GIANT LEFT ATRIAL APPENDAGE ANEURYSM AND RIGHT VENTRICULAR DIVERTICULUM: A VERY RARE ASSOCIATION

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Abstract

Left atrial appendage aneurysm (LAAA) is a rare cardiac anomaly, the association with right ventricular diverticulum (RVD) is extremely rare. LAAA is characterized by aneurysmal dilatation of left atrial may be congenital or acquired; RVD is an outpouching from the right cardiac chamber. Often fortuitous discovery between fetal period and adult age, these two entities are usually asymptomatic but can be complicated by arrhythmia and thrombo-embolism. We report the case of a 22-years-old which we have diagnosed LAAA associated with a RVD. The revelation mode was a pericardial effusion and the mechanism of pericardial effusion would be a pericardial dysgenesis or an inflammatory reaction. The diagnosis of these two anomalies is essentially posed by imagery test especially cardiac echography, cardiac RMI and scan. Treatment consists of resection of LAAA and RVD with pericardiocentesis.

Introduction:

Giant Left atrial appedange aneurysm is a rare cardiac malformation with less than 100 cases reported in the literature worldwide [1, 2], its association with a right ventricular diverticulum is still very rare. Giant left atrium is characterized by an aneurysmal dilatation of the left atrium, which may be congenital or acquired (secondary to mitral valve disease), whereas the diverticulum is defined as an evagination of one of the heart structures. These two clinical entities are associated with a high risk of rhythm disorders and thromboembolic complications, which make these pathologies so serious. Diagnosis is based on various imaging techniques, and management must be immediate after the diagnosis has been made.

Case Report

We report the case of a 22-years-old with no particular cardiovascular risk factors and no notable history of diseases. He consulted for dyspnea classified as NYHA II evolving for one month, with paroxysmal palpitations and no chest pain, no fever. The physical examination noted a patient in good general condition, normal BP (120/80 mmHg), high respiratory rate (34 cycles/min), spontaneous jugular vein distention and hepato-jugular reflux. The electrocardiogram was in regular sinus rhythm with electrical biauricular hypertrophy.

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Fig 1: Frontal chest X-ray showing moderate cardiomegaly with right overhang and left middle arch dilatation.

Fig 2: Transthoracic echocardiography showing a thickened pericardium with posterior localized pericardial effusion measuring 40 mm without significant respiratory variations (A); giant intrapericardial dog-eared left atrium measuring (20.6 mm x 73.3 mm) extending to the tip of left ventricle (B) associated with a diverticulum of the tip of right ventricle (C) and a small atrial septal defect.
Fig3: Posterior view of Cardiac CT scan showing a giant LAAA and a diverticulum of the tip of right ventricle (RVD).

The cardiac CT scan confirmed the diagnosis and did not find any coronary artery abnormalities. Moreover, the etiological assessment of the pericardial effusion was without particularity. We therefore retained the diagnosis of LAAA and right ventricular diverticulum, revealed by a pericardial effusion. In the absence of treatment, there is a risk of potentially fatal complications. The patient was referred to cardiovascular surgery to complete surgical cure.

Discussion:
The congenital giant left auricle is a very rare anomaly with less than 100 cases reported in the literature worldwide [1, 2], it results from a dysplasia of the pectineal muscle. It can be isolated or associated with other congenital malformations including the diverticulum but this association is rarely described. The intracardiac diverticulum is a saccular evagination of the ventricular wall, the embryological explanation of which is still poorly elucidated. It is most often found in the left ventricle [3], unlike in our case where it is found at the tip of right ventricle. In Africa, very few cases have been reported, in Morocco two pediatric cases have been described [4, 5]. These anomalies can occur at any age from the fetal period [6], young age [2, 5, 7, 8] to adulthood most often between the second and fourth decade [9, 10]. These two entities are most often asymptomatic, discovered incidentally, or sometimes symptomatic, revealing themselves by dyspnea, palpitations and above all serious complications such as rhythm disorders, thromboembolic complications and sudden death with vital prognosis involved [9-11], which is why it is necessary that surgical management follows the diagnosis [4]. The fact that revelation mode is a pericardial effusion in this case, is not rare as it has been described previously [4, 5, 12]. In literature, the presence of pericardial effusion was explained either by association with pericardial dysgenesis or by an inflammatory reaction. In this case, we did not find a pericardial abnormality and left atrium was intrapericardial with a dog-eared appearance. The diagnosis is made by cardiac ultrasound, cardiac CT or cardiac MRI [4, 10]. Surgical management consists of resection of the LAAA and diverticulum with pericardiocentesis [2, 6, 13]. The benefit of surgery is remarkable as reported in literature.

Conclusion:
Giant left atrium and right ventricular diverticulum are two rare cardiac anomalies requiring urgent surgical management due to the often fatal complications.

Informed consents of the patient
Oral informed consent has been obtained from the patient.
Conflicts of interest
The authors declare no conflicts of interest.

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