

Posterior Nutcracker Phenomenon in a Patient with Left Atrial Isomerism

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Abstract

Keywords

- ▶ congenital heart disease
- ▶ magnetic resonance imaging
- ▶ pediatric
- ▶ cardiovascular surgery

Background Nutcracker phenomenon (NCP) can occur due to various anatomical anomalies. Anterior and posterior NCP are defined in the literature. Posterior NCP combined with left atrial isomerism is a rare condition.

Case description We present a rare case of an asymptomatic posterior NCP involving the azygos vein in a patient with a complex cardiovascular pathology with left atrial isomerism, left ventricular outflow tract obstruction, interrupted inferior vena cava, and azygos continuation.

Conclusion Detection of the NCP especially involving such a rare anatomical anomaly as an azygos continuation has a crucial importance for diagnostic and surgical procedures.

Introduction

Left atrial isomerism is an uncommon condition associated with complex cardiovascular abnormalities requiring surgical correction and results in significant morbidity and mortality. Its combination with a compression of the azygos vein between aorta and vertebral column is even rarer. Because of this, knowledge of this anomaly and its anatomical variations play an essential role in cardiac surgery. Here, we present a unique case of interrupted inferior vena cava (IVC) with azygos continuation as a part of left atrial isomerism combined with posterior nutcracker phenomenon caused by compression of the azygos vein diagnosed by noncontrast-enhanced cardiac magnetic resonance imaging (MRI).

Case Description

A 24-year-old woman presented with a severe aortic regurgitation and moderate pulmonary stenosis for possible aortic and pulmonary valve surgery. She is status post Ross-Konno procedure for 14 years, performed to correct multilevel left ventricle outflow tract obstruction (previously two subaortic resections performed 19 and 22 years earlier).

She has left atrial isomerism with infrahepatic interruption of the IVC with azygos continuation, connecting to the right superior vena cava (SVC), left SVC continuation to the coronary sinus, common atrium (surgical correction 24 years earlier), abdominal situs inversus with malrotation and duodenal stenosis (surgical correction for 24 years).

She presented with NYHA (New York Heart Association) clinical stage II–III. Spiroergometry demonstrated no obstructive or restrictive ventilation disturbances, no dys- or arrhythmias or signs of myocardial ischemia were detected on further examinations. Cardiac exercise capacity was moderately reduced due to limited patient compliance.

Echocardiography confirmed left ventricular (LV) dilatation, severe aortic insufficiency, and moderate pulmonary stenosis with a pressure gradient of 45 mm Hg; LV ejection fraction was 54%. Long-term electrocardiogram monitoring showed regular rhythm with increasing rate of ventricular extrasystole, which had also been noticed by the patient. Noncontrast-enhanced cardiac MRI revealed sufficient LV pump function with LV dilatation and hypertrophy. Aortic valve (pulmonary autograft) demonstrated severe regurgitation, and the pulmonary homograft showed moderate stenosis. The hepatic segment of the IVC was absent, and hepatic veins drained directly into the right atrium. A large azygos

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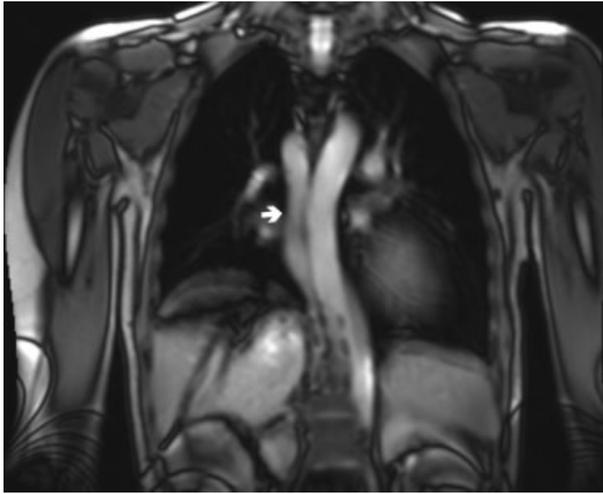


Fig. 1 Magnetic resonance imaging of the azygos vein (white arrow) passing between the aorta and spinal column and conflues with the right superior vena cava (coronal view).

vein crossed the vertebral column from left to right posterior to the thoracic aorta, where it is compressed between these two anatomical structures and then connects to the right SVC (→Figs. 1 and 2). No symptoms related to this nutcracker phenomenon were found in our patient.

Discussion

Left atrial isomerism is a subset of heterotaxy syndrome due to the disorder of left-right axis determination and may present with a variety of cardiac lesions and thoracoabdominal organ abnormalities, which determines a high mortality of up to 75% before 5 years of age.^{1,2} Left atrial isomerism is often associated with persistent left SVC, interruption of the hepatic part of the IVC with azygos/hemiazygos continuation, common atrium, LV outflow tract obstruction, and gastrointestinal abnormalities,²⁻⁴ which were also present in our patient. Interrupted IVC combined with azygos continuation is a rare condition with an incidence of 0.6 to 2.0% in patients with congenital

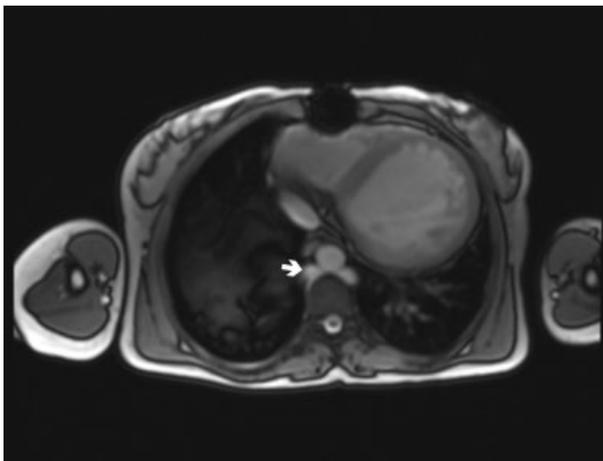


Fig. 2 Magnetic resonance imaging of the azygos vein (white arrow) compressed between the aorta and spinal column (horizontal view).

heart disease and less than 0.3% among healthy population.⁵ Abnormalities of the IVC have an asymptomatic course and are recognized incidentally during surgical procedures, radiological investigation or postmortem.⁶

Nutcracker phenomenon refers primarily to compression of the left renal vein (LRV) between aorta and superior mesenteric artery with impaired blood outflow to the IVC also known as anterior nutcracker in contrast to posterior nutcracker which occurs due to compression of the LRV between aorta and vertebral body.⁷

To the best of our knowledge, no case of posterior nutcracker phenomenon involving azygos vein compression in the context of left atrial isomerism has been described in the literature. Imaging of this condition on MRI presents a nonradiating diagnostic option in patients.

Although in the present case no significant symptoms of the azygos vein compression have been observed, this finding has a great clinical importance for planning cardiac surgery in terms of the application of the extracorporeal circulation and particularly adequate and safe venous drainage. Furthermore, symptoms may occur in the future, and follow-up is necessary.

Funding

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Conflict of Interest

All authors declare that they have no conflict of interest.

Ethical Approval

This article does not contain any studies with human participants performed by any of the authors.

Informed Consent

Informed consent was waived off because this study included no personal information about the patient.

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