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Brief Report

Congenital giant aneurysm of the left atrial appendage in an infant

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Abstract Aneurysm of the left atrial appendage is a rare pathological condition. We describe the diagnostic work-up and surgical management of a child with giant congenital aneurysm of the left atrial appendage.

Keywords: Atrial appendage dilatation; aneurysm; off pump

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CONGENITAL LEFT ATRIAL APPENDAGE ANEURYSM IS a very rare congenital heart disease,[1–7] with potentially lethal complications.[1–3] We report a case of congenital left atrial appendage giant aneurysm, leading to left ventricular compression and progressive mitral valve regurgitation, which was successfully operated off pump by means of surgical excision.

Case report

A 3-month-old girl, with a body weight of 6.2 kilograms was referred to our department for surgical management of a giant compressive aneurysm of the left atrial appendage. The child was asymptomatic at birth, but a cardiac murmur was detected at routine physical examination. The chest roentogenogram showed a nonspecific cardiomegaly with an unusually prominent left heart border. Electrocardiogram showed regular sinus rhythm, with a heart rate of 146 beats per minute, and no episodes of arrhythmia or thromboembolism were ever reported. Soon after birth, a two-dimensional cross-sectional echocardiographic and Doppler examination revealed a large aneurysm of the left atrial appendage (45×60 millimetres), communicating with the left atrium through a large orifice (Fig 1).

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The patient was followed up clinically by serial echocardiographic examinations that showed progression of mitral regurgitation from mild to moderate. In addition, when compared with the first assessement, the most recent echocardiographic detections showed that the left ventricle was rightwardly dislocated, as if the enlarged atrial appendage was pushing on it. Thus, because of echocardiographic evidence of left ventricular dislocation and

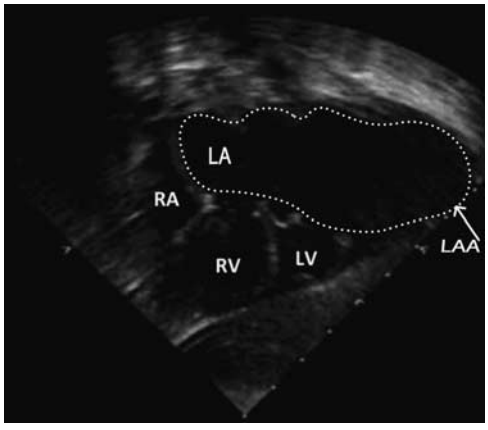

Figure 1.
Two-dimensional echocardiographic four-chamber view, showing the giant aneurysm originating from the left atrium (RA = right atrium; RV = right ventricle; LV = left ventricle; LA = left atrium, LAA = left atrial appendage; arrow pointing at the LAA aneurysm).

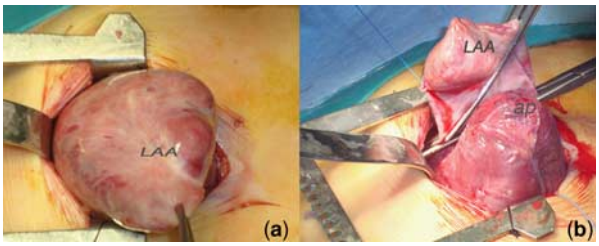

Figure 2.
*(**a**) Operative view of the aneurismatic left atrial appendage after mini sternotomy; (**b**) the LAA aneurysm is clamped off pump before excision (ap = apex of the left ventricle; LAA = left atrial appendage).*

progressively worsening mitral valve regurgitation, despite absence of symptoms, early surgical repair was indicated.

Through a mini sternotomy, the enlarged left atrial appendage was identified (Fig 2a). Intraoperatively, it was evident that the left atrial appendage compressed the left ventricle, displacing it to the right side of the chest. Coronary anatomy was unremarkable, and the coronary artery course appeared normal, with no evidence of kinking or distortion. The aneurysmatic left atrial appendage was clamped at its base (Fig 2b) and resected off pump. The left atrial appendage remnant was closed with a double-running 6.0 polypropylene suture uneventfully. No thrombus was found within the aneurysm. Post-operative course in Intensive Care Unit was uneventful, and the patient was extubated the day after operation. Her following clinical course in the wards was uncomplicated. No evidence of arrhythmias was detected at pre-discharge electrocardiogram. Histopathological examination did not reveal any evidence of malignancy. The child was discharged home in good general condition, 5 days after operation, on oral aspirin as anti-aggregation therapy for 3 months. Pre-discharge cross-sectional echocardiography showed a significant decrease of mitral valve regurgitation (1+). At 1-year follow-up, the patient is alive and growing well, without any medications, and without echocardiographic signs of either left atrial appendage dilatation or mitral regurgitation.

Discussion

Congenital aneurysms of the left atrium or the left atrial appendage are very rare entities of both anatomical and surgical fascination, and published literature documents only limited description of such cases and their management.[1–8] This case of giant congenital aneurysm of the left atrial appendage represents the only case ever reported at our institution in more than 40 years of activity. Morphologically, the genesis of the left atrial appendage aneurysm has been attributed to congenital dysplasia of musculi pectinati.[1] Congenital absence of pericardium has also been correlated with this anomaly.[5]

Life-threatening thrombo-embolic events[1–6] and supraventricular tachyarrhythmias are the most frequently reported complications.[1,8] Despite its congenital cause, symptoms usually do not arise until the second or third decade of life.[1–7] These aneurysms probably increase in size as the patient ages. Once they reach a larger size, they predispose the patient to supraventricular arrhythmias, cardiac dysfunction, atypical chest pain episodes due to compression of the left coronary artery or any of its divisions, an increased risk of intra-atrial thrombus formation, and systemic embolisation.[1–7] This did not occur in our experience, in which the patient had a complete diagnosis in early infancy. We believe that the early detection and surgical treatment have prevented from onset of these events, which are more common in older patients.[5,6]

Three-dimensional echocardiographic assessment has been reported as a valid technique for a fine diagnosis of this unusual anomaly.[8] However, in our experience, conventional two-dimensional echocardiographic assessment was accurate and allowed our experienced paediatric cardiologists to achieve a complete diagnosis, with no need for other diagnostic investigations. In particular, the colour Doppler echocardiogram was very helpful in diagnosis, by showing the exchange of blood between the left atrium and the left atrial appendage.

Strecker et al[9] have reported a similar case of a giant compressive aneurysm of the left auricle in a 1-day-old asymptomatic neonate, who underwent emergent surgery on cardiopulmonary bypass because of the presence of an intra-atrial thrombus, and was discharged home 1 month after surgery. In our experience, as the patient at birth was asymptomatic, clinical follow-up was decided. The child was monitored periodically by echocardiography, and surgical treatment was scheduled at 3 months of age. The operation was performed off pump, and post-operative course was smooth enough to discharge the patient home 5 days after surgery. The left atrial appendage aneurysm resection was feasible off pump, avoiding the deleterious effects of cardiopulmonary bypass, and the fast post-operative recovery allowed early discharge. The indication for surgery was left ventricular displacement by the left atrial appendage aneurysm. The surgical procedure was uncomplicated and did not require cardiopulmonary bypass. This quick recovery and fast discharge may have not been possible in the patient in the neonatal age.

We conclude that early diagnosis and early surgical resection of left atrial appendage aneurysm

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is a low-risk and effective procedure, which is mandatory to prevent the potentially life-threatening complications of this rare anomaly. However, surgical resection should be based on symptoms or echocardiographic signs of compression or intra-atrial thrombus, and avoiding cardiopulmonary bypass may be helpful to enhance a successful result and reduce post-operative stay.

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