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Case Report

Successful Heart Transplantation Following Decompressive Craniectomy in a Patient with Restrictive Cardiomyopathy and Extensive Stroke in the Region of the Right Middle Cerebral Artery

Salih Gulsen*

Baskent University Medical Faculty Hospital - Neurosurgery, Maresal Fevzi Cakmak cad. 10, sok. No: 45, Ankara 06540, Turkey

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Key words: Decompressive Craniectomy; Middle Cerebral Artery; Restrictive Cardiomyopathy; Stroke; Decompressive Craniectomy; Middle Cerebral Artery; Restrictive Cardiomyopathy; Stroke.

***Correspondence:** Salih Gulsen, MD. Department of Neurosurgery, Faculty of Medicine, Baskent University Medical Faculty Department of Neurosurgery, Fevzi Cakmak Caddesi 10. Sokak no: 45, Cankaya, Bahcelievler, Ankara 06490. Turkey. Business Phone: 0090 312 212 68 68 /1362. FAX: 0090 3122237333. E.mail: salihgulsen07@gmail.com

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Abstract

Restrictive cardiomyopathy (RCM) in children is associated with a greater risk of embolic stroke than are other congenital heart diseases. After diagnosis, 50% of children with RCM die within 2 years without heart transplantation. As such, all RCM patients are placed on the heart transplantation list and must wait for an appropriate heart for transplantation. Every type of embolic stroke can occur while waiting for a donor heart; therefore, the cardiovascular team must initiate antithrombotic therapy at time RCM is diagnosed. Some pediatric RCM patients experience embolic stroke (50% are the cerebral type) despite antithrombotic therapy, including acetylsalicylic acid, warfarin, and heparine. Neurosurgeons working in hospitals that perform organ transplantation expect to see RCM cases with restrictive large cerebral infarct. We think that decompressive craniectomy should be performed as soon as possible after determining the clinical condition of any patient with RCM and a large right middle cerebral artery (MCA) infarct.

Introduction

The annual incidence of restrictive cardiomyopathy (RCM) is approximately 0.035 per 100,000 children in Australia and the US [1]. Biatrial enlargement, relatively normal systolic function, and poor ventricular filling due to restrictive physiology are characteristic of this cardiac disorder [2]. Intracardiac thrombus and embolic thrombus occur in 0%-42%and 12%-33% of children with RCM, respectively [3-7]. Cardiac surgeons and cardiologists favor performing cardiac transplantation as soon as possible following the diagnosis of RCM due to poor prognosis and poor response to medical therapy, and the risk of rapidly progressing elevated pulmonary vascular resistance and sudden death. Moreover, the risk of embolism is high and cerebrovascular embolic events account for 50% of all embolic events in RCM patients [3, 5, 8, 9]. Cardiac transplantation should be performed as soon as possible in patients with RCM experiencing embolic events. Following the diagnosis of RCM >50% of patients die within 2 years without cardiac transplantation [3, 5, 8, 9]. Neurosurgeons prefer performing decompressive craniectomy soon after

extensive middle cerebral artery (MCA) region infarct, even in patients without midline shift or herniation, because development of midline shift and extensive edema eventually occur due to ischemia and energy failure [10-12]. A delay >48 h can result in a vegetative state or death in patients with an occluded MCA. As such, decompressive craniectomy should be performed within 48 h of an embolic event [10-12]. A Medline search of the English-language literature conducted in February 2013 using various combinations of 10 key words regarding cardiac transplantation following decompressive craniectomy in a patient with RCM and total occlusion of the right MCA did return any results. To the best of our knowledge the first such case is presented herein.

Case Report

A 16-year-old male RCM patient (height: 160 cm; weight: 54 kg) with left dominant hemisphere was hospitalized for cardiac transplantation. Cardiac surgeons and pediatric cardiologists (CSPS) had diagnosed his disease in 2008, at which time he was placed on the waiting list for heart transplantation and antithrombotic therapy (acetylsalicylic acid 300 mg in a day) was initiated. Acetylsalicylic acid treatment was stopped and low-molecular-weight heparin (LMWH) was started during the cardiac transplantation preoperative period. While waiting for cardiac transplantation in the cardiovascular surgery department he experienced numbness and tingling in his left arm and left leg. The cardiovascular surgeons then consulted the patient to the neurology and neurosurgery departments overnight.

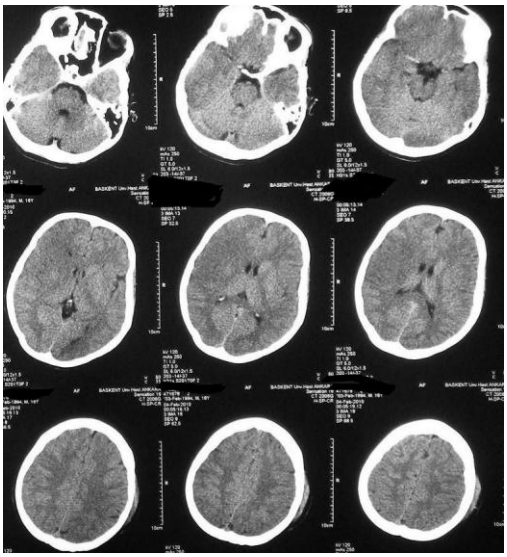

Figure 1: Axial CT shows a hyperdense MCA and subtle low density in the territory of the right MCA; the right Sylvian cistern is not observed.

Cranial CT (CCT) showed early abnormal findings - hemispheric sulcus effacement in the right cerebral hemisphere, loss of a hypodense appearance of the right Sylvian cistern, and a hyperdense right MCA (Fig. 1) - and his Glasgow Coma Scale (GCS) score was 15/15, with no loss of power. Diffusion weighted MRI (DW-MRI), apparent diffusion coefficient (ADC) mapping, and magnetic resonance angiography (MRA) were performed 2 hours after the stroke; which showed complete occlusion of the right MCA via MRA (Fig. 2), a bright right MCA acute infarction via DW-MRI (Fig. 3), and a low signal in the same location via the corresponding ADC map (Fig. 4), confirming the presence of restricted diffusion in the territory supplied by the right MCA, but no midline shift.

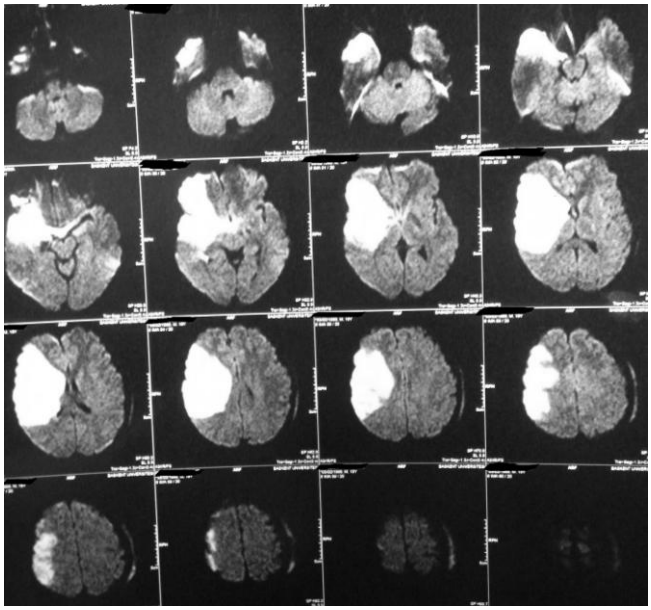

Figure 2: Axial DW-MRI shows a bright right MCA acute infarction.

Approximately 27 hours later, the patient developed left sided hemiplegia, and his GCS score dropped from 15/15 to 12/15. We performed CCT again (Fig. 5), which showed an 11-mm midline shift from the right to the left and a hypodense appearance of the region supplied by the right MCA. Once we determined that his GCS score was 12/15, we started rapid mannitol infusion while transferring of the patient to the surgical suite. We then performed a large right frontotemporoparietal craniectomy using a high-speed drill in order to decompress the cerebrum. Next, we opened the duramater; we observed no pulsation of the cerebrum, which was edematous and much harder than usual. We did not close the duramater with stiches, but covered the duramater to the cerebrum. The scalp was then closed with sutures. Lastly, we incised the skin over the left lower abdominal quadrant and arranged a place below the skin and above the muscle fascia of this region and inserted the bone that was removed during craniectomy.

The time from the onset of stroke to decompression was about 28 hours. Post surgery the patient remained intubated and on ventilator support,

and was transferred to the ICU. The patient's arterial blood gas parameters were as follows: pH: 7.42; pCO_2: 39 mmHg; pO_2: 93 mmHg; HCO_3: 14 mEQ L O_2 saturation: 99%, confirming that his status was stable.

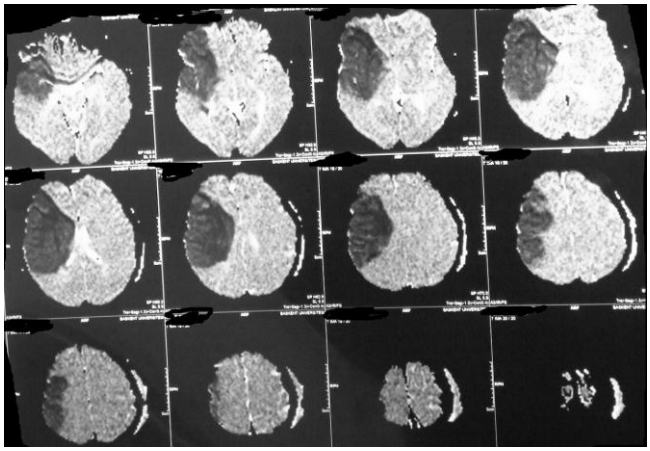

Figure 3: The correspondingADC map shows low signal intensity on the right side,location, confirming the presence of restricted diffusion.

The ventilator was set to SIMV mode and Diprivan® (propofol) 250 mg h and high-dose mannitol infusion was started. After 24 hours of ventilation we stopped Diprivan® and mannitol, and 1 hours later we extubated the patient. He was able to move his left leg and could raise it slightly when asked, and he was cooperative, but could not move his left arm hand 2 hours post extubation.

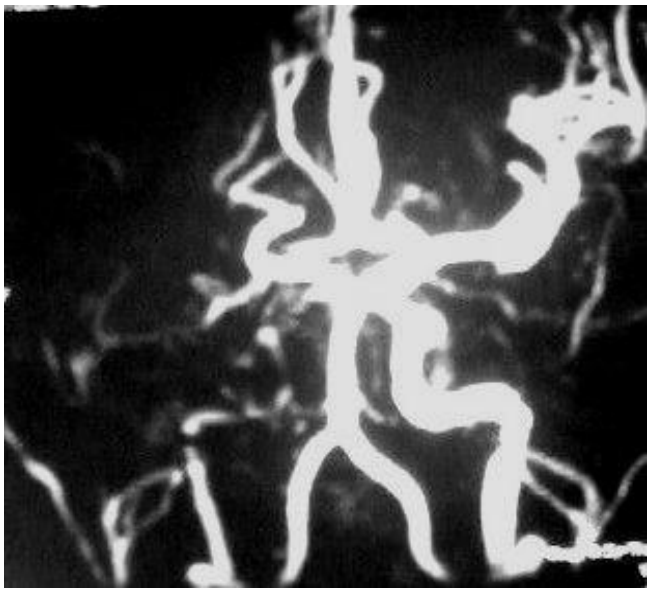

Figure 4: MRA frontal view shows non-visualization of the right MCA.

The patient began physiotherapy 4th day of post decompressive craniectomy. In the physiotherapy department the patient was fully conscious, oriented, and communicated well. CCT performed on 4th day of the craniectomy showed no edema or midline shift, but did show a large cystic encephalomalacic region previously supplied by the right MCA (Fig. 6). The patient underwent daily physiotherapy for his chest and limbs, and intensive exercise for his left hand and left leg. He improved steadily, and he was able to walk and 32nd day of the surgery without assistance and began to move his left arm and forearm. But he still could not move his left hand or fingers.

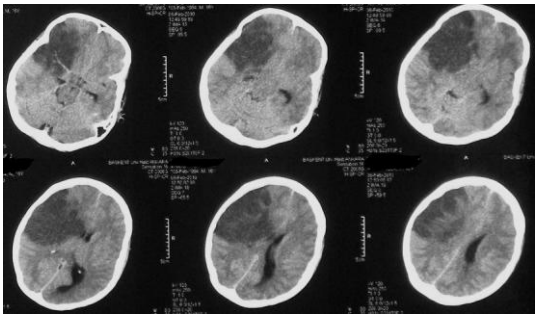

Figure 5: Axial CT shows a large low-density infarction in the region supplied by the right MCA, midline shift from right to left, and closing of the right lateral ventricle due to increased intracranial pressure.

The patient was recovering well and 42nd day of post decompressive craniectomy, so we performed cranioplasty and used the bone that was inserted in his abdomen. Three days later, we performed CCT, and it showed the bone that was replaced and the encephalomalasic region previously supplied by the right MCA, but no midline shift or other pathological signs (Fig. 7). Following his stroke, the cardiovascular team had removed the patient from the cardiac transplantation wait list due to his clinical condition, but his recovery was sufficient to persuade them to again place him on the list.

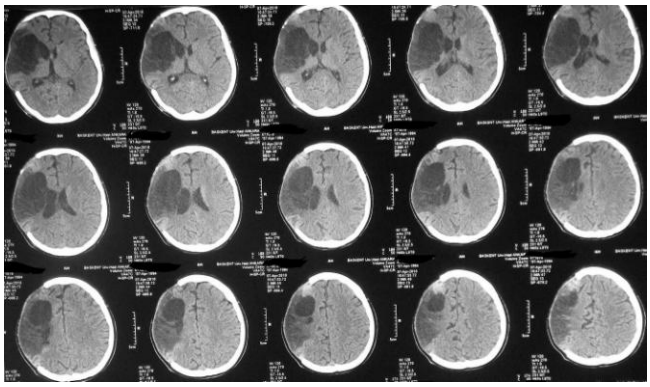

Figure 6: Axial CT shows the large cystic encephalomalacic region that was supplied by the right MCA before the embolic stroke, and no signs of elevated intracranial pressure. Both lateral ventricles appear symmetrical and there is no midline shift. In addition, the defect in the craniectomy region is seen.

The patient was discharged with prescriptions for physiotherapy and oral antiepileptic medicine and subcutaneous LMWH 1 mg/kg/d. A donor was eventually located for the patient and the cardiovascular team successfully transplanted a heart on 28 October 2010. The patient was discharged after

his recovery with immunosuppressive and antithrombotic therapy, including acetylsalicylic acid and clopidogrel. At the time this report was written (28 months post transplantation), the patient was attending high school, playing soccer (despite his mother's objection), and was able to raise his left arm as high as his head, but could not use his left hand or fingers.

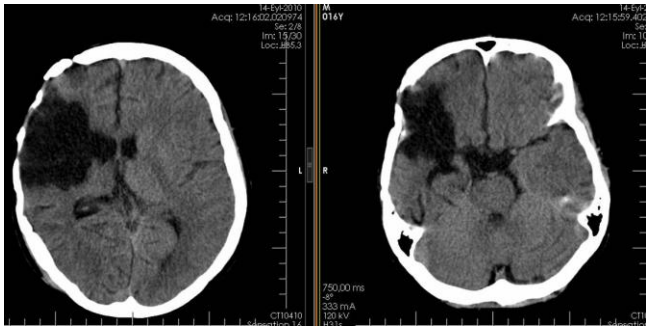

Figure 7: Axial CT appears similar to the figure 6, but does show the bone that was replaced during cranioplasty.

Discussion

Cerebrovascular stroke accounts for 50% of all embolic events in children with RCM, whether or not they have had intracardiac thrombus [3-7]. Children with RCM are at risk of thrombus formation due to turbulent blood flow and blood stasis - consequences of the contours of a dilated atrium, in terms of blood flow mechanics [3-7].

In addition, an abnormal heart rhythm causes even greater turbulence and may also cause a thrombus to embolize. As such, antithrombotic therapy should be initiated at the time RCM is diagnosed in children due to the high risk of embolism, but no comparative study has reported the effectiveness of antiplatelet therapy against vitamin K antagonists in children with RCM [3-7]. Nonetheless, any child with RCM that has had an embolic stroke should be given anticoagulant therapy instead of antiplatelet therapy in order to prevent further embolic strokes, and should be immediately placed on a transplant wait list [5, 7, 8].

Moreover, the sequelae of embolic strokes can preclude heart transplantation in children with RCM. Cardiac surgeons and pediatric cardiologists diagnosed the presented patient's disease in 2008 and placed him on the wait list for heart transplantation. They initiated antithrombotic therapy using acetylsalicylic acid 300 mg per day. When he was consulted to our department we decided to follow-up via observation for a while because his examination showed minimal loss of strength on the left side and no cognitive dysfunction. We thought that giving subcutaneous LMWH would be preventive, and it would not hinder in case of acute surgical intervention necessity would come up for the patient, and we did not start mannitol treatment at the beginning of the stroke. Clinical investigations reported that use of LMWH for acute ischemic stroke in children is safe [13-17], but no comparative study showing the efficacy or superiority of these drugs including acetylsalicylic acid,warfarin,heparine and ultra-fractionated heparine [18-21].

We decided that thrombolysis was not an appropriate treatment for the presented patient because intra-arterial use of thrombolytic treatment is not recommended for children outside of clinical trials due to the high risk of complications, and due to the lack of consensus on the use of thrombolytic treatment in adolescents with acute ischemic stroke [22-24]. We promptly performed decompressive craniectomy, because delaying the procedure and performing it with patients after their clinical condition declines negatively affects functional rehabilitation in patients with right MCA occlusion and can even result in a post surgical vegetative state [25-27]. The decision to perform decompressive craniectomy in cases of left MCA occlusion due to embolic stroke is more compelling for neurosurgeons than in cases of right MCA occlusion in patients with a dominant left hemisphere [25-27]. If the presented RCM patient had had an occluded left MCA, we would not have performed the surgery. Moreover, patients with RCM and an occluded right MCA have a greater risk than those without RCM of shorter postoperative survival and lack of independent functioning following cranial surgery. We think that a multidisciplinary approach is essential for the treatment of patients with RCM and right MCA occlusion. Once right MCA occlusion is verified via CCT or DW-MRI, ADC, and MRA, neurosurgeons should perform decompressive craniectomy without waiting for clinical deterioration and the development midline shift (based on cranial MRI or CCT) in order to prevent further complications, including subfalcine and transtentorial herniation, and brainstem hemorrhage.

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