Case report - Cardiac general

The acute chest syndrome of sickle cell disease following aortic valve replacement

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Abstract

The acute chest syndrome (ACS) of sickle cell disease (SCD) is a leading cause of death in SCD, with a high incidence following surgery, though only one case has been reported following cardiac surgery. We present a case of ACS in an adult undergoing aortic valve replacement (AVR) despite instituting established peri-operative optimization measures to prevent sickling. Early diagnosis of this condition in our patient as a distinct clinical entity facilitated appropriate, specific therapy and a good subsequent postoperative recovery. Greater recognition of this syndrome in the growing number of adult sickle cell patients presenting for cardiac surgery may help improve their outcome.

Keywords: Aortic valve; Cardiopulmonary bypass; Lung pathology; Hematology; Genetics

1. Introduction

Acute chest syndrome (ACS) is an acute pulmonary disorder associated with sickle cell disease (SCD) that leads to considerable morbidity and mortality. We describe an adult SCD patient undergoing aortic valve replacement (AVR) who developed ACS and describe key diagnostic features of the syndrome as well as the management strategy that led to a good subsequent recovery.

2. Case report

A 59-year-old Afro-Caribbean lady with homozygous hemoglobin S (HbS) SCD was admitted for AVR for symptomatic moderate–severe aortic stenosis (aortic valve area 0.88 cm²; peak gradient 48 mmHg). She was an ex-smoker with a history of multiple sickle ‘crises’ and took warfarin for superior vena cava thrombosis. Biventricular function was good, estimated pulmonary artery systolic pressure was 65 mmHg and coronary arteries were unobstructed. Pulmonary function tests showed FEV₁ of 1.05 liters (1; 64% predicted) and FVC of 1.27 liters (1; 64% predicted); corrected transfer factor for carbon monoxide (TLCOc) 35% predicted. Peri-operative management followed protocols to minimize risk of sickling [1]: good hydration, oxygenation and analgesia, and a preoperative exchange transfusion which reduced her HbS fraction from 43.3% to 20.9%.

The patient underwent uneventful AVR with a 21-mm mechanical prosthesis (St Jude Inc, Minnesota, USA) and was kept at 37 °C, with diastolic arrest induced and main-tained using anterograde warm blood cardioplegia. Cardiopulmonary bypass (CPB) time was 77 min and cross-clamp time 58 min. No inotropes were required and the patient was extubated after 5 h, with a fentanyl infusion for analgesia. She made a good initial recovery with aggressive management: oxygen therapy; strict pain control, maintenance of low HbS fraction; incentive spirometry post-extubation; broad-spectrum intravenous antibiotics and bronchodilators. She deteriorated, however, on the day of discharge, postoperative day (POD) 6, with acute chest wall and abdominal pain, pyrexia, tachypnea and ensuing type II respiratory failure. White blood count (WBC) was 19.9 × 10⁹/l (baseline WBC 15.3 × 10⁹/l), and platelet count 413 × 10⁹/l. Arterial blood gas analysis showed: pH 7.2, pO₂ 10 kPa, pCO₂ 8.4 kPa, lactate 0.7 mmol/l, HCO₃⁻ 20.2 mmol/l. The WBC increased to a peak of 32.6 × 10⁹/l by POD 9. She required re-admission to the intensive care unit (ICU) and mechanical ventilation. Chest X-ray (CXR) and computed tomography (CT) scan of the chest and abdomen (Fig. 1) showed bilateral lower lobe lung consolidation with pleural and pericardial effusions. A diagnosis of ACS was made and initial postoperative respiratory measures re-instituted; the patient was also commenced on inhaled nitric oxide (NO) and then oral sildenafil. She required a tracheostomy for respiratory weaning. Sputum cultures were negative. She improved gradually over the next 14 days, and was discharged after rehabilitation.

3. Discussion

The ACS of SCD is a leading cause of death in patients with SCD [2, 3]. It is defined as the occurrence of new
pulmonary infiltrates with accompanying symptoms and signs including: fever, cough, chest wall pain, abdominal pain, dyspnea, tachypnea and chest rales [2, 4]. Homozygous HbSS patients with SCD typically have a reduced life expectancy and history of frequent sickle vaso-occlusion (V-O) ‘crises’ with severe chest, back, abdominal or limb pain [5]. ACS appears to be a V-O event that often occurs on a background of chronic lung disease (CLD) in SCD patients either spontaneously or in response to triggers such as surgical stress [3–5]. The CLD may, in turn, be manifested by hypoxia, pulmonary fibrosis or pulmonary hypertension [4].

The pathogenesis of post-surgical ACS in SCD appears to involve several initiating events, including atelectasis and ventilation–perfusion mismatch related to anesthetic agents, both of which lead to sickling of erythrocytes within the pulmonary microcirculation [2, 3, 5]. Peri-operative systemic inflammation, hypoxia, acidosis, hypovolemia and hypothermia exacerbate these factors and neutrophils are involved both in CPB-mediated inflammation as well as sickling [1, 5]. Specific therapy in sickle cell ACS targets some of these mechanisms [2]: bronchodilators, daily incen tive spirometry, aggressive respiratory support, broad-spectrum antibiotics and NO/sildenafil (reduced NO in the pulmonary endothelium of SCD patients leads to enhanced erythrocyte sequestration [5]). Bronchodilators appear to be beneficial as many patients with ACS have a component of dynamic airway hyperreactivity [2, 4] and broad-spectrum antibiotics are recommended even though a causative infectious agent is often not identified [2].

Despite its high incidence and mortality, ACS has been reported in only one case following cardiac surgery [6]. The reported incidence following abdominal surgery is up to 10% [3]. Our patient fulfilled a number of criteria for ACS [2, 4]: severe chest pain, fever, cough, hypercapnea, pleural effusions and new bilateral lower lobe infiltrates on CXR and CT-scan (Fig. 1). The differential diagnosis of ACS includes acute asthma, pulmonary thromboembolism, pneumonia, atelectasis and acute respiratory distress syndrome. Our patient had a number of risk factors for ACS [1–3]: HbSS, pulmonary hypertension, reduced FEV and TLCOC, elevated baseline WBC and trigger stimulus of surgery and CPB. She also had at least two risk factors which predict the need for ventilation in ACS: cardiac disease and multi-lobar involvement [2]. Further, the platelet count > 400 × 10⁹/l on POD 6 is a predictor of prolonged hospitalization and respiratory failure in sickle patients with ACS [2].

We suggest that improved recognition of patients at high risk of ACS, targeted therapy and a multidisciplinary approach in adult SCD patients undergoing cardiac surgery and CPB may help improve outcomes in these patients.

References