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Pulmonary embolism presenting as disseminated intravascular coagulation

因散佈的血管內凝結物而出現的肺部栓塞

We report an unusual case of disseminated intravascular coagulation. Occult pulmonary embolism is a recognised cause of disseminated intravascular coagulation. Unexplained shock should prompt the physician to search for a thrombotic cause such as pulmonary thromboembolism.

我們報告了一宗不尋常的散佈血管內凝結物病例。潛隱性的肺部栓塞被認為起因於散佈的血管內凝結物。對原因不明的休克，醫生應去查找諸如肺部栓塞之類的血栓形成原因。

Case report

An 81-year-old woman with a history of rheumatic heart disease, atrial fibrillation, gouty arthritis, and chronic renal insufficiency (creatinine level 278 mmol/L [normal range, 53-106 mmol/L]), was admitted with a 4-day history of fever and malaise. Physical examination revealed mitral regurgitation (MR), tricuspid regurgitation (TR) with giant V wave, pyrexia of 38°C, and blood pressure of 140/70 mm Hg. The patient's haemoglobin level was 90 g/L (normal range, 120-150 g/L), with a white cell count (WCC) of 11 x 10^9/L (normal range, 4.5-11.0 x 10^9/L), platelet count 147 x 10^9/L (normal range, 150-450 x 10^9/L), and erythrocyte sedimentation rate (ESR) 84 mm/h (normal range, 0-20 mm/h). Blood culture grew *Staphylococcus aureus*. There was no intravenous catheter in situ at the time of admission. Transthoracic echocardiogram revealed a mass in the right atrium, dilated right and left atria, moderate MR and TR, and no vegetations. Transoesophageal echocardiogram showed an immobile mass of 2.7 x 4.6 cm in size, pedunculated and attached to the posterior wall of the right atrium; the tricuspid valve was mildly fragmented. A diagnosis of probable right atrial myxoma was made. In view of her frail condition, the myxoma was treated conservatively. She was treated for *S aureus* right-sided endocarditis with intravenous cloxacillin for 4 weeks. Erythrocyte sedimentation rate decreased to 35 mm/h after antibiotic treatment. Repeated blood culture was negative and the patient remained well. In view of excessive fall risk in this frail woman, together with other co-morbidities and poor drug supervision at home, a clinical decision was made that anticoagulation was not an appropriate treatment for her atrial fibrillation.

Two months later, the patient was admitted with a 2-day history of dizziness. On clinical examination she was dehydrated and afebrile, with a blood pressure of 88/44 mm Hg; MR and TR were present as before. Auscultation of the chest was clear, and there was no ankle oedema. Rectal examination was normal. The patient's haemoglobin level was 61 g/L, with WCC 4.7 x 10^9/L, platelet count 61 x 10^9/L, and ESR 22 mm/h. Blood film showed schistocytes and reticulocytes (1.1%; normal range, 0.5-1.5%). Prothrombin time (PT) was 26.8 s (normal range, 10-13 s) and activated partial thromboplastin time (APTT) was 74.8 s (normal range, 25-40 s). D-dimer levels were elevated above 0.5 mg/mL (normal level, <0.5 mg/mL), and fibrinogen was decreased to 0.5 g/L (normal range, 1.5-3.5 g/L). Arterial blood gas measurement showed pH 7.34 (normal range, 7.35-7.45), PO_2 59 mm Hg (normal range, 80-100 mm Hg), PCO_2 24 mm Hg (normal range, 35-45 mm Hg), and bicarbonate level 13.5 mmol/L (normal range, 21-28 mmol/L). Oliguria was present, with urea level of 16.7 mmol/L (normal

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range, 2.9-8.2 mmol/L), creatinine level of 277 mmol/L, and a mildly elevated anion gap of 18.4 mmol/L (normal range, 8-16 mmol/L). Spot glucose was 4.5 mmol/L, and morning cortisol level 544 nmol/L (normal range, 110-520 nmol/L). No organism was isolated from blood, urine, and sputum cultures. Chest X-ray showed cardiomegaly with clear lung fields. An electrocardiogram showed atrial fibrillation of 100 beats/min—this was unchanged from that performed 2 months previously, with no regional hypokinesia or pericardial effusion detected.

The patient was transferred to the high dependency unit for further management. Intravenous fluids, blood transfusion, and ceftazidime were administered, with a clinical diagnosis of septic shock complicated by disseminated intravascular coagulation (DIC). Blood pressure and urine output improved initially. On the fourth day, blood was oozing from drip sites; expiratory rhonchi were heard in the chest, with central cyanosis and acrocyanosis. Repeat chest X-ray was unremarkable. Hydrocortisone, salbutamol nebulising solution, and oxygen (5 L) were added. Pulmonary arterial and central venous catheterization were not attempted because of the risk of dislodging the right atrial mass. A further impaired clotting profile was found, with PT >60 s, APTT >109.4 s, platelet count 20 x 10^9/L, and fibrinogen level 0.3 g/L. The patient continued to deteriorate, with oliguria and hypotension unresponsive to dobutamine and dopamine. She died on day 8 after admission. At autopsy, a 1 cm long, 0.3 cm thick, large circumferential pulmonary thrombus occluding 50% of the left pulmonary artery was found. The right atrial mass was a thrombus (4 cm x 3 cm) with pustular material inside. Culture was negative. No thrombi were seen in other organs or leg veins. The tricuspid valve was fibrotic, and culture was negative.

Discussion

When a patient presents with DIC and shock, the most appropriate treatment is to restore blood pressure and urine output by fluid replacement, and treat the underlying cause.

The use of anticoagulants in permanent atrial fibrillation is associated with a reduction in the relative risk of stroke of 62% versus placebo. In primary prevention, the number of patients who need to be treated with warfarin for 1 year to prevent one stroke is 37.[1] In this frail elderly patient with a propensity to fall, and the risk of development of an intracranial haemorrhage,[2] anticoagulation was not commenced. The patient in this study died of a large pulmonary embolism (PE). The right atrial thrombus was clearly the source of the PE, since no clot was found in the peripheral venous system. The atrial thrombus was possibly acting as a septic focus, accounting for *S aureus* bacteraemia in the previous admission, although no organism was isolated from the thrombus.

Pulmonary embolism is a recognised cause of DIC. However, it is not well documented in modern textbooks. It is postulated that DIC is initiated by the release of thrombin from the impacted clot in the pulmonary artery.[3] Release of thromboplastin from infarcted tissue, and endothelial damage, also contribute to the coagulation process. During clot formation, serotonin is released from platelets into the plasma. Serotonin has been shown to cause spasm of the pulmonary vessels and dyspnoeic attacks.[4] This may explain the expiratory rhonchi detected in the patient during the clinical course. Disseminated intravascular coagulation accounts in part for the hypotension observed clinically. The endothelial damage activates clotting factor XII (Hageman factor), and subsequently the intrinsic clotting system.[5] Factor XII directly transforms kallikreinogen into kallikrein,[6] resulting in the release of bradykinin, a potent peptide that lowers blood pressure by vasodilation.

The patient in this study is an example of PE causing rather than complicating DIC. In DIC, there is de novo thrombus formation in the pulmonary venous system, whereas a thrombus in the pulmonary arterial system may represent an embolus from the right heart and venous circulation.[7] No other cause could be found at autopsy to account for DIC. Giant atrial thrombus causing mechanical obstruction has been reported to cause DIC,[8] but the atrial thrombus in this case was relatively small in size. In a patient with unexplained DIC, occult PE should be considered. Spiral computed tomography of the thorax is able to confirm the PE. Full-dose heparin, if administered early, will prevent further DIC,[9] acting at the first step of the chain reaction in which the endogenous fibrinolytic system lyses the clot before the irreversible stage of severe DIC is reached.[10]

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