Inversion of the left atrial appendage following cardiac surgery

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Inversion of the left atrial appendage is a rare complication after open-heart surgery. To our knowledge only 16 cases, besides that reported herein, have been described so far. Echocardiographically, the inverted left atrial appendage appears as a mass mimicking a thrombus, a vegetation or a tumor of the left atrium. Lack of awareness of this entity can result in a misdiagnosis and unnecessary procedures. The case here reported deals with an inverted left atrial appendage occurring in an infant after repair of an atrial septal defect.

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Introduction

Inversion of the left atrial appendage is a rare complication following open-heart surgery and only 16 cases have been reported so far in the literature[1-13]. The echocardiographic features of this condition mimic a left atrial mass and, therefore, it may be misdiagnosed as a tumor, a thrombus or a vegetation[5,10,11,13,14].

Failure to recognize this condition may lead to unnecessary and even dangerous consequences such as biopsies with the risk of systemic embolism, further invasive diagnostic procedures and even a resternotomy[4,6].

The natural history, complications and optimal therapeutic management of an inverted appendage remain to be fully elucidated. For this reason, intraoperative recognition of such a condition and, therefore, its immediate treatment are desirable.

This case report concerns a child submitted to surgical repair of a fenestrated defect of the atrial septum and a patent ductus arteriosus.

Case report

An 18-month-old male child with cardiomegaly was admitted to our unit for repair of an atrial septal defect and a patent ductus arteriosus. On physical examination the patient, weighing 13.5 kg, presented with a significantly reduced effort tolerance, but no shortness of breath at rest. On cardiovascular examination, a soft systolic murmur was audible along the left lower sternum. S1 was normal, S2 was split. The oxygen saturation in room air was 99% at rest. An initial chest X-ray revealed cardiomegaly with a cardiothoracic ratio of 0.64 for right ventricular enlargement, a well represented pulmonary trunk arch, and an increased pulmonary vascularity.

Echocardiography revealed a large fenestrated defect in the atrial septum, the posterior rim of the interatrial septum measuring < 5 mm. The atrial septal defect consisted of multiple fenestrations of the septum with a sizable color Doppler shunt through the holes. A moderate-to-severe overload of the right ventricular chamber, on the short-axis view, determined a change in the ventricular geometry that became globular in shape with a significant shift of the ventricular septum leftward and towards the center of the left ventricle at end-diastole. Tricuspid incompetence was detected and graded as mild (a right atrioventricular peak gradient of 32 mmHg). Moreover, a patent ductus arteriosus was detected and, on color Doppler analysis, the left-to-right shunt across the duct was considered to be significant. There were no masses in the left atrium.

Sternotomy was carried out and the ductus arteriosus ligated before commencing hypothermic cardiopulmonary bypass. The left heart was vented by a catheter in-

serted through the right superior pulmonary vein. After aortic cross-clamp and injection of cold blood cardioplegia, an oblique right atriotomy was performed and the defect exposed (Fig. 1). The remnants of limbic tissue were removed and the fenestrated defect of the atrial septum became a single large defect that was closed with a Dacron-Sauvage patch (1.8 × 1.3 cm).

Since the hemodynamic conditions were stable and the prognosis of the cardiac lesion repair was good, intraoperative echocardiography was deemed unnecessary and the patient was referred to the intensive care unit where he was weaned from mechanical ventilation 4 hours later.

On the third postoperative day, routine echocardiography was performed within the ward; this demonstrated a good surgical result with no residual shunt through the duct or interatrial septum. However, it also revealed an inhomogeneous mass in the left atrium, suggesting a cardiac tumor. On the parasternal long-axis view, the mass (Fig. 2), visualized in cross-section, appeared in the middle of the left atrial cavity close to the interatrial patch. Both in the systolic and diastolic phases, the mass appeared ovoid in shape with indented edges and a finger-like appearance when it was cut longitudinally. A diagnosis of tumor was excluded since no intracardiac mass had been detected at preoperative echocardiography. A thrombus in the left atrial cavity could not be ruled out and therefore anticoagulant therapy with heparin was started. No infectious disease or coagulation disorders were revealed upon clinical and laboratory examinations.

In view of the need for cardiac re-exploration, the mass was monitored at serial echography with no change being revealed in structure, size and location. The entire heart was carefully inspected to exclude the presence of multiple masses. On the short-axis view, repeated examinations failed to reveal the left atrial appendage: this anomalous finding led us to suspect intussusception of the left atrial auricle. The patient was therefore scheduled for transesophageal echocardiography. On the short-axis view, the mass was visible adjacent to the plane of the mitral annulus and below the pulmonary vein. The longitudinal view also showed the mass to be attached to the lateral atrial wall (Fig. 3). The entire left appendage was not visible in any view. The suspected diagnosis of inversion of the left atrial appendage was confirmed and therefore surgical exploration was cancelled in favor of a conservative approach.

The potential arrhythmogenicity of this mass in the left atrium, was studied by means of continuous 24-hour serial ECG recordings which demonstrated a normal sinus rhythm for 97% of the time and a wandering pacemaker for the remaining 3%. Moreover, a mean 24-hour heart rate of approximately 110 b/min with sporadic supraventricular beats but no ventricular arrhythmias was recorded.

Since hemodynamic conditions were stable and no unfavorable prognostic events emerged, further surgical procedures were deemed unnecessary.

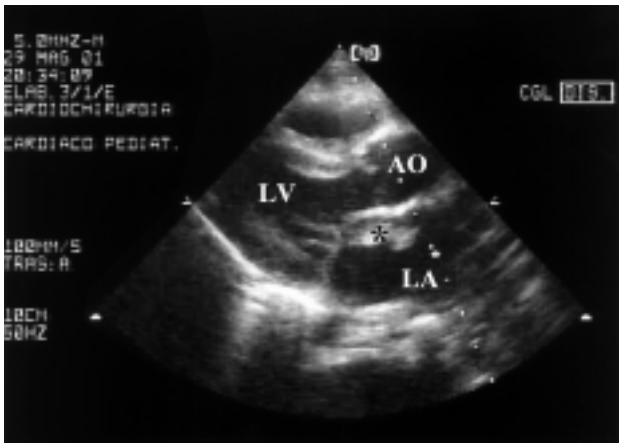

Figure 2. *Transthoracic parasternal long-axis view showing the mass (*) in the left atrial cavity close to the interatrial septum. AO = aorta; LA = left atrium; LV = left ventricle.*

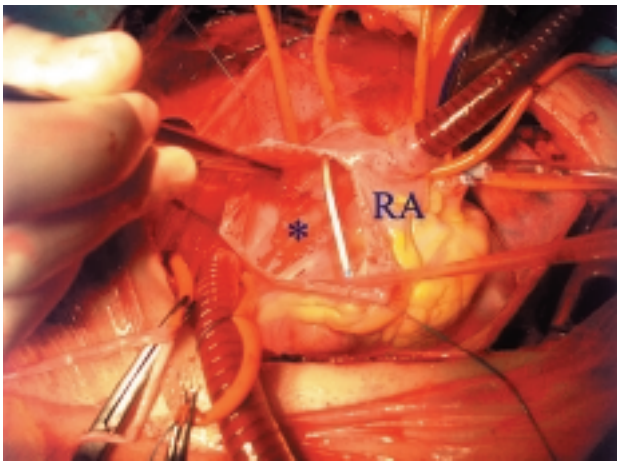

Figure 1. *Intraoperative view of the posterior atrial septal defect through the right atriotomy (*). RA = right atrium.*

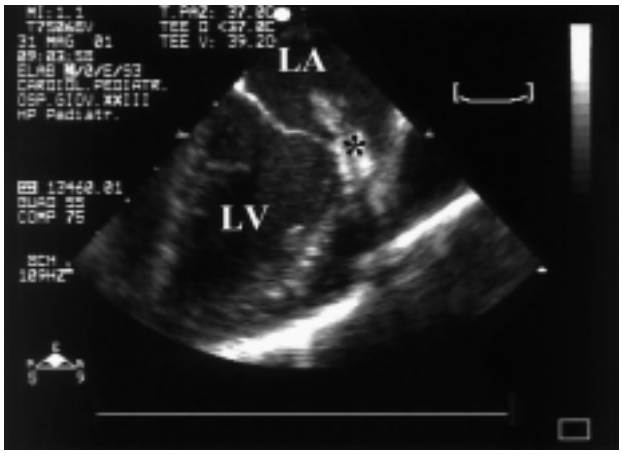

Figure 3. *Transesophageal long-axis view showing the mass (*) in the left atrium (LA) attached to the lateral wall. LV = left ventricle.*

The child was discharged on oral anticoagulant treatment for a period of 6 weeks and antiplatelet therapy for 6 months. At 14 months of follow-up, he was found to be in good clinical conditions with no symptoms of cardiac imbalance and/or clinical systemic embolism.

Discussion

Inversion of the left atrial appendage is a potential complication, after cardiac surgery, both for acquired and congenital heart diseases. Therefore, when a mass in the left atrium is discovered at postoperative echocardiography, an inverted left atrial appendage should be included in the differential diagnosis, together with a thrombus, vegetation and intracardiac tumor.

The most common cause of a left atrial mass is reported to be left atrial thrombi. The typical identifying features of thrombi are a laminated appearance with irregular or lobulated borders, often associated with spontaneous microcavitations at echocardiography. Thrombi, moreover, do not have a narrow stalk and are not of atrioseptal origin.

Primary tumors may vary considerably, according to their histotype and the patient's age at the time of diagnosis[15]. The most frequent tumors in pediatric age (< 15 years of age) are, in order of frequency: rhabdomyomas, fibromas, myxomas, and teratomas. Rhabdomyomas are derived from cardiac muscle (hamartoma). In approximately one third of cases they are associated with tuberous sclerosis. Such tumors may be localized throughout the heart (left ventricle, right ventricle, right atrium, left atrium, epicardium, and endocardium) but never originate from the cardiac valves. Besides, they tend to regress spontaneously as the patient approaches adulthood. Fibromas are composed of connective tissue derived from fibroblasts. The majority of tumors encroach on, or invade, the conduction system. Sometimes they occur in the atrium and usually present as an incidental finding on a routine chest X-ray. Myxomas arise from the endocardium as a polypoid, often pedunculated mass extending into a cardiac chamber. The most frequent location is in the left atrium and, from a clinical point of view, they are difficult to distinguish from mitral valve disease. A murmur of varying intensity together with systemic findings, such as an increased erythrosedimentation rate, may be useful for diagnostic purposes. Embolic phenomena represent the second most common situation.

In agreement with these findings and considering that our patient had been submitted to transthoracic echocardiography prior to surgery, we felt that it was reasonable to presume that the intracardiac mass within the left atrium was a *de novo* formation and therefore we excluded the presence of a tumor from the differential diagnosis.

The lack, upon clinical and laboratory examinations, of a postoperative infectious disease allowed us to definitely exclude the suspect of endocardial vegetations from the differential diagnosis.

In this case, even though the echocardiographic appearances of the mass were not in keeping with typical thrombi features, the presence of a thrombus could not be excluded. After cardiopulmonary bypass, the development of these types of masses in patients with congenital or acquired prothrombotic disorders who have undergone open-heart surgery, may be exacerbated by the iatrogenic coagulation imbalance associated with the use of non-biological materials such as the tubing of roller pumps and artificial oxygenators. In our patient, neither acquired nor congenital thrombophilic disorders had been detected prior to, or at the more detailed evaluation performed after detection of the intracardiac mass. However, as already pointed out, since a clot in the left atrial cavity could not be excluded, anticoagulant therapy with heparin was started, which, as would be expected, did not lead to any change in the structure and size of the mass.

Transesophageal echocardiography is the diagnostic gold standard for the evaluation of the left appendage and of its anomalies, in view of the high resolution of the images and of the possibility of performing multiplane scans[16]. In most of the cases of an inverted left atrial appendage reported in the literature[1-7,9,12], the diagnosis has been made or suspected by means of this technique performed both intra- and postoperatively. Magnetic resonance imaging has been used only in 3 cases and always after echocardiographic evaluation[8,10,11,13]. On account of its invasiveness with respect to conventional transthoracic echocardiographic imaging, transesophageal echocardiography is to be resorted to only in dubious cases. In the case reported here, atypical findings of a thrombus or of a tumor in conjunction with the absence of the left atrial appendage during transesophageal echocardiographic evaluation led us to diagnose the inversion of the left atrial appendage without the need of further diagnostic procedures.

It is tempting to hypothesize that anatomical and/or procedural factors may be responsible for the invagination of a left atrial appendage. Aronson et al.[1] were the first authors to propose that an unusually narrow neck of the left atrium appendage was implicated. Meanwhile, Ankersmit et al.[11,13] reported the only case of spontaneous intussusception of the left atrial appendage as a possible cause of an ischemic cerebral attack. No predisposing factor was speculated as the main cause of invagination of the auricle in this recent report. Later, the operative reports[1,4,9,10,12] of all cases in which the inverted appendage was surgically repositioned, confirmed that an unusually long and narrow base of the appendage might be the main cause of invagination and, despite adequate atrial preloading, of the lack of spontaneous repositioning.

As far as the procedural causes of the inversion are concerned, this is likely to occur either during the deairing maneuvers or due to suction of the vent. As we do not routinely perform manipulation of the left appendage, it is, in our opinion, feasible to exclude this as the cause of appendage inversion in the reported case (obviously attributing the intussusception of the appendage to a strong vent suction). Following this case of invagination of the appendage, in our Department, our perfusionists have started to take the precaution of inserting a depression modulator on the vent line in order to avoid the possibility that venting heart sections might turn the left auricle inside-out and lead to permanent invagination. Moreover, following the detection of this peculiarity, intraoperative echocardiography screening has been protocolled even in those cases with a simple diagnosis in which the corrective procedure was uncomplicated. Despite the drawbacks this modification in the protocol might have led to in our Surgical Unit, this problem has been easily overcome by resorting to epicardial echocardiography which is less time consuming, avoids esophageal intubation and necessitates limited human and technological resources[17], allowing a rapid and efficacious intraoperative monitoring in all cases submitted to cardiopulmonary bypass.

The limited number of cases of postoperative appendage inversion reported in the literature does not allow an estimate of the reduced risk of reoperation versus conservative treatment to be made. Besides, even though postoperative appendage inversion may potentially disturb the mitral valve[12], promote thrombus formation[4,8,11,13] and cause impaired ventricular preload[1,9] and arrhythmias[3,8], no such a complication has been demonstrated in cases of postoperative appendage inversion. Others consider this anomaly as a benign complication which is not dangerous for the cardiovascular system and may even resolve spontaneously[7,10]. Therefore, further surgery may not be required. Hence, in our opinion the fact that we prescribed oral anticoagulation treatment for several weeks and indeed for the whole period of transient postoperative myocardial dysfunction secondary to cardiopulmonary bypass and then antiplatelet therapy for 6 months should not give rise to controversy.

In agreement with Allen et al.[7], we believe that the inner surface of the inverted appendage in contact with blood, which is covered by an intact endothelium and therefore does not constitute a procoagulation factor, does not favor *per se* a thromboembolic status[18]. We think that a conservative approach is preferable if serial echocardiographic monitoring shows the same images of the appendage at outpatient controls, instead of scheduling the patient to invasive surgery.

To conclude, in the presence of a newly formed left atrial mass detected at echocardiography, especially during the early postoperative period, we strongly recommend that the possibility of an inversion of the left atrial auricle be included in the differential diagnosis.

Inversion of the left atrial appendage may be avoided with appropriate control of the force of the suction vent which, in a small heart, may be life-saving so long as suction of the vital structures is avoided.

Direct inspection of the left atrial appendage after cardiopulmonary bypass performed for any cardiac procedure and before closure of the sternotomy is always recommended.

The rarity of this complication does not allow an estimate of the potential hemodynamic consequences of an inverted atrial appendage to be made. Hence, choosing the appropriate therapeutic approach may be rather problematic. For this condition, intraoperative diagnostic evaluation for a prompt and safe repositioning of the appendage is highly recommended.

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References

1. Aronson S, Ruo W, Sand M. Inverted left atrial appendage appearing as a left atrial mass with transesophageal echocardiography during cardiac surgery. Anesthesiology 1992; 76: 1054-5.
2. Robertson DA, Arcilla RA, Sachsteder W, Ilbawi MN. Transesophageal echocardiography diagnosis of intussusception of the left atrial appendage. Echocardiography 1993; 10: 619-22.
3. Danford DA, Cheatham JP, Van Gundy JC, Mohiuddin SM, Fleming WH. Inversion of the left atrial appendage: clinical and echocardiographic correlates. Am Heart J 1994; 127: 719-21.
4. Slavik Z, Salmon AP, Lamb RK. Unusual left atrial mass following cardiac surgery in an infant. Eur J Cardiothorac Surg 1994; 8: 566-7.
5. Toma DM, Stewart RB, Miyake-Hull CY, Otto CM. Inverted left atrial appendage mimicking a left atrial mass during mitral valve repair. J Am Soc Echocardiogr 1995; 8: 557-9.
6. Minich LL, Hawkins JA, Tani LY, Judd VE, McGough EC. Inverted left atrial appendage presenting as an unusual left atrial mass. J Am Soc Echocardiogr 1995; 8: 328-30.
7. Allen BS, Ilbawi M, Hartz RS, Kumar S, Thoele D. Inverted left atrial appendage: an unrecognized cause of left atrial mass. J Thorac Cardiovasc Surg 1997; 114: 278-80.
8. Corno AF. Inverted left atrial appendage. J Thorac Cardiovasc Surg 1998; 114: 1223-4.
9. Cohen AJ, Tamir A, Yanai O, Houri S, Schachner A. Inverted left atrial appendage presenting as left atrial mass after cardiac surgery. Ann Thorac Surg 1999; 67: 1489-91.
10. Fujiwara K, Naito Y, Noguchi Y, Hayashi H, Uemura S. Inverted left atrial appendage: an unusual complication in cardiac surgery. Ann Thorac Surg 1999; 67: 1492-4.
11. Ankersmit HJ, Kocher A, Frank H, Mohl W, Wolner E. Inverted left atrial appendage masquerading as myxoma. Circulation 2000; 101: E42-E43.
12. Kanemitsu N, Okabe M, Wariishi S, Nakamura T, Kitamura F. Inverted left atrial appendage. Jpn J Thorac Cardiovasc Surg 2000; 48: 597-8.

13. Ankersmit HJ, Mohl W, Moser B, et al. De novo inverted left atrial appendage: an unrecognized cause of left atrial mass with symptoms mimicking myxoma. J Thorac Cardiovasc Surg 2001; 121: 1211-3.
14. Tallano JV, Gardin JM. Vegetation, tumor, masses and thrombi. In: Tallano JV, Gardin JM, eds. Textbook of two-dimensional echocardiography. New York, NY: Grune and Stratton, 1993: 239-70.
15. McAllister HA Jr, Fenoglio JJ Jr. Tumors of the cardiovascular system. In: Atlas of tumor pathology. Second series, Fascicle 15. Washington, DC: Armed Forces Institute of Pathology, 1978.
16. Nanda NC, Pinhero L, Sanyal RS, Storey O. Transesophageal biplane echocardiography imaging: technique, planes and clinical usefulness. Echocardiography 1990; 7: 771-8.
17. Ungerleider RM, Greeley WJ, Sheikh KH, et al. Routine use of intraoperative epicardial echocardiography and Doppler color flow imaging to guide and evaluate repair of congenital heart lesions. A prospective study. J Thorac Cardiovasc Surg 1990; 100: 297-309.
18. Scarabeo V, Cernetti R, Zorzi C, et al. Interatrial septal aneurysm and genetic prothrombotic disorders: possible interaction in the pathogenesis of pediatric stroke. Ital Heart J 2001; 2: 854-7.