

Optimal Tactics for Surgical Treatment of Heart Damage with Burkitt's Lymphoma

RM Vitovskiy, VV Isaienko*, VF Onishchenko, OA Lozovyy, OA Pishchurin, DN Dyadyun, IV Martyshchenko and IG Yakovenko

N.M. Amosov National Institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine, Kyiv, Ukraine

***Corresponding Author:** VV Isaienko, N.M. Amosov National Institute of Cardiovascular Surgery of the National Academy of Medical Sciences of Ukraine, Kyiv, Ukraine.

Received: January 02, 2020; **Published:** February 13, 2020

Abstract

In the structure of the surgical pathology of the cardiovascular system, the tumor of the heart reach is less than 1%. The article describes an unusual case of the defeat of the heart with Burkitt's lymphoma in a male patient of 27 years old. The rapid and aggressive growth of Burkitt's lymphoma, which rapidly violates intracardiac hemodynamics, requires immediate diagnosis and adequate treatment, including surgical and antitumor therapy. In this case, attempts were made to use various diagnostic methods to determine the nature of the neoplasm and subsequent treatment methods with a positive long-term result.

Keywords: Heart Tumors; Burkitt Lymphoma; Surgical Treatment

Introduction

Primary malignant neoplasms originating directly from the heart tissue are extremely rare [4,10,11]. Tumors of the heart make up less than 1% in the structure of the surgical pathology of the cardiovascular system [1,9]. The literature mainly describes cases with a mixomas of the heart or metastatic lesions with primary localization of a malignant tumor in the lungs, pericardium, kidneys and other organs. At the same time, the problem of the effectiveness of surgical treatment of this type of pathology is of particular concern to cardiac surgeons.

Due to the difficulty of early diagnosis of malignant tumor of the heart (MHT), 50 - 80% of patients already have regional or distant metastases at the time of diagnosis, which significantly worsens their clinical condition and prognosis of surgical treatment [2,3,10,13,14]. A significant prevalence of the tumor process in the heart at the time of diagnosis leads to high (25 - 50%) hospital mortality in patients with malignant neoplasms. This is often associated with the need for a wider volume of surgical intervention, which may simultaneously include reconstruction of the heart chambers, prosthetics of its valves, coronary artery bypass grafting and, if necessary, implantation of a pacemaker [4,6,8,11].

The world literature describes successful cases of surgical treatment of malignant neoplasms of both the right and left parts of the heart with quite satisfactory long-term results against the background of repeated courses of chemotherapy and radiotherapy [1,10,12,14]. These reports indicate the possibility of successful surgical treatment of MHT at an early stage of the disease.

At present, cardiac surgeons from leading world clinics see a way out in the early differential diagnosis of malignant tumor growth,

as well as in immediate heart transplantation, which allows to achieve significantly better results [3,5-7]. The use of modern diagnostic tools, such as magnetic resonance imaging (MRI), computed tomography (CT), as well as a transthoracic and endovascular biopsy, allow to tentatively and sometimes absolutely accurately determine the nature of the neoplasm before surgery. Therefore, the problem of timely diagnosis and surgical treatment of MHT requires further study and maximum coverage.

In this article, we present our own observations about the features of the clinical course, methods of phased diagnosis and surgical treatment, as well as the immediate and long-term results of the operation of a patient with Burkitt's lymphoma of cardiac localization. As the primary MHT, this neoplasm is extremely rare.

In the available world literature, there are very few observations about heart damage with Burkitt's lymphoma. One study cites 22 reports of such tumor localization. Moreover, a fatal outcome had recorded in 10 (45%) cases. Most of these patients (9 out of 10) died within a few days, and one 6 weeks after being diagnosed. It is also reported that the longest survival period in patients with Burkitt's lymphoma in the heart was observed in 2 patients who were alive 36 months after the diagnosis of the neoplasm [3].

Aim of the Study

The aim of the work is to demonstrate the complexity of preoperative diagnosis and decision-making on surgical intervention in a patient with extensive tumor of the heart caused by Burkitt's lymphoma.

Case Report and Discussion

Patient Z., 29 years old, No 4218, admitted to the N.M. Amosov National Institute of Cardio-Vascular surgery of the National Academy of Medical Sciences of Ukraine on August 6, 2014, with a diagnosis of a heart tumor. It became known from the anamnesis that he considers himself sick since March 2014, when they began to worry about general weakness, shortness of breath and palpitations. Soon, paroxysms of flutter and atrial fibrillation joined in. Until that time, he was actively involved in sports, in particular, running. He entered the regional cardiology department on 07/31/2014, in a serious clinical condition with an attack of atrial fibrillation with a heart rate of 90-100 and signs of pericardial tamponade. Echo examination determined the presence of a large amount of fluid in the pericardial cavity, as well as the volumetric formation of the right atrium (RA), filling its cavity and extending along the walls of the atrium to the fibrous ring of the tricuspid valve (TV), which was regarded as a tumor tissue or blood clots. During puncture and drainage of the pericardial cavity, about 1000 ml of yellow turbid exudate was evacuated, which significantly improved the patient's condition. A cytological examination of the obtained fluid revealed atypical proliferation of mesothelium against the background of elements of inflammation. Additionally, a CT scan of the chest was performed. Moreover, in the basal parts of the lungs, the areas of consolidation of the lung tissue of irregular polygonal shape were determined on both sides. In the remaining parts of the lungs, focal and infiltrative changes were not detected.

At the time of admission to the N.M. Amosov National Institute of Cardio-Vascular surgery of the National Academy of Medical Sciences of Ukraine patient complained of severe general weakness, pain in the heart. During the examination, pallor of the skin, moderate swelling of the lower extremities attracted attention, the liver protrudes 2 cm below the costal arch. During auscultation, systolic murmur was heard in the projection of the apex of the heart of a small intensity. On an ECG, atrial flutter with a heart rate of 82 beats/min. Radiologically expressed deviations from the side of the contours of the heart shadow were not observed, cardiothoracic index of 0.45, pathological formations in the lungs were not detected. Complete blood count: hemoglobin - 115 g/l, red blood cells - $4,8 \times 10^{12}/l$, white blood cells - $14,2 \times 10^9/l$, white blood cell count within normal limits.

An echo was performed on the patient, revealing a sedentary fragmented tumor formation of $5,2 \times 5,8$ cm in size, with invasive lesions of the wall of the RA, the right ventricle (RV) and the fibrous ring of the TV. The formation partially fills the RA cavity and creates a moderate obstruction of the right atrioventricular opening with a pressure gradient of 15 mm Hg (Figure 1). Reverse current through the TV is not determined. The ejection fraction of the left ventricle (LV) is 66%. A small amount of fluid is detected in the pericardial cavity.

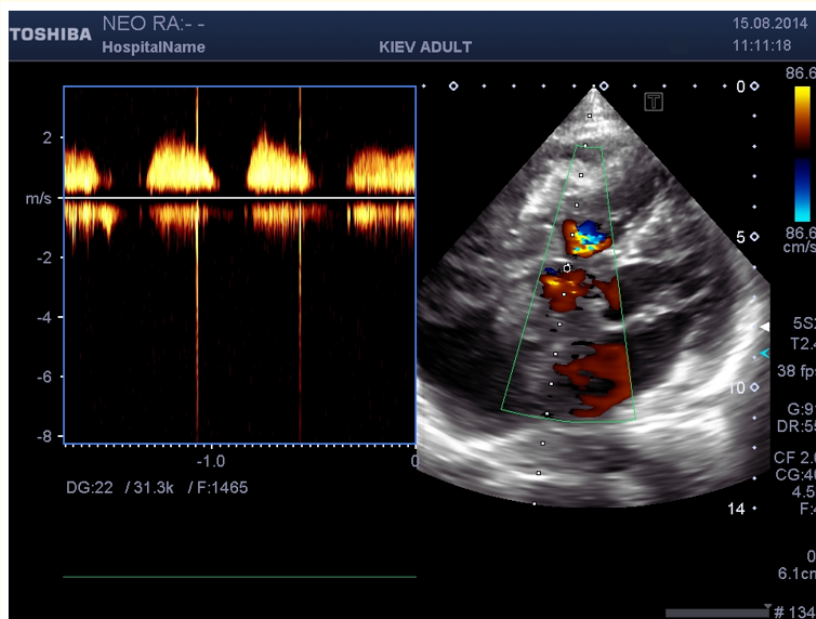


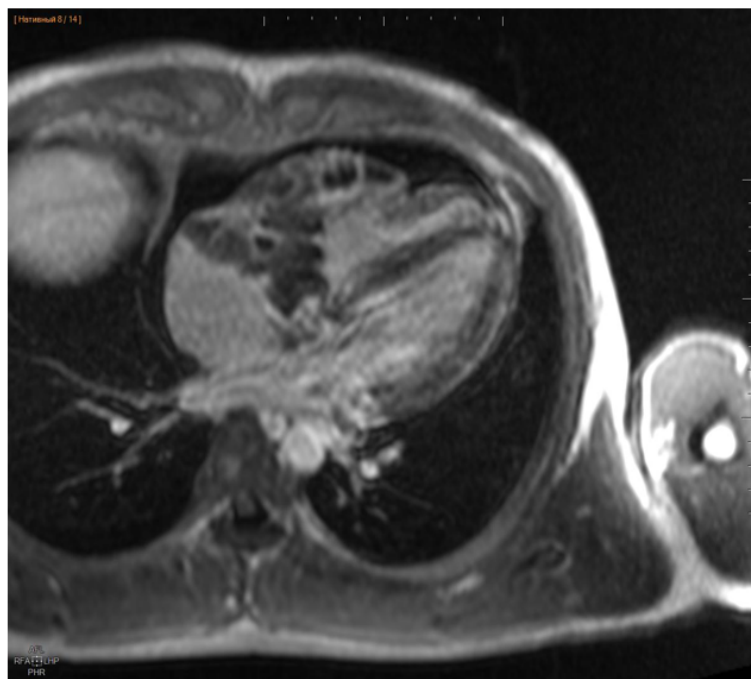
Figure 1: The color Doppler echo with the apical position of the ultrasound probe. Marked tumor of the right heart with obstruction of the right atrioventricular opening and systolic pressure gradient on it.

Thus, the results of the echo test showed that in this case we are talking about a possible malignant lesion of the heart.

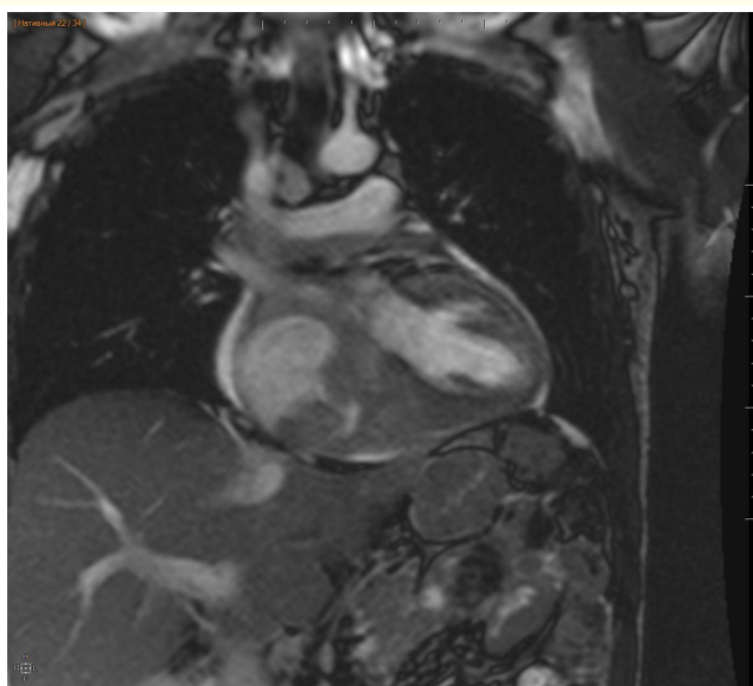
The spiral CT scan was performed to clarify the prevalence of the tumor process, given the suspicion of the malignant nature of the neoplasm. The examination showed the presence of a heart formation with a total size of 87 × 80 × 70 mm, located mainly in the region of the right departments, performing 1/3 of the RA cavity, sprouting into the right ventricle (RV) cavity and causing sharp stenosis of its inflow section, as well as deformation of the TV. The tumor germinates the lower third of the interatrial septum, the posterior wall of the left atrium (LA), the posterior and inferior septal LV segment at the basal level. The process extends to the trunk of the pulmonary artery (PA) and its right branch without signs of stenosis of the clearance. The neoplasm contour is clear, tuberous, differentiation of the adjacent myocardium is difficult.

The right coronary artery in the middle and distal third, as well as the distal third of the envelope branch of the left coronary artery, are “multiformly” surrounded by the formation. The coronary sinus was narrow in the proximal third by 30 - 50% due to compression formation. Also revealed lymphadenopathy of the posterior mediastinum and the root of the right lung. As a result of the study, a tumor of the heart with a suspected lymphoma was diagnosed.

For additional information about the distribution of the process, an MRI scan was performed on the patient. According to the data obtained, intramyocardially, in the projection of the atrioventricular sulcus, the formation is determined with a total size of 81 × 58 × 81 mm, which extends to the lateral wall of the right atrium and ventricle, the posterior basal LV, and also the germinating lower third of the atrial septum. The neoplasm performs the cavity of the RA and the RV, causing their reduction and deforming the TV. The inflow of the RV is slit-like. The distribution of the formation to the lower part of the PA trunk and its right branch, without signs of clearance stenosis, is also determined (Figure 2).



A



B

Figure 2: The MRI image showing the pronounced spread of the tumor process in the right heart with reduction of the RV cavity and deformation of the TV (A).

Damage to the lower part of the trunk of the PA and its right branch without signs of stenosis of the clearance (B).

In the future, an attempt was made to biopsy the neoplasm by the endovascular method, with the aim of differential diagnosis of tumor damage and to determine treatment tactics. Given the significant filling with the tumor tissue of the cavities of the RA and RV, the probability of successful removal of the tumor tissue with special forceps seemed rather high. The five-biopsy specimens collected from the RA and RV, by access through the right femoral vein. However, despite the accompanying procedure with echo, histological examination in the biopsy determined exclusively myocardial tissue, and tumor cells were absent.

The expediency of surgical intervention in this clinical case was largely determined by the histological nature of the tumor. The desire to get an accurate diagnosis prompted us to resort to another method for identifying the neoplasm - a transthoracic transcutaneous puncture biopsy.

Computer remodeling of tumorous lesions of the heart demonstrated the presence of tumor tissue with myocardial replacement on the anterior wall of the heart. Based on this, a biopsy sample has taken from the RV wall trans-thoracically and under the control of the echo using a biopsy needle. Since this manipulation may accompanied by the development of hemopericardium up to cardiac tamponade, sometime after its completion, an echo study of the pericardial sac cavity was repeated. No signs of bleeding were detected. It was also not possible to detect tumor cells in the biopsy obtained by this method.

At the same time, the next echo control of the neoplasm revealed signs of its rapid growth compared with the data obtained when the patient was admitted to the clinic. Almost complete filling of the RA cavity with a tumor with significant progressive obstruction of the right atrioventricular opening and high pressure in the RA segment above the tumor was noted. The critical condition of the patient associated with this necessitated an emergency operation, despite the lack of reliable information about the histological nature of the neoplasm.

The patient underwent a palliative operation on August 18, 2014, aimed at eliminating the obstruction of blood flow to the right atrioventricular hole. The longitudinal median sternotomy was chosen as access to the heart. The pericardial cavity is free of adhesions, up to 50 ml of hemolysis blood identified (a consequence of a biopsy). The dense, tuberous formation is observed, when examining the heart, towering above the RV and occupying almost its entire front surface. The tumor invasively invades the ventricular wall, passes to the diaphragmatic surface of the heart and to the RA, leaving the small lateral surface of the latter intact in the region of the inflow of the upper and lower vena cava. Palpation is determined by the tumor invasion of the basal LV, trunk and right branch of PA (Figure 3).

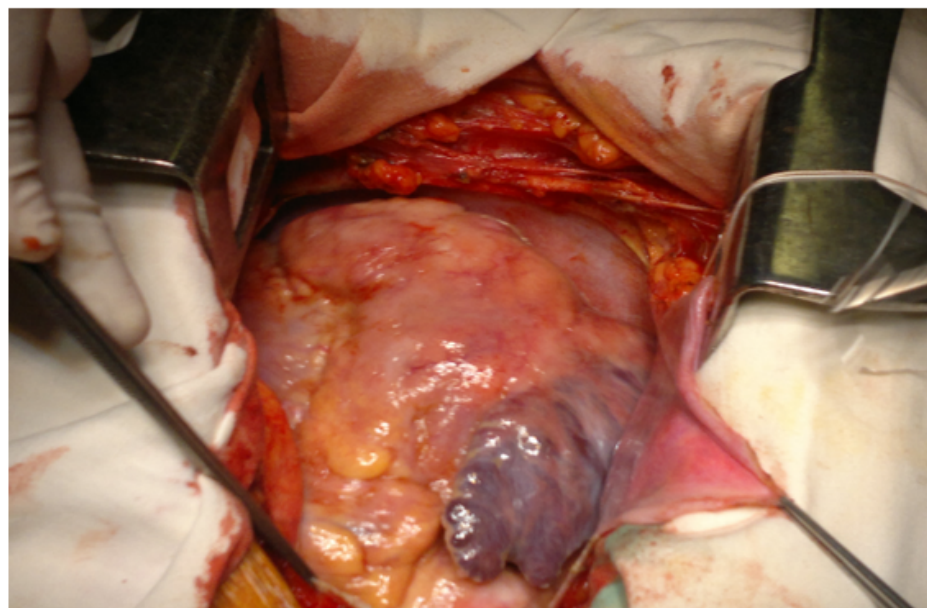


Figure 3: The tumor damage to the right heart.

Despite significant filling of the RA cavity by the neoplasm, the inferior vena cava was cannulated through its wall, the superior vena cava was cannulated 2,5 cm above the point of inflow into the RA. After connecting the cardiopulmonary bypass, the collapsed wall of the RA marked the contours of the tumor (Figure 4).

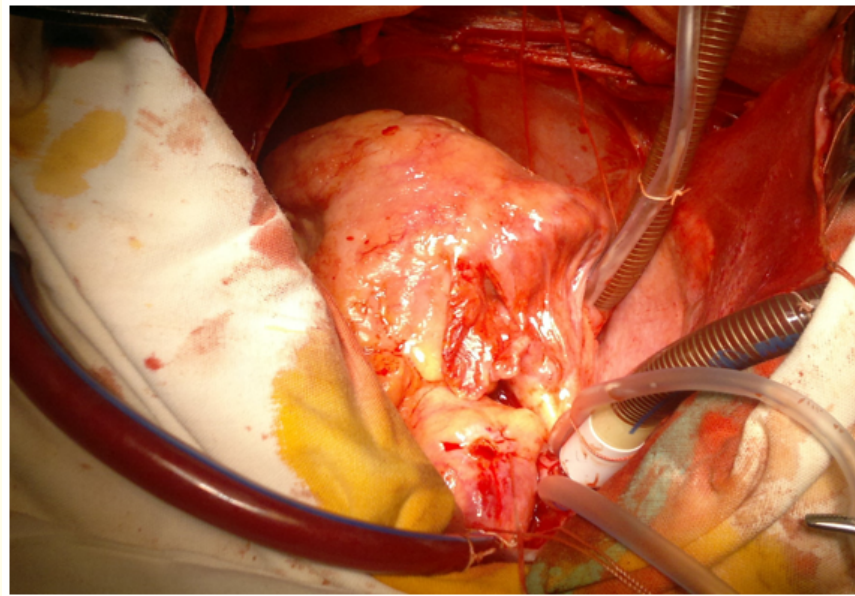


Figure 4: The view of the heart damaged by the tumor after connecting the cardiopulmonary bypass and blood sampling from the RA cavity (the tumor rises above the myocardium).

After dissection of the anterior wall of the RA in its unchanged part, it became evidently that the tumor tissue fulfills the entire atrial cavity, has multiple attachment sites and unequal density - from tissue similar to unorganized blood clots to dense connective tissue. Leaving only a narrow slit-like opening, the neoplasm tightly covers the TV, thereby significantly preventing the free flow of blood to the RV (Figure 5). Wherein, TV was not involved in the pathological process.

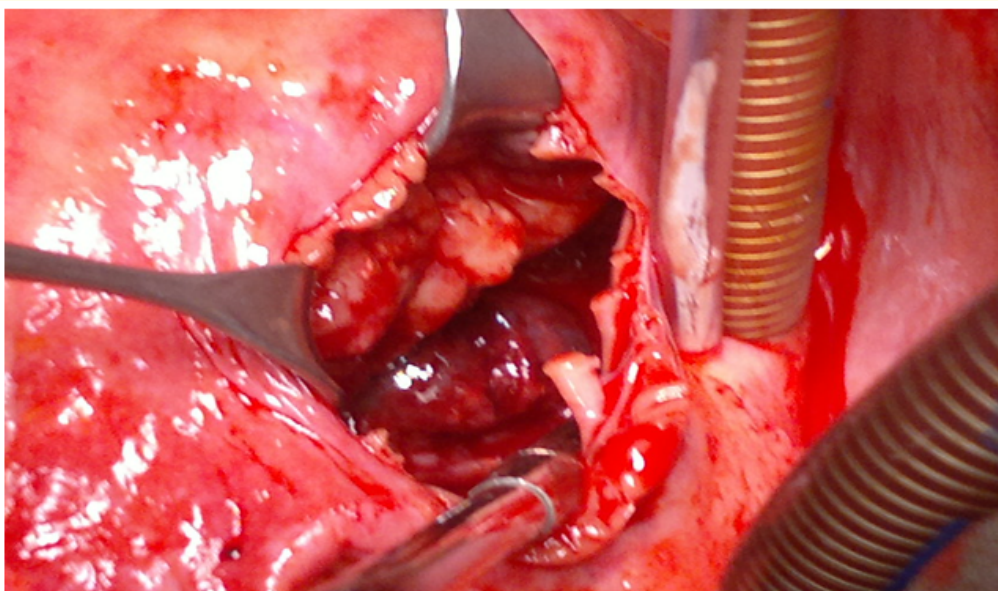


Figure 5: The tumor tissue filling the cavity of the RA in the clearance of its incision.

The tumor tissues are maximally removed by the “acute” method, from the RA in such a way as to ensure free blood flow to the TV (Figure 6).

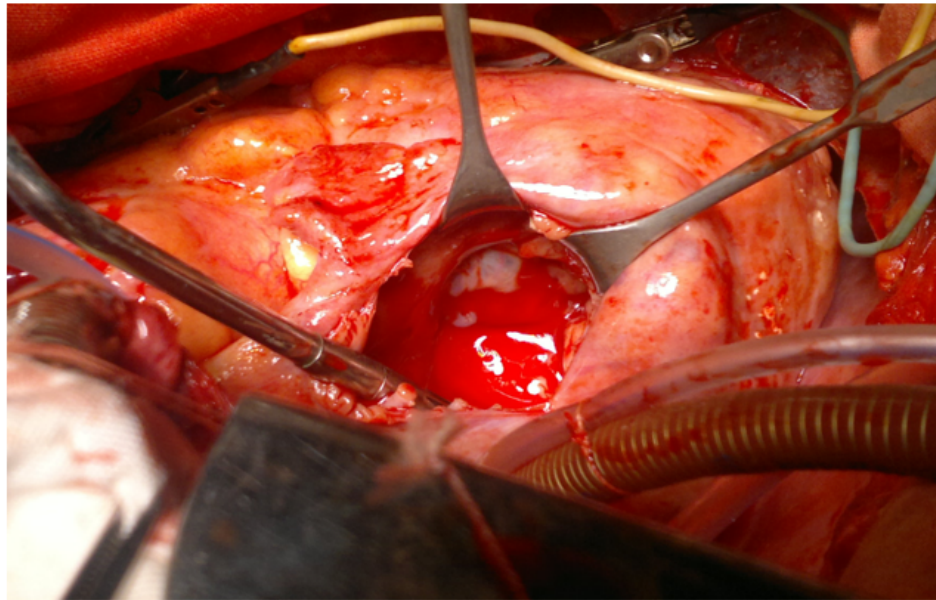


Figure 6: The stage of operation - RA is freed from tumor tissue, through its incision, TV is freely visualized.

The performed hydraulic test demonstrated the competence of the TV, and examination of the subvalvular space testified to a sufficient value of the free RV cavity. Further manipulations on the “open” heart were deemed inappropriate. After the restoration of independent cardiac activity and with minimal inotropic support, the operation was completed.

The histopathological study of the distant neoplasm classified it as lymphoma. The patient repeatedly needed detoxification therapy in the postoperative period, in the intensive care unit and in the department; otherwise, it proceeded without complications.

The immunohistochemical study of the material obtained during the operation was carried out at the Kiev City Clinical Oncology Center and at the R.E. Kavetsky Institute of Experimental Pathology, Oncology and Radiobiology of the National Academy of Medical Sciences of Ukraine. The patient was also consulted at the National Cancer Institute of Ukraine, where a molecular cytogenetic analysis of tumor tissue was performed to verify the diagnosis and determine the tactics of further treatment. The neoplasm in question is defined as Burkitt's lymphoma, as a result of all the studies.

The patient was discharged on the 14th day after the operation in satisfactory condition and sent for further treatment to the hospital at the place of residence, where he underwent several courses of antitumor therapy, including complex chemotherapeutic, antiviral, hormonal and immunomodulators treatment. Six-fold courses of antitumor therapy carried out according to the DA-R-EPOCH program (cyclophosphamide, vincristine, doxorubicin, prednisolone, etoposide, “MabThera”). The patient's condition improved significantly as a result of treatment, as indicated in the epicrisis of case histories, manifestations of heart failure decreased.

The study of the four month long-term results of surgical and conservative anti-relapse treatment using echo and MRI showed not only the absence of relapse of the neoplasm in the RA cavity, but also a pronounced decrease in the total volume of heart damage by the tumor process (Figure 7).

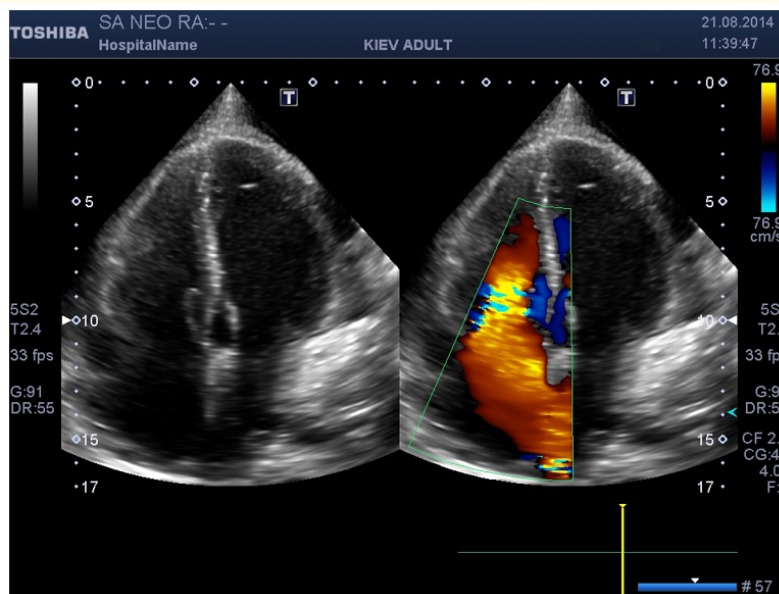


Figure 7: The color Doppler echo at the apical position of the ultrasound probe, demonstrating significant tumor regression with no TV obstruction.

Moreover, the study of long-term results 3 and 5 years after surgery using the same diagnostic methods determined the complete absence of tumor tissue in the heart.

Thus, the obtained data of instrumental research methods, as well as the satisfactory condition of the patient in the long-term follow-up period, testified to the effectiveness of the treatment and the right optimal tactics.

Conclusion

1. Burkitt's lymphoma is an extremely rare neoplastic heart disease that must be consider when conducting differential diagnosis of intracardiac neoplasms.
2. The rapid and aggressive growth of Burkitt's lymphoma, which rapidly violates intracardiac hemodynamics, requires immediate diagnosis and adequate treatment, including surgical and antitumor therapy. Only in this case should one count on a positive result with a possible complete recovery of the patient, even with a significant amount of myocardial damage by the tumor process.

Bibliography

1. A Hoffmeier JR., et al. "Cardiac tumors - diagnosis and surgical treatment". *Deutsches Aerzteblatt International* 111.12 (2014): 205-211.
2. A Gowdamarajan., et al. "Therapy for primary cardiac tumors: is there a role for heart transplantation?" *Current Opinion in Cardiology* 15.2 (2000): 121-125.

3. Chan., *et al.* "Burkitt lymphoma presenting as an intracardiac mass: case report and review of literature". *American Journal of Case Reports* 3.17 (2016): 553-558.
4. SM Yuan A., *et al.* "Imaging morphology of cardiac tumors". *Cardiology Journal* 16.1 (2009): 26-35.
5. JM Jimenez Mazuecos R., *et al.* "Is heart transplantation for primary cardiac sarcoma a useful therapeutic option?" *Revista Española de Cardiología* 56.4 (2003): 408-411.
6. A Kośmider., *et al.* "23-year experience on diagnosis and surgical treatment of benign and malignant cardiac tumors". *Archives of Medical Science* 9.5 (2013): 826-830.
7. Habertheuer A., *et al.* "Primary cardiac tumors on the verge of oblivion: a European experience over 15 years". *Journal of Cardiothoracic Surgery* 10 (2015): 10.
8. Cresti A., *et al.* "Incidence rate of primary cardiac tumors: a 14-year population study". *Journal of Cardiovascular Medicine* 17 (2016): 37-43.
9. Burazor I., *et al.* "Primary malignancies of the heart and pericardium". *Clinical Cardiology* 37 (2014): 582-588.
10. Isogai T., *et al.* "Factors affecting in-hospital mortality and likelihood of undergoing surgical resection in patients with primary cardiac tumors". *Journal of Cardiology* 10 (2016): 1016.
11. Barreiro M., *et al.* "Primary cardiac tumors: 32 years of experience from a Spanish tertiary surgical center". *Cardiovascular Pathology* 22 (2013): 424-427.
12. Padalino MA., *et al.* "Surgery for primary cardiac tumors in children: early and late results in a multicenter European Congenital Heart Surgeons Association study". *Circulation* 126 (2012): 22-30.
13. Dias RR., *et al.* "Mortality and embolic potential of cardiac tumors". *Arquivos Brasileiros de Cardiologia* 103 (2014): 13-18.
14. Barnes H., *et al.* "Clinicopathological and surgical experience with primary cardiac tumors". *Asian Cardiovascular and Thoracic Annals* 22 (2014): 1054-1058.

Volume 7 Issue 3 March 2020

©All rights reserved by VV Isaienko., *et al.*