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

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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 erythrocytasia. 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, aPTT, PT, AST, ALT, ALP, uric acid and bilirubin levels were within normal ranges. The results of arterial blood gas analysis (ABG) were as follows: paO₂ 52 mm Hg, paCO₂ 30 mm Hg, sO₂ 87%, BE –3.1 mmol L⁻¹. Other parameters were normal. In urinalysis, bacteriuria, significant leucocyturia and proteinuria (9.68 g L⁻¹) were discovered. A chest ultrasound in urinanalysis, bacteriuria, significant leucocyturia and proteinuria (9.68 g L⁻¹) were discovered. A chest ultrasound

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(cultures of blood, tracheal aspirates and urine, tests for influenza, cytomegalovirus (CMV), Epstein-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. Procalcitonin 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 midaflom along that the patient received were 0.13 mg 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 immunoinsufficiency, levels of immunoglobulins 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 contrast of vena cava superior occurred, contrast 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 to LA 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).

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References:

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