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Simultaneous Pulmonary Embolism and Ischemic Stroke in a Patient with Cor Biloculare after Glenn Anastomosis

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ABSTRACT

Background: Cor biloculare, two-chambered heart due to the absence of atrial and ventricular septa, is a rare congenital heart anomaly. For Cor biloculare and other cardiac defects with single ventricle physiology, Glenn anastomosis has been developed as a palliative procedure. Thrombosis secondary to Glenn anastomosis in the patient with Cor biloculare could pose a serious threat to the survival, and has never been reported before.

Case report: We report the case of a 27-year-old patient, with past history of Glenn anastomosis that was performed 7 years ago for the palliation of Cor biloculare. She presented with pulmonary embolism and ischemic stroke simultaneously at 7 days after Cesarean section. Due to her critical status, systemic anticoagulation with low-molecular-weight heparin was started immediately, followed by lifelong warfarin therapy. Pulmonary embolism regressed and neurological symptoms were considerably diminished after the anticoagulation treatment.

Conclusion: This case illuminates the potential risk of thrombotic events in this patient cohort and demonstrates that anticoagulation therapy is an effective, secure, and appropriate for the management of this disease.

INTRODUCTION

Cor biloculare is a rare congenital heart defect characterized by a single atrium and a single ventricle being communicated through an atrioventricular valve. This defect is usually associated with various other malformations [Wang 2014]. Advances in medical and surgical care greatly improve the survival and quality of life for cor biloculare patients. Glenn anastomosis has evolved into a standard procedure for intermediate or long-term palliation of single ventricle physiology, in which end-to-side shunt between superior vena cava and right pulmonary artery is created [Yuan 2009; Jonas 1994]. Parallel to the progress on surgical intervention, thrombosis is a fatal complication which has drawn more attention than ever before.

In this report, we present a case of simultaneous pulmonary embolism and ischemic stroke in a cor biloculare female after Glenn anastomosis

CASE REPORT

A 27-year-old female patient was admitted to the hospital because of antepartum hemorrhage accompanied by lower abdominal pain in 2014. She had a history of cyanotic congenital heart disease. In 2007, she complained of progressive exercise intolerance and increased cyanosis. DSA revealed dextrocardia, single atrium and single ventricle of the right ventricular type, stenosis in pulmonary artery and malposition of the great artery. To increase pulmonary blood flow and thereby, increase oxygen saturation, the Glenn anastomosis was performed.

Premature labor was diagnosed and she underwent Cesarean section at the gestational age of 34 weeks plus 2 days. The procedure was uneventful and she showed no apparent distress. On day 7 following the delivery, the patient developed a sudden onset of dizziness, chest tightness, slurred speech and weakness in right extremities when using the toilet. She had a blood pressure of 125/70 mmHg, with a heart rate of 92 bpm, a respiratory rate of 24 breaths per minute, and her blood oxygen saturation level was 85% on 3 L/min of oxygen. Upon physical examination, she was noted to have pronounced clubbed-fingers and toes. In addition, bluish color to the skin and mucous membranes were also observed. The neurologic examination revealed central facial paralysis and hemiparesis on the right side with muscle strength grade 0/5. On cardiovascular system examination, systolic murmurs of grade 3/6 could be heard over the 2nd right intercostal space.

Laboratory tests showed that the pH value for arterial whole blood was 7.457 (reference range, 7.350 to 7.450), the blood level of carbon dioxide partial tension was 58 mmHg (reference range, 35.0 mmHg to 45 mmHg), the level of D-dimer was 3.31 mg/L (reference range, 0.00 mg/L to 0.5 mg/L), and the elevated fibrinogen level was 5.90 g/L (reference range, 2.00 g/L to 4.00 g/L). Tests of complete blood

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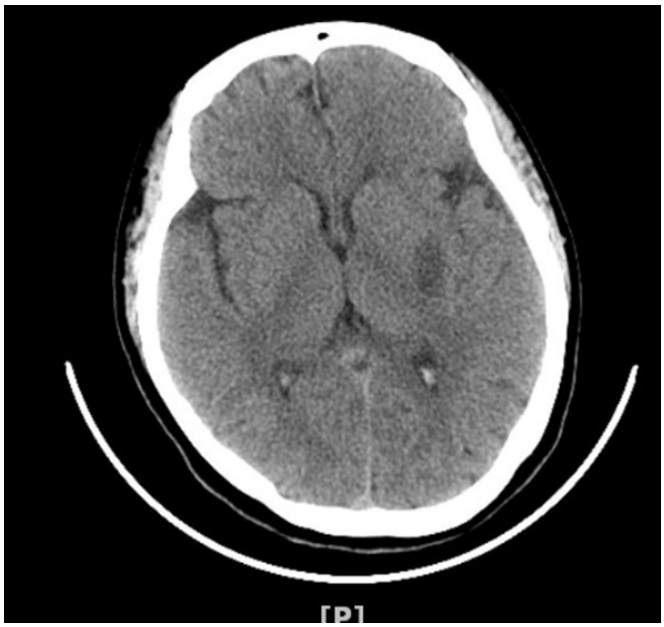

Figure 1. Head CT: left basal ganglia infarct.

count, renal function and liver function were normal in conjunction with the blood biochemistry screening.

The electrocardiogram showed sinus rhythm with abnormal Q waves and ST-T changes. A wedge-shaped infarction in left basal ganglia could be observed on cranial computed tomography. Computer tomography pulmonary angiography (CTPA) demonstrated filling defects within the left inferior pulmonary artery and the right lower lobe branches due to pulmonary embolism. Lower extremity venous ultrasound was negative for venous thrombosis.

She was diagnosed with pulmonary embolism, cerebral infarction, and transferred to the ICU for treatment. The patient was placed on systemic anticoagulation with low-molecular-weight heparin at 4000IU q12h. After 5 days, warfarin was taken orally.

The patient's clinical condition improved gradually, and she was discharged after 16 days with slight symptoms of dyspnea and muscle strength grade 5-/5 in right-sided extremities. No recurrence of stroke and pulmonary embolism was later found.

DISCUSSION

In previous studies, there is scant data relating to the simultaneous occurrence of ischemic stroke and pulmonary embolism. And in most reports available, researchers mainly focus on its association with patent foramen ovale [Gunta 2012; Omar 2013]. Herein, we demonstrate the simultaneous occurrence of pulmonary embolism and ischemic stroke in a female with complex congenital heart disease after delivery. This case is of great rarity as cor biloculare only comprises less than 1% of all cardiac anomalies [Temple 1981]. Furthermore, because of the poor hemodynamic condition, patients rarely survive to adulthood, not to mention fall pregnant.

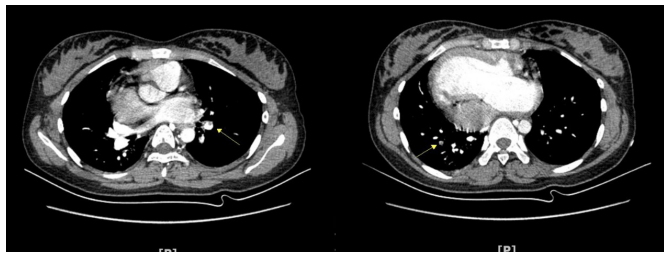

Figure 2. CTPA: embolism in the proximal branch of left lower pulmonary artery and distal branch of right lower pulmonary.

Thrombosis, secondary to Glenn anastomosis, mostly occurs around the anastomosis which mainly causes pulmonary embolism [Kozicka 2015; Oski 1996; Bonnel 2015]. Underlying mechanisms may include low venous flow state, elevated postoperative superior vena cava vein pressure, abnormal level of coagulation factors, presence of prosthetic material and increased platelet reactivity [Forbes 1997; Ravn 2001; Odegard 2002; Iannoli 2005]. While the abovementioned occurs, dysfunction of endothelium in vessels and shift towards hypercoagulable state in the postpartum period have been reported [Wiszniewska 2013]. Additionally, long-term bed rest after Cesarean section can result in static blood flow. According to Virchow's triad, hypercoagulability, blood flow stasis and endothelial injury could be responsible for the thrombosis.

Simultaneous thrombotic occlusions at two distant vascular territories are rare. However, in our opinion, it is not a mere coincidence. Although venous duplex ultrasonography and echocardiogram showed no obvious embolic source in this case, thrombosis in the anastomosis region and superior vena cava has been found in patients after Glenn surgery [Forbes 1997; Koutlas 1996; Kopf 1990; Pennington 1981]. Given the abnormal structure of the heart in our patient, it could be speculated that thrombi from the venous system could migrate into pulmonary circulation and systemic circulation through common atrium and ventricle, eventually causing arterial occlusion.

Considering the unfavorable prognosis of ischemic stroke with pulmonary embolism, treatment should be started as early as possible. At present, there are no standard recommendations for anti-thrombotic therapy in patients with concomitant PE and embolic stroke [Bonnel 2015]. Management modalities including anticoagulation, thrombolytic therapy or endovascular embolectomy have been used alone or in combination previously [Gunta 2012; Omar 2013; Liu 2015]. In the immediate postoperative period, thrombolytic therapy and endovascular embolectomy were believed to be contraindicated. Therefore, systemic anticoagulation was administered and we recommended the use of it in our case [Wang 2014]. Furthermore, systemic anticoagulation is an approach with a lower risk of bleeding.

Conclusion

Our case raises concerns about the late thromboembolic complications after Glenn surgery. Currently, thromboprophylaxis

with warfarin or antiplatelet agents in the immediate postoperative period is often recommended. However, there is still insufficient evidence and no consensus on their use in routine clinical practice. Further trials are mandatory to establish the ideal approach for prophylactic treatment.

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