VIA MEDICA

LETTER TO EDITOR

Anaesthesiology Intensive Therapy
2017, vol. 49, no 4, 320–322
ISSN 1642–5758
10.5603/AIT.a2017.0048
www.ait.viamedica.pl

Pathological drainage of the right superior vena cava into the left atrium diagnosed in a 37-year-old patient in postpartum period: a case report

Katarzyna Krysiak-Neneman, Karina Stefańska-Wronka, Jacek Galicki, Jolanta Miszczak-Śmiałek, Łukasz Matuszewski

Department of Anesthesiology and Intensive Therapy, Voivodeship Hospital in Poznań, Poland

Sir,

We would like to report the case of a 37-year-old patient with an undiagnosed heart defect, admitted to the Intensive Care Unit (ICU) with acute respiratory failure and suspicion of pulmonary embolism (PE) after a caesarean section performed one day earlier, in the 34th week of her 4th pregnancy. Eventually, the diagnosis of isolated drainage of the right superior vena cava (RSVC) to the left atrium (LA) was made.

Our patient had a history of two caesarean sections, one miscarriage, gestational diabetes, pregnancy-induced hypertension, obesity and erythrocythaemia. Initially, the patient was sent to the Obstetrics Department by her general practitioner because of a deterioration in her blood pressure (BP) control. On admission, the patient additionally presented upper respiratory tract infection (URTI) symptoms (voice hoarseness for a week and 2-day history of low-grade fever), dyspnoea on effort and peripheral oedema (ankles and feet). BP on admission was 160/100 mm Hg, Hb 17.1 g dL^{-1}, Ht 50%, RBC 5.4 T L^{-1}, PLT 108 G L^{-1}, WBC 11.9 G L^{-1}, fibrinogen 2.6 g L^{-1}. Urea, creatinine, Na, K, aPPT, PT, AST, ALT, ALP, uric acid and bilirubin levels were within normal ranges. The results of arterial blood gas analysis (ABG) were as follows: paO_2 52 mm Hg, $paCO_2$ 30 mm Hg, sO_2 87%, BE –3.1 mmol L^{-1}. Other parameters were normal. In urinanalysis, bacteriuria, significant leucocyturia and proteinuria (9.68 g L^{-1}) were discovered. A chest ultrasound showed nothing significant. During a spirometry, only PEF was below the normal value. The patient was treated with cefuroxime, betamethasone (to stimulate foetal respiratory system maturation), magnesium sulphate, diazepam, inhaled budesonide and supplemental oxygen. As after 4 days her condition had not improved, she was sent to University Gynaecology and Obstetrics Hospital with suspected preeclampsia and PE. In laboratory tests on admission, CBC and coagulogram did not change significantly, there was no troponin elevation, while procalcitonin was 0.17 ng mL^{-1}, and d-dimmer 731 ng mL^{-1} (the highest value was 1124 ng mL^{-1} — two days after admission). A chest X-ray showed opacities in right lower pulmonary region. She was treated with magnesium sulphate, methyldopa, nitrendipine, metoprolol, a subtherapeutical dose of enoxaparin and continued cefuroxime. In the 2nd day of hospitalization dyspnoea, tachypnoea, retrosternal chest pain and numbness of the upper extremities were observed. BP was 170/110 mm Hg, SpO_2 90% with supplemental oxygen, paO_2 60 mm Hg, $paCO_2$ 33 mm Hg. Markers of myocardial necrosis were at normal ranges. Although the next day her symptoms had improved, lower abdominal pain and vaginal bleeding occurred. An urgent caesarean delivery under general anaesthesia was performed. Intraoperatively, abruption of 25% of the placental surface was discovered. A hypotrophic female infant with an Apgar score of 8 in the 1st minute and 10 in the 5th minute was delivered. Evaluated blood loss was 500 mL. Although after the surgery the patient was extubated, symptoms of respiratory failure developed. After 4 hours she required endotracheal intubation and mechanical ventilation. Despite ventilation with 100% oxygen, SpO_2 was 89–92% and paO_2 was 60 mm Hg (other parameters in ABG were normal). Oliguria and elevation of creatinine by 0.4 mg dL^{-1} were observed, while CBC was as follows: haemoglobin 11.9 g dL^{-1}; haematocrit 35%; RBC 3.74 T L^{-1}; PLT 206 G L^{-1}; WBC 19 G L^{-1}. The patient required a dobutamine infusion and was transferred to the ICU of our hospital. On admission the patient was sedated and her lungs were mechanically ventilated with FiO_2 0.5. SpO_2 was 86% and HR 90 min^{-1}. She required a noradrenaline and dobutamine infusion. The results of laboratory tests were as follows: WBC 15.7 G L^{-1}; RBC 3.79 T L^{-1}; Hb 12.3 g dL^{-1}; Ht 36.5%; PLT 200 G L^{-1}; creatinine 1.04 mg dL^{-1}; urea 46 mg dL^{-1}; C-reactive protein (CRP) 84.1 mg L^{-1}; procalcitonin 1 ng mL^{-1}; and paO_2 53.8 mm Hg. Moreover, PT, aPTT, d-dimer, troponin, Na, K and antithrombin III were normal. She had a CT scan of the head, the abdomen, the pelvis and a CT pulmonary angiogram. Contrast was administered via a central venous catheter (CVC) inserted through right subclavian vein. Discovered abnormalities were small postischaemic lesions in the subcortical nuclei of the brain, the presence of fluid in the pleural cavities (up to 10 mm), opacities in the lower pulmonary regions, dilation of the ascending aorta to 45 mm and features of gross heart defect with right-to-left shunt at the level of atria. There was no enlargement of the right heart and pulmonary trunk or arteries. Pulmonary arteries could not be assessed for PE. A transthoracic echocardiography showed no significant abnormalities — only left ventricle hypertrophy and insignificant hydropericardium. There were no features of a septal defect, any other heart defect or right ventricular strain. A transoesophageal echocardiography also did not explain our patient's condition. As there was no proof for PE, an intermediate dose of low-molecular-weight heparin was administered. In the succeeding days, the patient underwent broad bacteriological and virological diagnostics

(cultures of blood, tracheal aspirates and urine, tests for influenza, cytomegalovirus [CMV], Ebstein-Barr Virus [EBV], *Mycoplasma pneumoniae*, *Chlamydiophila pneumoniae*). Broad-spectrum intravenous antibiotics and antiviral therapy were started, namely: piperacillin with tazobactam; ciprofloxacin; oseltamivir (until negative influenza test results); acyclovir (switched to gancyclovir after a positive test result for CMV-DNA in blood plasma). Additionally, intravenous fluconazole was started after identification of *Candida albicans* in tracheal aspirate. The highest value of CRP was 192 mg L^{-1} (in the 4th day of treatment). This decreased to 14 mg L^{-1} in the 9th day. Procalcithonin was 1.7 ng mL^{-1} in the 2nd day but only 0.4 in the 4th day. Low-grade fever was observed. The maximum doses of catecholamines that the patient received were 0.13 μg kg^{-1} min^{-1} of noradrenaline and 8 μg kg^{-1} min^{-1} of dobutamine. Amines were continued until the 8th day of treatment. Arterial hypertension was observed. Despite high doses of midazolam (6 mg h^{-1}) and fentanil (300 μg h^{-1}), the patient's Ramsey Sedation Scale score was 1 or 2. After short period of oliguria, kidney function improved. The last, but not least remaining problem was isolated respiratory failure, which was mechanically ventilated with FiO_2 45%, while her PaO_2 values were around 50–60 mm Hg. Despite low levels of CMV-DNA in blood plasma (485 copies mL^{-1}) and a lack of characteristic radiographic features, there was no other perceptible reason for isolated hypoxaemia — the diagnosis of CMV pneumonia was made. Searching for evidence for immunoincompetence, levels of immnoglobulins were measured, and the patient was tested for HCV, HIV, HBV viruses, adrenal insufficiency and hypothyroidism. Although HCV-RNA was found in the blood plasma, there were no features of hepatitis. In addition, a decreased level of cortisol was detected. Hydrocortisone supplementation was introduced. Despite the decrease of inflammatory markers levels, there was no improvement in respiratory system function. A decision was made to repeat a CT of the chest with image acquisition after 20 and 50 seconds following contrast administration. Contrast was injected into a CVC inserted through the right subclavian vein. In the early phase, when contrasting of vena cava superior occurred, contrasting of the left heart was observed at once. The image was suggestive of drainage of the superior vena cava to the left atrium. A transoesophageal echocardiography with contrasting substance (0.9% NaCl injected under pressure to CVC) was performed. A stream of solution flowing from the superior vena cava to the left atrium was observed. Despite there being apparently no justification for blood transfusion (Hb value 9.1 g dL^{-1}, Ht 27.8%), two units of red cell concentrate were administered. This was justified by erythrocythaemia having been observed before the C-section. On the same day the patient was extubated. As anticipated, PaO_2 values did not fall significantly, despite a considerable decrease in FiO_2. Within a few days, the patient was discharged from the ICU in a good general condition and transferred to the hospital where the caesarean section had been performed. She was advised to contact University Department of Cardiology after 2–3 months.

Pathological drainage of the RSVC to the LA is a very rare heart defect. In 1975, De Leval *et al.* [1] reported only 28 cases of such disease among 5,099 other heart defects while only 4 patients presented with an isolated defect. According to the best of our knowledge, at the present time only 21 cases of isolated RSVC to LA drainage have been reported. It is far more often encountered in conjunction with other heart defects. In the majority of cases, reports were on patients that presented with cyanosis, clubbed fingers, dyspnoea, brain abscesses or emboli in well-vascularized organs [2, 3]. We found only 3 case reports on asymptomatic patients. One of them was a patient in postpartum period. In this case, reported by Baggett *et al.* in 2009 [4], symptoms of the heart defect had also become apparent after an urgent caesarean section under general anaesthesia. The patient was also suspected of PE. In this case, however, it was already suggested after CT pulmonary angiogram that there was pathological drainage of RSVC to LA. It is difficult to find information explaining why some patients remain asymptomatic. Explanation for the ability of the patients to function for a long time with symptoms but without correction of the heart defect arise from the fact that most of the cardiac output comes back via the inferior vena cava to the right atrium and becomes oxygenated. Rosenkranz *et al.* [5] estimated that, at rest, the right-to-left shunt arising from pathological drainage of RSVC is around 15% of cardiac output. It is hard to tell what precisely decompensated the cardiovascular function of the patient discussed here. Before the caesarean section, it was probably worsening PIH and URTI. After the surgery, these were, additionally, a decrease in Ht, respiratory dysfunction arising from positive pressure ventilation during general anaesthesia and perhaps pneumonia, which had been recognised as the cause of respiratory failure before the real reason became clear. Regardless of the explanation, the above-discussed case should be a cautionary tale. After making the right diagnosis, all pieces of the jigsaw fell into place. However, it took 9 days to put this puzzle together. The patient received antibiotics and antiviral drugs that probably could have been avoided. Short before making the right diagnosis, indications for performing a tracheostomy started to be analysed as there was no short term perspective for extubation. The authors admit that they displayed too little perseverance in analysing the reason for a lack of contrasting of the right heart and pulmonary arteries in the CT pulmonary angiogram performed on the day of

admittance. Despite the apparently negative TTE and TEE images, in the context of any heart defect, the diagnostics in that direction should have been extended much earlier. We are justified only by the fact that discussed defect is very rare and additional difficulties in making the diagnosis are caused by misleading results of echocardiography, which may show no pathology when performed without contrast administration. The previously asymptomatic course of the disease in the above-discussed patient was even more deceptive.

Some time after the patient had been discharged from the University Gynaecology and Obstetrics Hospital, she was admitted to the Cardiology Department where a heart MRI was performed and in which the previous diagnosis was confirmed. Additionally, minor partial pathological drainage of the pulmonary veins to the superior vena cava was discovered (three small pulmonary venous branches draining into the superior vena cava).

ACKNOWLEDGEMENTS

1. Source of funding: none.
2. Conflict of interest: none.

References:

1. de Leval MR, Ritter DG, McGoon DC, et al. Anomalous systemic venous connection. Surgical considerations. Mayo Clin Proc. 1975; 50(10): 599–610, indexed in Pubmed: 1165650.
2. Ezekowitz MD, Alderson PO, Bulkley BH, et al. Isolated drainage of the superior vena cava into the left atrium in a 52-year-old man: a rare congenital malformation in the adult presenting with cyanosis, polycythemia, and an unsuccessful lung scan. Circulation. 1978; 58(4): 751–756, indexed in Pubmed: 688585.
3. Usalp S. Right superior vena cava draining into the left atrium. Clinical Medical Reviews and Case Reports. 2016; 3(10), doi: 10.23937/2378-3656/1410134.
4. Baggett C, Skeen SJ, Gantt DS, et al. Isolated right superior vena cava drainage into the left atrium diagnosed noninvasively in the peripartum period. Tex Heart Inst J. 2009; 36(6): 611–614, indexed in Pubmed: 20069093.
5. Rosenkranz S, Stäblein A, Deutsch HJ, et al. Anomalous drainage of the right superior vena cava into the left atrium in a 61-year-old woman. Int J Cardiol. 1998; 64(3): 285–291, indexed in Pubmed: 9672410.
6. Al-Biltagi MA, Kouatli A, Al-Mousily F. Right superior vena cava draining in the left atrium associated with tetralogy of Fallot and pulmonary atresia. Ann Pediatr Cardiol. 2013; 6(1): 65–67, doi: 10.4103/0974-2069.107237, indexed in Pubmed: 23626439.

Corresponding author:
Katarzyna Krysiak-Neneman
Department of Anesthesiology and Intensive Therapy,
Voivodeship Hospital in Poznań, Poland
e-mail: ait-lutycka@wp.pl