary to inflow obstruction are most common, occurring in more than 50% of patients having atrial myxomas.⁴

Embolic phenomena occur in 30% to 40% of patients and involve the central nervous system, kidneys, coronary arteries, and extremities. The embolic fragments usually consist of myxomatous tissue and may also be related to the thrombus dislodged from the surface of the myxoma. Ischemic strokes and vision defects secondary to occipital lobe involvement have also been reported. The clinical presentation may vary from hemodynamic compromise to neurological deficits and sudden cardiac death.⁴ Our patient presented with signs and symptoms of acute stroke, which was of unknown origin until suspected and diagnosed at our institution three weeks after the acute event.

Nonspecific constitutional symptoms, such as fever, weight loss, and rheumatic pain, are commonly associated with atrial myxomas. Abnormal laboratory findings include anemia, thrombocytopenia, elevated erythrocyte sedimentation rate, and high serum levels of interleukin. These nonspecific and often vague symptoms result in late diagnosis in many cases.^{4,5,7} However, our patient did not present any of these findings.

Cardiac myxomas are typically diagnosed by Doppler color echocardiography, allowing a rapid and precise description of the intracardiac mass, giving the surgeon all the necessary information to plan and subsequently perform a safe and complete resection of the mass.⁶

The brain CT, MRI, and angiography are valuable modalities to determine the presence of central nervous system pathology, especially if neurological signs and symptoms are present. A high degree of clinical awareness would help to obtain a timely diagnosis. Although a cardiac myxoma is an

unusual source of a cerebral embolic event in children, case reports are well described in the pediatric population and, therefore, it should be part of the differential diagnosis. Neurological complications resulting from cardiac myxomas are seen in 20% to 35% of patients.⁸ In our case, the diagnostic work-up, including 2D color Doppler echocardiogram to assess possible sources of embolism, led us to discover this infrequent lesion.

After the diagnosis has been established, surgery should be performed promptly because of the possibility of embolic complications including sudden death. The treatment of choice is the complete surgical excision of the myxoma with removal of a substantial portion of healthy adjacent endocardial tissue. Survival after surgery has been reported to be excellent.¹ The importance of follow-up must not be underestimated. Although resection of atrial myxoma results in a rapid resolution of symptoms, long-term follow-up is required in young patients because of a reported recurrence rate of 5% to 7%.⁴ Although most cases of recurrence are thought to be related to incomplete resection, there is another group of patients with recurrence in multiple sites, which are associated with Carney syndrome that includes a constellation of myxomas, spotty pigmentation, and endocrine overactivity.⁸

We present this case to increase awareness and to stress early evaluation of secondary causes of ischemic cerebrovascular accident, other than the AVMs and hypercoagulability states. This case also exhibits the need for basic cardiac work-up in young individuals who present with stroke, to minimize further significant morbidity or mortality.

This case report demonstrates an unusual presentation of a cardiac tumor in a child where further neurologic damage was prevented by suspicion, identification, and surgical resection of the left atrial myxoma.

Declaration of Conflicting Interests

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