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A CASE OF ORTHOTOPIC BICAVAL HEART TRANSPLANTATION IN A PATIENT WITH PERIPARTUM CARDIOMYOPATHY

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This paper presents a clinical case of orthotopic bicaval heart transplantation in a patient with end-stage chronic heart failure that developed against the background of peripartum cardiomyopathy. The disease manifested itself in the second trimester of pregnancy, complicated by severe preeclampsia and progressive heart failure. In the absence of an available donor organ, the patient was implanted with a left ventricular assist device HeartMate 3 as a «bridge to transplantation». Four years after left ventricular assist device implantation, when a donor appeared, orthotopic heart transplantation was successfully performed. The case demonstrates the effectiveness of a multidisciplinary approach in conditions of a limited donor resource and emphasizes the need for early diagnosis, systemic prevention and development of transplantation services to improve outcomes in patients with peripartum cardiomyopathy.

Key words: transplantation; peripartum cardiomyopathy; chronic heart failure; left ventricular assist device

INTRODUCTION

Chronic heart failure (CHF) is a serious problem in modern cardiology, being one of the leading causes of disability and death among patients with heart disease. One of the most effective treatments for late stages of CHF is orthotopic heart transplantation (OHT). This treatment method improves the quality of life and prognosis of patients with end-stage heart failure, but requires solving a number of complex medical, organizational and ethical problems.

In recent years, orthotopic heart transplantation has become increasingly important in our country. The shortage of donor organs, complex transplantation processes and the need to comply with strict medical and legal standards create additional difficulties for the development of the transplant program in the country.

Peripartum cardiomyopathy (PPCM) is a secondary cardiomyopathy, usually with a decrease in LVEF ≤45% and manifested towards the end of pregnancy (third trimester) or in the first months after childbirth without any other identifiable cause [1].

The pathogenesis of this cardiomyopathy is associated with increased oxidative stress, prolactin cleavage into the angiostatic 16 kDa N-terminal fragment of prolactin (16 kDa Prl), and impaired VEGF signaling due to upregulation of sFlt-1. The 16 kDa fragment of prolactin causes significant endothelial injury and myocardial dysfunction. Similarly, sFlt-1 further damages the vasculature, supporting the idea that angiogenic imbalance and endothelial injury may play a role in the development of PPCM [2].

A nationwide retrospective study of periportal cardiomyopathy in Japan in 2007 – 2008 reported that elevated sFlt-1 was associated with the development of preeclampsia and PPCM(3). According to this study, the incidence of PPCM in Japan was 1 case per 20,000 deliveries (102 cases in absolute numbers), and the mean age of patients was 32.7 years. The mean values of laboratory and instrumental studies were as follows: Left Ventricular Ejection Fraction (LVEF) – 31.6 ± 12.0%, the mean level of brain natriuretic peptide (BNP) in serum was 1,258 ± 1,028 pg/mL. Only 4 patients had BNP levels below 100 pg/mL. The mortality rate was 4%. Most notably, 2% of patients had severe LV dysfunction requiring mechanical support (Left Ventricular Assist Device (LVAD).

Although the causes of preeclampsia, defined as new hypertension (diastolic blood pressure >90 mmHg) and significant proteinuria (>300 mg per 24 hours) after 20 weeks of gestation, remain largely unknown, the leading hypothesis strongly suggests a disruption of placental function early in pregnancy, possibly due to a failed interaction between two genetically distinct organisms.

This paper considers not only the medical aspects of this case, but also the problem of donation, which forces the country to resort to expensive treatments to save the patient's life.

Aim – to reveal the problems of treating end-stage chronic heart failure by heart transplantation. Description of a clinical case of treating end-stage chronic heart failure by implantation of the LVAD Heart Mate 3 mechanical left ventricular assist device as a «bridge to therapy» and heart transplantation as the final primary surgical treatment.

Description of the clinical case:

This case report presents a clinical case of a patient who underwent orthotopic heart transplantation as a treatment for terminal stage chronic heart failure. The phenotype of this case of CHF is PPCM. According to statistics,

a total of 101 heart transplants have been performed in Kazakhstan, including 3 patients with peripartum cardiomyopathy.

Patient L., a woman, born in 1980, was first hospitalized at 20 weeks of her third pregnancy in a district hospital with a diagnosis of «Gestational arterial hypertension» and was discharged with improvement and recommendations. In February 2016, the patient was diagnosed with severe preeclampsia (blood pressure – 170/110 mm Hg, urine protein – 7.5 g/l) and from the same period, symptoms and signs of CHF appeared.

In the same year, he was hospitalized on a planned basis at the JSC «National Scientific Cardiac Surgery Center» (hereinafter JSC «NSCC»), during hospitalization, laboratory and instrumental studies were carried out. According to echocardiography data: end-diastolic volume (EDV) – 289 ml, end-systolic volume (ESV) – 241 ml, LVEF – 17%, Tissue Doppler Imaging, medial mitral annular systolic velocity (TDI S'med) – 3.8 cm/s, lateral mitral annular systolic velocity (S'lat) – 4.2 cm/s, severe mitral valve insufficiency (MR+++), right heart catheterization of pulmonary artery (PA) – 26/17-20 mm Hg, pulmonary vascular resistance (PVR) – 2.21 Wood Units, cardiac output (CO) (according to Fick) – 2.87 l/min, cardiac index (CI) (according to Fick) – 1.59 l/min/m^2, also coronary angiography was performed to exclude ischemic genesis of the disease: vessels are intact. Considering the positive dynamics against the background of drug therapy, a decrease in heart failure symptoms, an increase in tolerance to physical activity, it was recommended to continue optimal drug therapy for CHF, as well as disaggregant and gastroprotective therapy.

In March 2017, she had an outpatient consultation with a cardiologist at JSC «NNCC», during echocardiography: LVEF – 24%, EDV – 236 ml, right posterior descending artery – 40 mm Hg. Surgical treatment of CHF was recommended, which the patient refused. In 2018, the patient was hospitalized with decompensated CHF in JSC «NNCC», taking into account CHF functional class III according to New York Heart Association (Functional Classification, INTERMACS 3, heart condition (left ventricular end-diastolic volume – 224 ml, EF 22%), progression of heart failure, despite the background of maximum optimal drug therapy, the impossibility of transplantation of a donor heart (lack of a donor) at that time, as the «bridge to transplantation» therapy by the decision of the council, implantation of the left ventricular mechanical support device LVAD was recommended.

In the same year (June 29, 2018), the LVAD Heart Mate 3 left ventricular support device was implanted under artificial circulation. The early postoperative period was complicated by renal failure (creatinine – 2.29 mg/dl, urea – 51.1 mg/dl, SCF according to CKD-EPI: 43.3 ml/min/1.73 m^2, CKD stage 3B). Against the background of the therapy, renal function was restored. Otherwise, the postoperative period was uneventful. She was discharged with improvement and recommendations (optimal drug therapy for CHF, anticoagulant, disaggregant and gastroprotective therapy). Subsequently, the patient was observed in this center, was hospitalized several times on a planned basis for catheterization of the right heart.

Four years after LVAD implantation, the patient was admitted to the cardiology department with an infection at the LVAD cable exit site: Serratia marcescens 10x5 colony forming units were found in the cable discharge, etiologic antibacterial therapy was performed. Heart systolic function indices improved: CO (according to Fick) – 3.94 l/min, CI (according to Fick) – 2.18 l/min/m^2. The patient was put on the waiting list for donor heart transplantation.

In 2024 (17.07./2024), due to the presence of a donor heart, resternotomy was performed, LVAD Heart Mate 3 was explanted, orthotopic bicaval heart transplantation was performed under artificial circulation, hypothermia. Drive line was explanted. During hospitalization, a full range of therapeutic measures was carried out (standard cardiotonic, immunosuppressive, antibacterial, symptomatic, rehabilitation).

The patient's well-being improved dynamically. According to the protocol for managing patients with heart transplantation, an endomyocardial biopsy was performed: morphological picture of acute cellular rejection of the graft: pathologic Antibody-Mediated Rejection stage 2 (2013 International Society for Heart and Lung Transplantation, Antibody-Mediated Rejection, both histological and immunopathological data are present; deposition of immune complexes IgG, fibrinogen, kappa and lambda +, rejection of IgA and IgM +) 1R (2004) – low grade. Immunosuppressive therapy was enhanced. In August 2024 (07.08.2024) the patient was discharged with recommendations in a satisfactory condition.

At the moment, the patient is undergoing outpatient monitoring and rehabilitation measures.

Main disease: Peripartum cardiomyopathy. Implantation of the LVAD Heart Mate 3 left ventricular assist device under artificial circulation on June 29, 2018. Condition after resternotomy, explantation of LVAD Heart Mate 3, orthotopic bicaval heart transplantation under artificial circulation, hypothermia on July 17, 2024. Drive line explantation on July 17, 2024. Complication of the main disease: CHF with preserved EF, stage B (ANA/ACC), FC II (NYHA).

Outcome: Currently, the patient is being observed on an outpatient basis at the UMC Heart Center and at her place of residence. A clinical effect has been achieved in the form of an improvement in the patient's condition, quality of life, heart function (LVEF – 56%, EDV – 72 ml), and an increase in tolerance to physical activity (6-minute walk test – 390 m).

The patient's written informed consent for the dissemination of her medical information was obtained.

DISCUSSION

According to the clinical protocol of the Ministry of Health of the Republic of Kazakhstan and the European Society of Cardiology from 2019, peripartum cardiomyopathy (PPCM) is a form of DCM, which is characterized by the development of signs of heart failure during the last month of pregnancy or the first 5 months after childbirth, in the absence of any other causes of heart failure.

Also, according to a retrospective study conducted in South Korea between January 1, 2010 and December 31, 2012, it was concluded that patients with PPCM

were older, they were more likely to have preeclampsia and gestational diabetes, they were more often primiparous and had multiple pregnancies. In addition, these patients more often had cesarean sections, pregnancy-related complications, and had higher in-hospital mortality.

Like all cardiomyopathies, the severity of PPCM is determined by NYHA functional classes. Based on complaints, clinical, anamnestic, laboratory and instrumental data, a diagnosis of peripartum cardiomyopathy was established, as indicated by the manifestation and progression of the disease during the 2nd trimester of pregnancy. Due to the ineffectiveness of optimal drug therapy, as well as the lack of a donor heart, the patient was implanted with a left ventricular support device LVAD Heart Mate 3 as a «bridge to transplantation», which is a common practice in Kazakhstan given the underdevelopment of this type of treatment. Subsequently, with the presence of a posthumous donor, the patient underwent orthotopic bicaval heart transplantation. At the moment, heart transplantation remains the gold standard for the treatment of progressive CHF, which significantly improves the quality of life and functional status of the patient.

One-year survival after HT is ~90%, and the median survival is 12.5 years according to the European Society of Cardiology.

CONCLUSIONS

The clinical case clearly illustrates the complexity of managing patients with peripartum cardiomyopathy complicated by terminal chronic heart failure. Despite modern drug therapy and temporary improvement in the condition with the use of a left ventricular mechanical support device, the patient required orthotopic heart transplantation. The positive results obtained confirm the relevance of transplantation as a method of choice for progressive CHF, and also demonstrate the capabilities of modern cardiac surgery and a multidisciplinary approach in the context of a limited donor resource.

This case emphasizes the need for a systemic approach to the prevention of cardiovascular diseases in women of childbearing age, including risk factor control and competent pregnancy planning. Early diagnosis, comprehensive monitoring and individualized therapy of such patients can significantly reduce the risk of severe complications, improve the prognosis and increase the quality of life. In addition, the development of transplantation programs and increasing the availability of donor organs remain the most important areas for improving cardiac care in Kazakhstan.

Authors' contribution:

A. Kushugulova, M. Bekbossynova, A. Sailybaeva – concept and design.

S. Jetybayeva – data collection and preparation.

Zh. Aldanush, A. Taukelova – statistical analysis, writing.

A. Sailybaeva – editing.

Conflict of interest:

There are no among all authors.

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СЛУЧАЙ ОРТОТОПИЧЕСКОЙ БИКАВАЛЬНОЙ ТРАНСПЛАНТАЦИИ СЕРДЦА У ПАЦИЕНТКИ С ПЕРИПАРТАЛЬНОЙ КАРДИОМИОПАТИЕЙ

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Представлен клинический случай ортотопической бикавальной трансплантации сердца у пациентки с терминальной стадией хронической сердечной недостаточности, развившейся на фоне перипартальной кардиомиопатии. Заболевание манифестировало во втором триместре беременности, осложнилось тяжелой формой преэклампсии и прогрессирующей сердечной недостаточностью. На фоне отсутствия доступного донорского органа пациентке было имплантировано устройство механической поддержки левого желудочка HeartMate 3 в качестве «моста к трансплантации». Через четыре года после имплантации устройства механической поддержки левого желудочка, при появлении донора, успешно проведена ортотопическая трансплантация сердца. Случай демонстрирует эффективность мультидисциплинарного подхода в условиях ограниченного донорского ресурса и подчеркивает необходимость ранней диагностики, системной профилактики и развития трансплантационной службы для улучшения исходов у пациенток с перипартальной кардиомиопати.

Ключевые слова: трансплантация; перипартальная кардиомиопатия; хроническая сердечная недостаточность; устройство механической поддержки левого желудочка

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ПЕРИАНТАЛЬДЫ КАРДИОМИОПАТИЯСЫ БАР НАУҚАСҚА ОРТОТОПИЯЛЫҚ БИКАВАЛЬДЫ ЖҮРЕКТІ ТРАНСПЛАНТАЦИЯЛАУ ЖАҒДАЙЫ

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Бұл мақалада перинатальды кардиомиопатия (PPCM) фонында дамыған созылмалы жүрек жеткіліксіздігінің соңғы сатысы бар науқаста ортотопиялық бикавальды жүректі трансплантациялаудың клиникалық жағдайы ұсынылған. Ауру жүктіліктің екінші триместрінде көрінді және преэклампсияның ауыр түрімен және үдемелі жүрек жеткіліксіздігімен асқынды. Қолжетімді донор органы болмаған кезде науқасқа «трансплантацияға көпір» ретінде HeartMate 3 сол жақ қарыншаға көмекші құрылғы (LVAD) имплантацияланды. LVAD имплантациясынан кейін төрт жыл өткен соң, донор қолжетімді болған кезде ортотопиялық жүрек трансплантаты сәтті орындалды. Бұл жағдай шектеулі донорлық ресурстар контекстіндегі мультидисциплинарлық тәсілдің тиімділігін көрсетеді және PPCM бар науқастарда нәтижелерді жақсарту үшін ерте диагностикалау, жүйелі алдын алу және трансплантация қызметтерін дамыту қажеттілігін көрсетеді.

Кілт сөздер: трансплантация; перинатальды кардиомиопатия; созылмалы жүрек жеткіліксіздігі; сол жақ қарыншаға көмекші құрылғы