Case report - Congenital

Hemolytic anemia: an unusual presentation of cor triatriatum sinistrum

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1. Introduction

Cor triatriatum sinistrum is a rare congenital cardiac malformation, constituting about 0.1–0.4% of congenital heart disease [1]. It is characterized by the presence of a fibromuscular membrane that subdivides the left atrium into two chambers, a superior common pulmonary venous chamber and an inferior true left atrial cavity. The hemodynamics of cor triatriatum is similar to that of mitral stenosis. Patients usually present with signs and symptoms of heart failure. In this report, we will focus on an unusual presentation of a two-month-old child with cor triatriatum, who presented with heart failure and hemolytic anemia. To the best of our knowledge, the constellation of cor triatriatum and hemolytic anemia was not reported before.

2. Case report

A two-month-old female patient was referred to our hospital due to heart failure. The patient was full-term, product of normal spontaneous vaginal delivery with no postnatal complication. Shortly after birth, the parents noticed shortness of breath and sweating on feeding that was progressive and then she developed cough and chest infection. The patient was treated in a local hospital and referred to our hospital for further management. Clinically, the patient was distressed, respiratory rate of 60/min with intercostals and subcostal retraction, pulse 160 beats/min, blood pressure 82/51 mmHg, and oxygen saturation 99% on room air. The cardiac apex was felt in the 5th intercostal space outside the mid-clavicular line, the second heart sound was prominent and there was systolic murmur grade 3/6 at the left lower sternal border. The liver was 3 cm below the costal margin. A chest X-ray revealed pulmonary venous congestion. Preoperative investigations showed that the patient had normochromic normocytic anemia with high serum bilirubin level. The patient had hemoglobin of 7.2 g/dl, mean corpuscular volume of 77 femtoliter, mean corpuscular hemoglobin of 24 picogram, mean corpuscular hemoglobin concentration of 34%, and reticulocyte count of 13.2%. The patient’s lactate dehydrogenase was 2295 IU/l, her Coombs test was negative, and her renal function was normal. Hemoglobin A2 was 88%, hemoglobin A3 was 2.3% and hemoglobin F was 11.2%. Peripheral blood smear examined by a consultant hematopathologist showed normochromic normocytic red blood cells. The blood group of the patient and her mother was the same (A+). With these laboratory findings, we concluded that her anemia was due to the intravascular hemolysis. Transthoracic ECHO revealed situs solitus, levocardia, normal systemic venous connections; all pulmonary veins were clearly seen draining to the left atrium with a patent foramen ovale and intact interventricular septum. There was a fibromuscular membrane dividing the left atrium to a superior chamber to which all pulmonary veins were drained into. The membrane had a small defect through which pulmonary venous drainage was passed from the superior chamber to a smaller inferior one with turbulent flow and a mean pressure of 25 mmHg and a peak pressure gradient of 36 mmHg across the defect.

The patient developed severe respiratory distress and required an urgent surgical operation through a median sternotomy. Cardiopulmonary bypass was initiated after aortic and bicaval venous cannulation. Aortic cross-clamp...
was applied and the heart was arrested with cold blood cardioplegia, and mild systemic hypothermia was used throughout the procedure. The atrial septum was determined and the cor triatriatum was reached via transseptal approach. The membrane was successfully excised around the circumference of the atrial septum and left atrial free wall. Care was taken to avoid injury to the mitral valve apparatus and atrial free wall.

The patient had an uneventful postoperative course. Postoperatively, her hemoglobin returned to normal, her lactate dehydrogenase gradually approached the normal range, the reticulocyte count was normalized. Postoperative echocardiography revealed no residual gradient in the left atrium. The patient was discharged in good condition on postoperative day 5. Follow-up shows still normal hemoglobin of 14.5 g/dl.

3. Comment

In patients with stenotic lesions, the mechanism for intravascular hemolysis is considered to be related to turbulence and shear stress produced by flow through stenosed or incompetent orifices. In the adult, red cell shearing stress above 3000 dynes/cm² can increase hemolysis [2, 3]. There is a report of a 50-mmHg peak gradient through left ventricular outflow tract or aortic valve calculated to have a shear stress of 4000 dynes/cm² [4]. Our patient is an infant and had a maximal peak gradient of 35 mmHg with echocardiography. This high gradient in an infant may be enough to cause hemolysis. We determined that cor triatriatum might cause hemolytic anemia in our case, as was evidenced by the laboratory findings and clinical presentation. In our case, the stenotic orifice of the cor triatriatum caused hemolysis which was surgically fully corrected.

References