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Case report

Cor triatriatum dexter: a rare incidentaloma

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Summary. Cor triatriatum dexter (CTD) is an extremely rare finding resulting from the persistence of the right valve of sinus venosus. It is a congenital cardiac anomaly defined by an abnormal septation of the atrium leading to inflow obstruction to the respective ventricle. Multimodal diagnostic modalities are necessary to characterize it for an optimal patient management. We report the case of a 68-year-old woman who presented to our clinic for further feedback of ventricular ectopic beats.

Key words: cor triatriatum dexter (CTD), atrial sept defect, cardiac magnetic resonance

Introduction

Cor triatratum dexter (CTD) is a partitioning of the right atrium (RA) to form a triatrial heart. It is an extremely rare congenital anomaly caused by the persistence of the right valve of the sinus venosus (1). The prevalence of CTD is only approximately 0.025% of cases of all congenital heart diseases (2). It has no gender predilection and commonly presents in infancy but may remain undetected till death (3). Symptomatology may mimic tricuspid stenosis, producing signs of systemic venous congestion (4). This paper presents a case of CTD in a 68-year-old woman without hemodynamic disturbances.

Case Report

A 68-year-old woman was referred to our clinic for further feedback of ventricular ectopic beats. Physical exam revealed irregular rhythm and no signs of heart failure. Transthoracic echocardiogram was performed and showed preserved left ventricular ejection fraction, dilated left atrium, and severely dilated RA. We didn't find any interatrial communication. Mild tricuspid and mitral regurgitation was reported. A diagnostic study was requested by means of cardiac magnetic resonance (CMR) to obtain in the first case a response to ventricular ectopic beats. To answer to the diagnostic suspicious of cardiologists about the ventricular ectopic beats, we performed a contrast CMR to eventually demonstrate the presence of ventricular fibrosis with the Late Gadolinium Enhancement (LGE) technique, which ruled out the presence of fibrosis. The sequences cine SSFP, most commonly used to study the cardiac morphology, demonstrated the presence of a membrane and an accessory chamber in greater detail, which showed the presence of CTD (**Figure 1**).

Atrial chambers were dilatated and the ventricles were of normal dimensions with a normal functionally parameters. In the absence of any heart hemodynamic distubances CTD can be considered a rare incidentaloma. The patient was discharged with beta blockers therapy and the strategy of watchful observation was adopted.

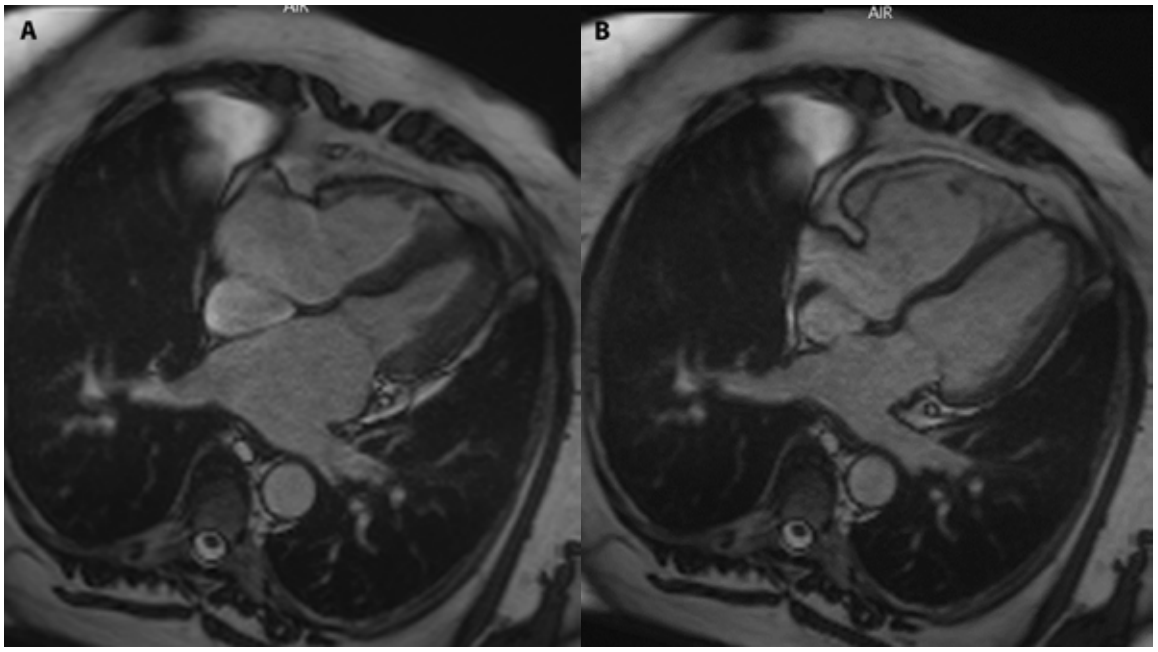

Figure 1. Four chamber visioncine SSFP. It is possible to view the presence of cor triatrium dexter during the atrial diastole and atrial systole.

Discussion: Cor triatriatum dexter is a rare congenital heart disease characterized by a separation between the smooth posterior part and the trabeculated anterior part of the RA due to the persistence of the right valve of the right horn of the sinus venosus. The right horn of the sinus venosus is incorporated into the right atrium, giving rise to the posterior smooth part of the right atrium called sinus venarum, the orifices of the superior vena cava and inferior vena cava. During development, the right valve of the sinus venosus divides the right atrium and then ends up leaving the crista terminalis, eustichian valve, and thebasian valve (5, 6). The membrane in CTD may range from a reticulum to a substantial sheet of tissue. The most common location of the membrane is at the right of superior vena cava, coronary sinus, and inferior vena cava, and the next common location is at the left of coronary sinus and to the right of superior and inferior vena cava (7). Right ventricular hypoplasia, pulmonary valve atresia, pulmonary valve stenosis, and Ebstein anomaly are communally associated with CTD (8,9). Multimodal imaging modalities with echocardiography, transesophageal echography and CMR are necessary to confirm this condition and may be required to delineate and differentiate it from others cardiac anomalies. Moreover, they also directly visualize the presence of the membrane that divides the atrial chamber into two separate chambers. CMR could play an important role where the transthoracic echocardiography can't show the real cardiac morphology. The degree of septation within the right atrium decides the age and severity of clinical manifestations, thus it may mimic tricuspid stenosis with signs of systemic venous congestion or when the septation is mild, the condition is often asymptomatic and represents an incidental finding frequently made at postmortem examination. In symptomatic patients the mainstay of treatment for CTD is surgical resection or percutaneous catheter disruption of the dividing membrane (10,11), conversely treatment is not required when the septum appears to have no hemodynamic significance.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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