LEUKOSTASIS: AN UNUSUAL CAUSE OF DESATURATION

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INTRODUCTION

Hyperleukocytosis, defined as a leucocyte count exceeding 100 x 10^9/L, is an oncologic emergency associated with leukemias. Leukostasis is a clinicopathological syndrome caused by sludging of circulating leukemic blasts in the tissue microvasculature. We report a pediatric case with relapsed acute myeloid lymphoma (AML) scheduled for elective Hickman line insertion who had persistent desaturation after anesthesia induction as a result of presumptive pulmonary leukostasis.

CASE REPORT

A 21 month old boy with relapsed AML presented for an elective Hickman line insertion under general anesthesia (GA) for the initiation of chemotherapy. He was first diagnosed with AML subtype M5 six months prior and was deemed to be in remission after completing 4 cycles of chemotherapy. A month later, he was admitted to our center with a relapse. Preoperatively, he maintained saturations of 97-100% on room air; chest X-ray and transthoracic echocardiography (TTE) were unremarkable. Of note, his leukocyte count was 116 x10^9/L. Two days prior, he underwent a bronchoalveolar lavage under GA uneventfully.

During the procedure, the patient desaturated immediately after intravenous induction. He was intubated and ventilated with inspired oxygen (FiO2) of 1.0. Lung auscultation and intra-procedural chest x-ray were unremarkable. Blood pressure post-induction was 70/30mmHg. Despite conventional corrective measures, his saturations hovered around 88-90%. He was extubated after four days of uneventful stay in the ICU and transferred to the general ward subsequently.

At the CICU, the presumptive diagnosis was hyperleukocytosis with pulmonary leukostasis. The patient was started on hyper-hydration therapy and chemotherapy. A repeated TTE showed improvements in pulmonary pressures. After starting chemotherapy, his ventilatory requirements decreased in parallel with the leukocyte count. He was extubated after 20 minutes of lung recruitment. His saturation improved to 95% and blood pressure was 85/35mmHg. However, efforts to wean FiO2 below 0.6 were unsuccessful. The line insertion proceeded in view of need to initiate chemotherapy and the patient was transferred to the children’s ICU (CICU) for ventilatory support.

DISCUSSION

Hyperleukocytosis is an oncologic emergency and if left untreated, can cause mortality in 40% of patients1. It forms a poor prognostic marker and increases risks of severe complications arising from hyperviscosity of the blood. Leukostasis gives rise to symptomatic hyperleukocytosis, usually involving the pulmonary and cerebral microvasculature. The occlusion of small blood vessels arises from abnormally high leukocyte count and their decreased deformity, leading to clumping and stasis from sluggish flow. In addition, formation of microthrombi, release of toxic granules from endothelial damage and massive oxygen consumption by the leukocytes all contribute to the pathogenesis of leukostasis.

Pulmonary symptoms include dyspnea, tachypnea, hypoxia, pulmonary infiltrates, and overt respiratory failure. Neurological symptoms involve dizziness, delirium, papilloedema, retinal hemorrhage, coma and intracranial hemorrhage. Other manifestations involve the heart, testes and peripheral circulation.

A retrospective study by Westmead Hospital in 2009 showed mortality and morbidity rates of 3.8% and 32.7% respectively in pediatric patients with hyperleukocytosis undergoing anesthesia2; contrasting with nonfatal perioperative morbidity rates of 2.9-3.1% in the general pediatric group (excluding open-heart surgery and neurosurgery)3,4.

In our patient, it was likely that there was pre-existing ventilation-perfusion (VQ) mismatch resulting from sluggish pulmonary blood flow due to hyperleukocytosis prior to the anesthesia. Post-induction vasodilatation of the peripheral vasculature resulted in systemic hypotension, reducing preload to the right heart. This further compromised pulmonary blood flow, worsened VQ mismatch and contributed to the desaturation. In addition, the high metabolic demands of the leukocyte load increased oxygen consumption. Pulmonary vascular resistance had increased due to the sluggish pulmonary flow and preexisting hypoxemia, hence explaining his echocardiography findings.

Lung recruitment and fluid resuscitation improved his blood pressures and saturation marginally. Institution of chemotherapy, which resulted in a downward trend of his leukocyte count, gave rise to the most significant improvement in oxygenation.

Leukocytoreduction either via chemotherapy or leukapheresis remains the mainstay of management for leukostasis. Leukapheresis acutely reduces tumor load and improves symptoms, but its long-term benefit is lacking and its precise role in pediatric practice is not clearly defined5. Other supportive measures include hyperhydration, tumor lysis prophylaxis with allopurinol and the administration of Rasturibase, a recombinant urate oxidase. Red cell transfusions should be used cautiously, if at all, to minimize increasing the blood viscosity and hence risks of adverse outcomes.

CONCLUSION

This case highlighted an unusual cause of post-intubation desaturation and offered a differential diagnosis to pulmonary embolism in leukemic patients. It served to alert practitioners to the risks of hyperleukocytosis which can be compounded by the effects of general anesthesia.

REFERENCES


Day Procedure

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<th>Day</th>
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<th>At CICU</th>
<th>At CICU Day 1</th>
<th>At CICU Day 2/Post-chemo</th>
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<td>60.6</td>
<td>50.3</td>
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</table>

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