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Case Report

Complete Atrio-Ventricular Block with Atrio-Ventricular and Ventriculo-Arterial Discordance in Adults: About a Case

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ABSTRACT

Introduction: The double atrioventricular and ventriculoarterial discordance is characterized by an aberrant connection between a right atrium and a ventricle of left morphology, from which anterior ventricle emerges a dividing vessel: the pulmonary artery. From the left ventricle with right morphology emerges a large vessel not dividing, the aorta. It is a rare congenital heart disease that can be associated with disorders of atrioventricular conduction.

Case: It was a 45-year-old patient with no cardiovascular risk factors and no pathological history. He had a dyspnea of progressive worsening for two months initially at the usual efforts to then become a respiratory gene at the least effort. He did not report any notion of chest pain, dizziness or loss of consciousness. At admission, blood pressure was 150/60 mmHg with bradycardia at 40 beats/min, polypnea at 26 cycles/min and oxygen saturation at room air at 96%. The physical examination noted a right ventricular paraesternal heave and fine crackles at the pulmonary bases.

The EKG enrolled a complete atrioventricular block with narrow QRS complex and right atrial hypertrophy. Transthoracic ultrasonography showed atrioventricular and ventriculoarterial discordance with significant dilation of the left atrium and preserved biventricular systolic function.

A double chamber pacemaker implantation was performed. The ventricular lead was placed in the left ventricle instead of the right ventricle. We associated Spirinolactone and Ramipril in the treatment. The evolution was favorable with a considerable regression of the dyspnea two weeks after the pacemaker implantation.

Conclusion: Double atrioventricular and ventriculoarterial discordance is a rare congenital anomaly. It can induce disorders of atrioventricular conduction and compromise the functional and vital prognosis. Hence the need for implantation of a pacemaker.

Keywords: Pulmonary artery; Congenital heart disease; Bradycardia; Echocardiography; Myocardial perfusion

INTRODUCTION

The double discordance atrio-ventricular and ventriculo-arterial (DD) is characterized by an aberrant connection between a right atrium (OD) with a ventricle of left morphology, therefore not trabeculated, of this anterior ventricle comes out a vessel dividing: the pulmonary artery. From the left posterior ventricle of right morphology (trabeculated) emerges a large vessel not dividing: the aorta [1]. It is a rare congenital heart disease that can be associated with atrioventricular conduction disorders [2]. The objective of this case is to describe the clinical and therapeutic features of the complete atrioventricular block during the double discordance.

CASE PRESENTATION

This is a 45-year-old patient with no cardiovascular risk factors and no medical history. He had a progressive worsening dyspnea for two months, first at the usual efforts and then become a dyspnea at least efforts. He did not report any notion of chest pain, dizziness or loss of consciousness.

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At admission in our unit, the general condition was good. The blood pressure was 150/60 mm Hg. There was bradycardia at 40 beats per minute, a superficial polypnea at 26 cycles per minute, and oxygen saturation in ambient air at 96%.

The physical examination found a right ventricular paraesternal heave and fine crackling rattles at the pulmonary bases. The EKG recorded an atrioventricular dissociation with narrow QRS complex, right atrial hypertrophy (Figure 1).

The echocardiography showed atrioventricular and ventriculo-arterial discordance with significant left atrium dilatation with a volume of 30 m^1/m^2, biventricular systolic function preserved (Figure 2).

In this context of complete auriculo-ventricular block symptomatic with adult congenital heart disease, the implantation of pacemaker double chamber was performed (Figures 3). The ventricular lead was placed in the left ventricle instead of the right ventricle after catheterization of the cephalic vein. We associated Spirinolactone and Ramipril in the treatment. The evolution was favorable with a considerable regression of the dyspnea two weeks after the pacemaker implantation.

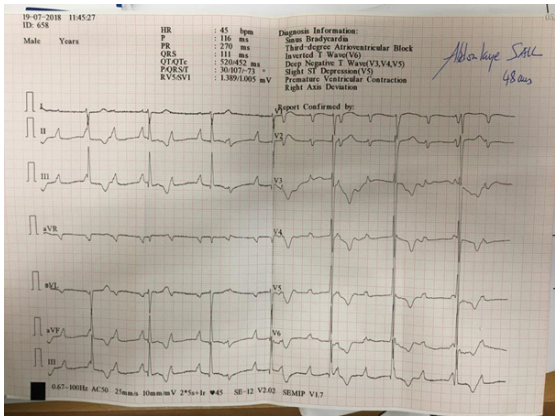

Figure 1: Surface ECG showing complete atrial ventricular block.

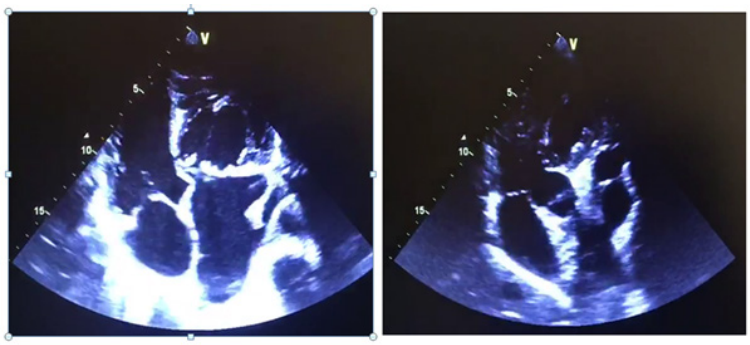

Figure 2: Transthoracic echocardiography apical 4 chambers (A) and 5 chambers (B) showing atrioventricular and ventriculo-arterial discordance.

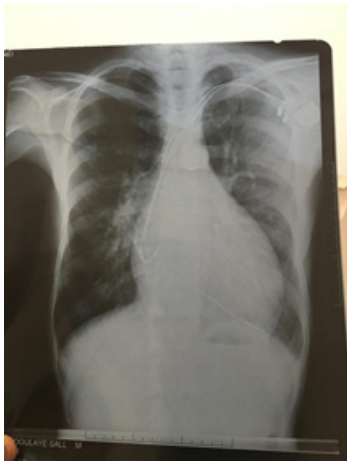

Figure 3: Thorax chest ray after pacemaker implantation.

DISCUSSION

Atrioventricular discordance is a rare congenital abnormality with an incidence of 1/33,000 live births, representing approximately 0.05% of congenital heart defects [3]. A pathophysiological contrary to the transposition of large vessels, the ventriculo-arterial discordance is corrected by the auriculo-ventricular discordance. Indeed, venous blood from the right atrium goes to the pulmonary artery via the left ventricle and oxygenated blood, via the left atrium, goes to the ventricle must then evacuated by the aorta [4].

It can cause conduction disorders. About one-tenth of infants born with dual discordance have complete atrial ventricular block [5,6]. In patients born with normal atrioventricular conduction, the risk of passing through complete atrial ventricular block over time is 2% per year and can reach 10-15% in adolescence or even 30% in adulthood [7]. The origin of atrioventricular conduction disorders is related to the malposition of the atrioventricular node on the axis of the conduction tissue. Gradually over time, the PR interval extends until the conduction blockage is complete and becomes symptomatic [8].

In our case, the major symptom was dyspnea. This respiratory gene could be explained in part by the poor tolerance of the conductive disorder also by the dysfunction of the systemic right ventricle. Indeed, several multicentre studies have shown an increasing incidence of heart failure with age [9,10]. After 45 years old, half of patients with associated lesions and one third of subjects without significant associated lesions will develop systemic right ventricle dysfunction. This dysfunction is also related to volume overload due to atrioventricular valve regurgitation or abnormal myocardial perfusion, during adolescence and adulthood.

Echocardiography has an important diagnostic role in the diagnosis of this condition, making it possible to determine the existence of associated malformations and to indicate the specific relationships between the different segments of the heart.

The therapeutic intake focuses on the management of the failing systemic right ventricle and the severity of associated malformations. It is a treatment with diuretics and inhibitors of the angiotensin converting enzyme. In case of atrioventricular conduction abnormalities such as the case of our patient, the implantation of a pacemaker is necessary especially when the latter is symptomatic [11]. The peculiarity of this stimulation lies in the fact that the ventricular probe is placed in the left ventricle instead of the right ventricle.

Our patient's short-term prognosis was good. But usually in the long run, volume overload of the systemic right ventricle will set in, leading to progressive heart failure as soon as hemodynamic stresses become significant.

CONCLUSION

The double atrioventricular and ventriculoarterial discordance is a rare congenital abnormality. It can induce atrioventricular conduction disorders and involve the functional and vital prognosis. Hence the need to implant a pacemaker.

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