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Case report
Prikaz slučaja
UDK 616.126.42-007-089.168:616.61-008.6
https://doi.org/10.2298/MPNS2402063K

RARE CASE OF UNILEAFLET MITRAL VALVE DIAGNOSED BY COMPUTED TOMOGRAPHY

REDAK SLUČAJ JEDNOLISNE MITRALNE VALVULE DIJAGNOSTIKOVAN KOMPJUTERIZOVANOM TOMOGRAFIJOM

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Summary

Introduction. Congenital malformations of the mitral valve encompass a broad spectrum of lesions, with an incidence of approximately 0.4% among patients with congenital heart diseases. **Case Report.** A 45-year-old female was admitted to our institution due to a confirmed mitral valve defect identified via echocardiographic examination. She presented with leg swelling and rapid fatigue. Her medical history includes systemic lupus erythematosus. Upon admission, she was in cardiopulmonary decompensation, with a systolic murmur detected in the mitral valve area, radiating towards the axilla. Transesophageal echocardiographic revealed severe mitral stenosis with moderate regurgitation, without clear visualization of the two cusps. Cardiac computed tomography confirmed a voluminous anterior mitral cusp and a hypoplastic posterior cusp, observed in diastole as a crescent-shaped, characteristic of a unicuspid mitral valve. Follow-up echocardiographic showed progression of mitral regurgitation with circumferential pericardial effusion, necessitating pericardiocentesis. Due to further hemodynamic instability, urgent surgical mitral valve replacement was indicated. The surgery proceeded without complications, but the immediate period was marked by exacerbation of renal impairment, requiring hemodiafiltration on several occasions. Following cardiological stabilization, the patient was transferred to the Nephrology Clinic for further management of renal insufficiency and systemic lupus. **Conclusion.** It is crucial to emphasize the significance of a multidisciplinary approach for each patient, with particular emphasis on detailed imaging diagnostics to definitively establish a congenital heart disease diagnosis.

Key words: Mitral Valve; Congenital Abnormalities; Tomography, X-Ray Computed; Echocardiography; Heart Valve Prosthesis Implantation; Signs and Symptoms; Hemodynamics; Diagnostic Imaging; Postoperative Complications; Renal Insufficiency

Sažetak

Uvod. Kongenitalne malformacije mitralne valvule predstavljaju širok spektar lezija, sa incidencijom od 0,4% među pacijentima sa kongenitalnom bolešću srca. **Prikaz slučaja.** Pacijentkinja starosti 45 godina je primljena u našu ustanovu zbog verifikovanog mitralnog defekta ehokardiografskim pregledom, što je bilo praćeno oticanjem nogu i brzim zamaranjem. Od komorbiditeta navodi sistemski lupus. Pri prijemu kardiopulmonalno dekompenzovana, sa auskultatorno verifikovanim sistolnim šumom u predelu mitralne valvule, koji propagira put aksile. Transezofagealnim ehokardiografskim pregledom verifikovana je teška mitralna stenoza sa umerenom regurgitacijom, bez jasne vizualizacije dva kuspisa. Kompjuterizovanim tomografijom srca verifikovano je postojanje voluminoznijeg anteriornog mitralnog kuspisa, dok je posteriorni kuspis hipoplastičan, što se u dijastoli videlo kao oblik polumeseca (karakteristika jednolisne mitralne valvule). Načinjen je kontrolni transtorakalni ehokardiografski pregled gde je evidentirana progresija mitralne regurgitacije, sa cirkularnim perikardnim izlivom, zbog čega je načinjena perikardiocenteza, ali usled dalje hemodinamičke nestabilnosti indikovana je urgentna hirurška zamena mitralne valvule. Kardiohirurška intervencija je protekla bez komplikacija, ali je neposredno postoperativno registrovana akutizacija bubrežne slabosti, zbog čega je načinjena hemodijafiltracija u nekoliko navrata. Nakon kardiološke stabilizacije, pacijentkinja je premeštena na Kliniku za nefrologiju radi daljeg tretmana bubrežne insuficijencije i sistemskog lupusa. **Zaključak.** Krucijalno je naglasiti značaj multidisciplinarnog pristupa kod svakog pacijenta, sa posebnim skretanjem pažnje na imidžing dijagnostiku kao definitivnu dijagnostičku metodu za uspostavljanje dijagnoze kongenitalne bolesti srca.

Ključne reči: mitralna valvula; kongenitalne abnormalnosti; CT; ehokardiografija; implantacija srčanog zaliska; znaci i simptomi; hemodinamika; dijagnostički imidžing; postoperativne komplikacije; bubrežna insuficijencija

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Abbreviations

LV – left ventricle
CHD – Congenital heart disease
MV – mitral valve
MR – mitral regurgitation
TTE – transthoracic echocardiography
TEE – transesophageal echocardiography
CT – computed tomography
CVVHDF – continuous veno-venous hemodiafiltration

Introduction

The mitral apparatus is a complex anatomical structure situated between the left atrium and left ventricle (LV). It comprises the annulus, and the anterior and posterior cusps, which are connected to the two papillary muscles of the LV via chordae tendineae, preventing valve prolapse during ventricular systole. Given the complexity of its anatomy and the importance of each segment, the integrity of the mitral apparatus is essential for adequate LV function.

Congenital heart disease (CHD) is the leading cause of mortality in infants, with anomalies of the heart and large blood vessels being the most common fetal abnormalities [1]. Congenital malformations of the mitral valve (MV) cover a wide spectrum of lesions, with an incidence of approximately 0.4% among patients with CHD, or 5 per 100,000 in the general population [2, 3]. These malformations can present in several forms, including parachute MV, double orifice MV, cleft MV, atresia, and unileaflet MV, with unileaflet MV being one of the rarest [4, 5]. Unileaflet MV typically diagnosed in early childhood and is generally incompatible with life due to the presence of severe mitral regurgitation (MR) [4]. In asymptomatic patients with unileaflet MV, severe hypoplasia of the posterior mitral cusp is often observed [6].

Patients with unileaflet MV experience symptoms due to severe MR, which manifests as rapid fatigue, dyspnea, inability to lie flat, palpitations, and pretibial edema. Auscultation over the MV reveals a pansystolic murmur that often radiates to the axilla. The diagnosis of unileaflet MV is usually made through clinical examination and transthoracic echocardiography (TTE), while transesophageal echocardiography (TEE) and cardiac computed tomography (CT) provide precise evaluation of the mitral apparatus.

Case Report

We present a case involving a 45-year-old female patient who was transferred to our institute from the Institute for Pulmonary Diseases of Vojvodina, where TTE revealed hypertrophic cardiomyopathy and combined mitral valve disease. The patient reported swelling of her legs over the past week, along with rapid fatigue upon mild physical exertion and difficulty breathing. Additionally, she had a persistent dry cough for the past month, for which she was prescribed systemic intravenous corticosteroid and antibiotic therapy. She had been treated for hypertension for several months. Other comorbidities included systemic lupus erythematosus accompanied by lupus rheumatoid arthritis, stage II renal insufficiency, hyperlipidemia, a history of smoking, and a family history of cardiac diseases, all risk factors for ischemic heart disease.

Upon admission to our institution, the patient was conscious, oriented, and communicative, with borderline hypertension (140/90 mmHg), a normal heart rate (80 bpm), and cardiopulmonary decompensation. Clinically, pretibial edema was observed, more pronounced in the left lower leg. Auscultation revealed bilaterally diminished breath sounds at the lung bases without other accompanying sounds, and a pronounced systolic murmur with radiation to the axilla was heard at the mitral area. Laboratory tests showed markedly elevated levels of NT-proBNP (21,663 pg/ml), increased levels of nitrogenous substances (urea 9.5 mmol/l, creatinine 135 μmol/l), and elevated inflammatory markers (C-reactive protein 53 mg/l, fibrinogen 10.7 g/l). Based on these findings, continuous intensive diuretic therapy was initiated along with empirical parenteral antibiotic therapy.

Transesophageal echocardiography revealed dilated LV with increased volume, severe mitral valve stenosis (MaxPg 56 mmHg/30 mmHg), and moderate MR. 4D reconstruction of the mitral valve did not clearly visualize the two cusps. Additionally, moderate tricuspid valve insufficiency with consequent pulmonary hypertension was noted. Cardiac CT was performed for a precise evaluation of the mitral valve, revealing a voluminous anterior mitral cusp while the posterior mitral cusp was not visualized, suggesting atresia. In diastole, the mitral orifice appeared crescent-shaped, consistent with a unileaflet mitral valve **(Figure 1).** Following all diagnostic procedures, a diagnosis of unileaflet mitral valve was established, warranting mitral valve replacement.

During the preoperative hospital course, the patient experienced intermittent febrile episodes with increased inflammatory markers, leading to the prescription of empirical parenteral antibiotic therapy. Due to significant orthopnea and tachypnea, non-invasive mechanical ventilation was applied several times, along with intensive parenteral diuretic and bronchodilator therapy. These symptoms subsequently improved, but due to right-sided pleural effusion, a pleural puncture was performed. Laboratory tests revealed an acute exacerbation of renal insufficiency, necessitating continuous venovenous hemodiafiltration (CVVHDF), which resulted in decreased levels of nitrogenous substances.

A follow-up TTE examination revealed the progression of MR along with circumferential pericardial effusion, with separation of the pericardial layers up to 2.2 cm, indicating impending tamponade. Consequently, pericardiocentesis was performed, evacuating 650 ml of hemorrhagic fluid. However, the patient's hemodynamic status deteriorated, necessitating an urgent transfer to the cardiac surgery operating room, where emergency mitral valve replacement with a mechanical bileaflet valve (St. Jude No. 23) was performed. The cardiac surgery proceeded without complications, with a total cardiac arrest time of 91 minutes and a cardiopulmonary bypass time of 103 minutes.

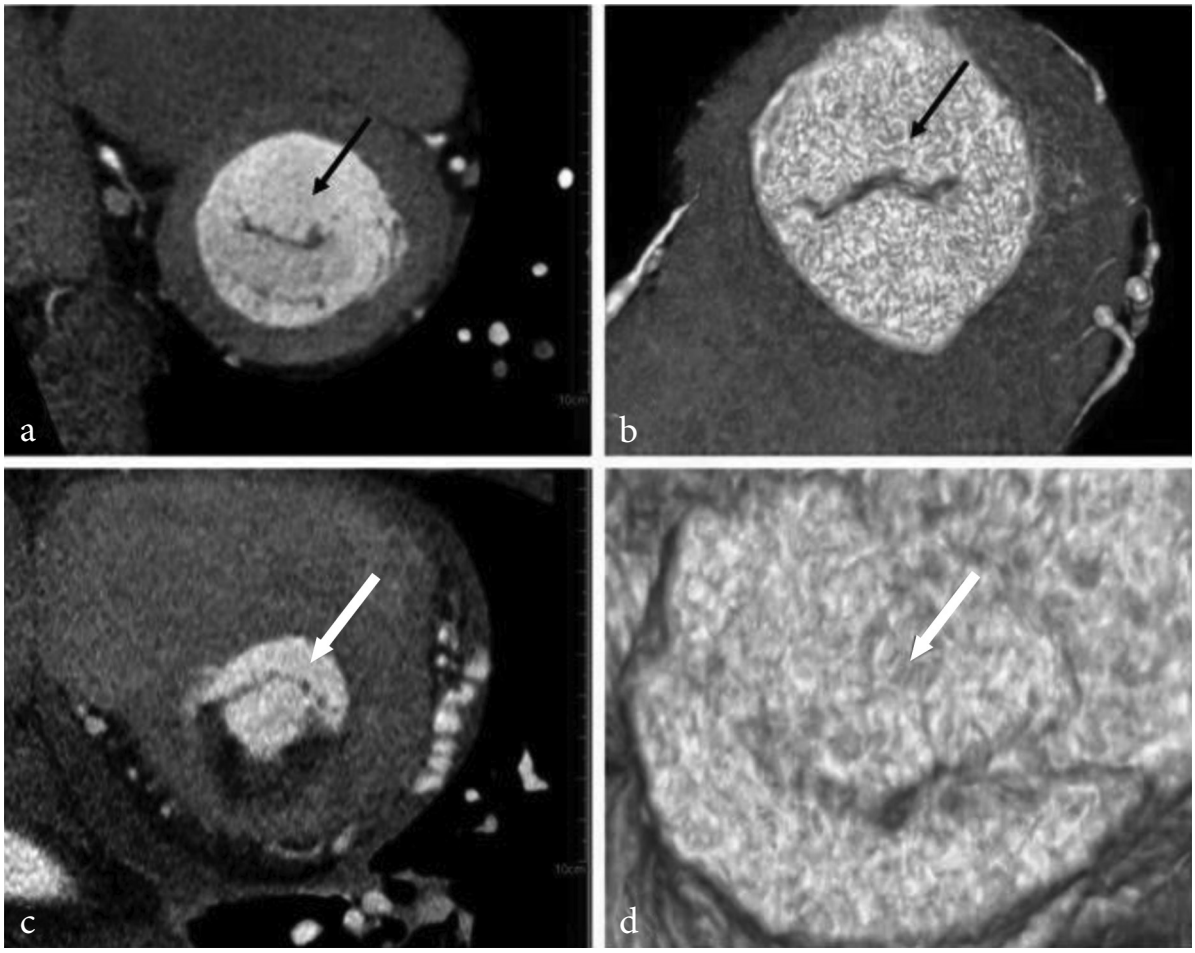

Figure 1. a) and b). Black arrow showing normal mitral valve – fish mouth shaped; **c) and d)** white arrow showing unileaflet mitral valve – crescent-shaped
Slika 1. a) i b) *Crna strelica je usmerena na mitralnu valvulu normalnog izgleda – oblika ribljih usta;* ***c) i d)*** *bela strelica pokazuje jednolisnu mitralnu valvulu naše pacijentkinje koja je oblika polumeseca*

In the immediate postoperative course, the patient was hemodynamically and rhythmically unstable, requiring high doses of vasopressor support. Arterial blood gas analysis revealed metabolic acidosis with elevated lactate levels. Based on hemodynamic parameters, inotropic support was initiated, gradually achieving hemodynamic stability. Due to significant pleural effusion detected on echocardiography and chest radiography, a left pleural drain was placed, yielding serous fluid.

After discontinuation of continuous sedation and stabilization of arterial blood gas exchange, weaning from mechanical ventilator support was initiated, and the patient was extubated on the third postoperative day. Febrile episodes prompted blood culture sampling, which returned negative results, while sputum culture was positive for Acinetobacter species, leading to antibiotic therapy according to the antibiogram.

Recurrent acute exacerbation of renal insufficiency with anuria and markedly elevated nitrogenous substance levels (urea 21.9 mmol/l, creatinine 317 μmol/l) necessitated postoperative CVVHDF on multiple occasions. Following complete stabilization of the cardiac condition, the patient was transferred to the University Clinical Center of Vojvodina, specifically to the Nephrology and Immunology Clinic, for further monitoring and treatment of renal insufficiency and systemic lupus erythematosus.

Conclusion

Although congenital heart diseases are most commonly detected in the neonatal period, some cases are identified later in life. Our case report illustrates such a scenario, where a congenital anomaly of the mitral valve was discovered in adulthood. Through a multidisciplinary and prompt approach, the patient was successfully managed from a cardiological perspective.

This case underscores the importance of a multidisciplinary approach for each patient, with particular emphasis on detailed imaging diagnostics to definitively establish a diagnosis of congenital heart disease.

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Rad je primljen 9. VI 2024.
Recenziran 4. VII 2024.
Prihvaćen za štampu
BIBLID.0025-8105:(2024):LXXVII:1-2:63-66.