CASE REPORT

Anesthetic Management of a Child with Hyper-IgE Syndrome for Pneumatocele Excision: A Case Report

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ABSTRACT

We describe a child of hyperimmunoglobulin-E syndrome (HIES) with bilateral pneumatoceles for an elective right thoracotomy for pneumatocele excision. HIES is a rare primary immunodeficiency characterized by the triad of markedly elevated serum levels of immunoglobulin E (>2000 IU/mL) with peripheral eosinophilia, recurrent staphylococcal infections of the skin and lungs, and chronic eczema. A 10-year-old male child was presented with recurrent skin abscesses and lung infections, leading to extensive bilateral pneumatocele formation. Such children present unique anesthetic challenges in terms of immunodeficiency, multiple hospital admissions, difficult intravenous (IV) access, and poor pulmonary compliance affecting intraoperative ventilation. Along with pediatric lung isolation techniques, thorough asepsis, good perioperative analgesia and high index of suspicion for pneumothorax are essential components of successful anesthetic management.

Keywords: Anesthesia, Hyperimmunoglobulin-E syndrome, Pneumatocele, Thoracotomy.

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INTRODUCTION

Hyperimmunoglobulin-E syndrome (HIES, Job’s syndrome) is a rare primary immunodeficiency first described by Davis et al.1 It is a multisystem disorder caused by genetic mutations.2–4 The incidence of HIES is <1:10,000,000 population.5–6 It presents as persistent cutaneous abscesses, recurring pneumonias, pneumatoceles, hypereosinophilia, craniofacial and bone growth changes, and elevated serum IgE levels.7–9 The objective of this report is to present a case of a rare multisystem disease and its anesthetic management in a patient undergoing right thoracotomy for a recurrent pneumatocele excision.

CASE DESCRIPTION

A 10-year-old male child diagnosed with HIES, (weight: 20 kg; height: 120 cm) with recurrent bilateral pneumatoceles was presented for an elective right thoracotomy for pneumatocele excision. The child was presented with high-grade fever and chronic productive cough. He had nine prior hospitalizations for infections, involving the skin, lung, and liver. Since birth, multiple times the drainage of cold abscesses from the neck, thigh, and face was done under antibiotic cover. He had a past history of empyema (three times) requiring chest drains. At 4 years of age, he was diagnosed to have HIES (IgE levels of >50,800 IU/mL, normal: 3–423 IU/mL). IgA, IgM, and IgG levels were normal. Following recurrent staphylococcal lung infections, he developed bilateral pneumatoceles. At nine years of age, he developed right sided spontaneous pneumothorax requiring emergency decompression with chest tube, liver abscess, and infective endocarditis. He had a prior history of right lower lobe pneumatocele excision under general anesthesia 4 months back with delayed extubation after 4 hours owing to a poor chest condition.

Physical examination revealed coarse facial features with flat nasal bridge, persistence of primary dentition, healed scars of chronic eczema, and grade II clubbing. He had difficult peripheral venous access. Child was normotensive with a pulse rate of 84/minute, respiratory rate of 20/minute, breath holding time of 15 seconds, SpO2 on room air 95% and systolic murmur in the tricuspid area without clinical evidence of pulmonary hypertension. He had decreased breath sounds in bilateral bases, and rhonchi with coarse crepitations (right > left side).

Investigations showed Hb 16.6 g/dL, white cell count 4700/mm3, high absolute eosinophil count 2920/mm3, normal platelets, normal liver, and renal function tests. Chest X-ray showed bilateral multiple pneumatoceles. High-resolution contrast CT chest demonstrated a large pneumatocele (8.9 × 7.4 × 10.6 cm) involving right middle and lower lobes extending along the anterior end of 2nd to 6th rib. An air-filled bulla (5.2 × 6.2 × 5.1 cm) involved posterior and lateral basal segments of the right lower lobe. The left lung parenchyma showed multiple thick-walled air-filled cavities measuring 3.7 × 2.8 × 2.4 cm (Figs 1 and 2). Fibronodular changes were also seen.

Pulmonary function test (PFT) revealed moderate restriction with small airway obstruction with forced vital capacity (FVC) 65% predicted, forced expiratory volume in 1 second (FEV1) 62% predicted, FEV1/FVC ratio 99% predicted, and peak expiratory flow rate (PEFR) 61% of predicted. Recent echocardiogram showed severe tricuspid regurgitation without pulmonary hypertension.

The child was on regular nebulization with ipratropium, salbutamol, and sulfamethoxazole–trimethoprim prophylaxis for optimization of the chest condition. Preoperatively, patient’s respiratory functions were optimized as far as possible. However, due to the presence of bilateral pneumatoceles occupying more...
than 50% of lung parenchyma, a further improvement in his respiratory parameters was unlikely.

Written informed consent (ASA III physical status) was obtained from parents on explaining the risk of intraoperative pneumothorax and postoperative ventilatory support. 6 hours of starvation to solids was confirmed. Infective endocarditis prophylaxis (ceftriaxone 50 mg/kg IV) was given. In the operation room, cardioscope, noninvasive blood pressure (NIBP), and pulse oximeter (SpO$_2$) were applied. Before induction of anesthesia, a surgical trolley with chest tube was kept ready, in order to manage an inadvertent pneumothorax. Injection of glycopyrolate (0.004 mg/kg), midazolam (0.05 mg/kg) and fentanyl (2 μg/kg) was given IV after monitoring baseline parameters.

Anesthesia was induced with propofol (50 mg) and sevoflurane in oxygen:air (50:50). The airway was secured with 26 Fr left-sided double lumen tube (DLT) after adequate muscle relaxation with vecuronium (2 mg). Lung isolation was confirmed by auscultation after alternate clamping of tracheal and bronchial lumens. Left radial arterial and right internal jugular venous canulation was done for invasive monitoring and fluid management. Muscle relaxation was maintained with vecuronium infusion (0.06 mg/kg/hour). Careful positioning of child was done perioperatively, with padding all pressure points. Anesthesia was maintained with oxygen-air (FiO$_2$ of 0.5) and sevoflurane (MAC 1–1.3) with volume-controlled ventilation (tidal volume 6–7 mL/kg, respiratory rate 30/minute). Double lung ventilation with intermittent single lung ventilation of dependant lung as and when required was done while maintaining peak airway pressures below 18–20 cm of H$_2$O. A stethoscope was placed under the left hemithorax for detection of pneumothorax. Intraoperative SpO$_2$ ranged from 91 to 98% during one lung ventilation, which was managed by increasing FiO$_2$ to 80%. End tidal CO$_2$ ranged from 35 to 45 mm Hg. Analgesia was maintained by intermittent intravenous fentanyl (total 6 μg/kg), 400 mg of paracetamol (infusion), and skin infiltration (10 mL of 0.125% bupivacaine). Intraoperatively, there was a sudden loss of tidal volume due to communication of right lower lobe pneumatocele with the lower lobe bronchus, which was managed by selective left lung ventilation at that time and surgical suturing. While excising the right upper lobe pneumatocele, the patient had few arrhythmias due to a close dissection near pericardium, which was managed conservatively. The chest wall was closed after insertion of the chest tube without air leak. The duration of the surgery was 4 hours with blood loss of 100 mL. After reversal of residual neuromuscular blockade, the child was extubated. Post-extubation arterial blood gas analysis showed a pH of 7.34, pO$_2$ of 167 mm Hg, pCO$_2$ of 47 mm Hg, HCO$_3^-$ of 25 mEq L$^{-1}$, and SaO$_2$ of 99.4%.

In the high-dependency unit, in a propped-up position, oxygen (5 L/minute) was administered with Hudson’s mask. Postoperative analgesia was maintained with IV tramadol 20 mg 6 hourly and IV paracetamol 300 mg 8 hourly. The patient recovered without any complications and was discharged on the eighth postoperative day.

**Discussion**

Hyperimmunoglobulin-E syndrome is a multisystem involvement of the immune system, skin and soft tissue, pulmonary, musculoskeletal, dental, cranial, and vascular systems. The National institutes of health (NIH) clinical scoring system is an important evaluation system; scores ≥40 suggest HIES. These patients may have repeated hospitalizations with multiple anesthesia exposures for various procedures such as drainage of abscess/empyema, bronchoscopies, lung resections, and scoliosis correction surgery. Perioperative assessment should focus on the severity of multisystem involvement.

Patients with HIES have distinct facial features such as prominent forehead with deep-set eyes, broad nasal bridge, a bulbous nose, and abnormal dentition. Dermatological manifestations include recurrent staphylococcal abscesses, eczematous rash, and mucocutaneous candidiasis. Skeletal anomalies include osteopenia, hyperextensibility, degenerative
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joint disease, pathological fractures, discrepancy in limb length, and scoliosis. This demands careful positioning of patient under anesthesia.

Pulmonary involvement includes recurrent abscesses and pneumonia, pneumatocele, bronchiectasis, empyema, spontaneous pneumothorax, and bronchopleural fistula. Incidence of post-infectious pneumatocele formation can be up to 80% in these patients.

Children presenting with recurrent pneumonias may have mild to severe respiratory distress. Preoperative assessment of respiratory function is very important to predict the postoperative recovery. This should include detailed history, clinical examination, chest X-ray, CT scan, arterial blood gases, and PFT to determine the cardiorespiratory reserve. PFTs are frequently abnormal because of a restrictive defect and difficult to perform in young children. The increased pressures in spirometry may increase the risk of rupture of pneumatocele. This child had pulmonary, cardiac, cutaneous, and dental involvement.

Indications for surgical intervention for pneumatocele are involvement of more than 50% of hemithorax with severe atelectasis, development of tension pneumatoceles leading to pneumothorax, a secondary infection of the pneumatocele, and cardiovascular compromise. Our patient had all these indications except cardiovascular compromise with recurrence of pneumatocele further complicated by immunodeficiency.

Induction of anesthesia is very challenging in cases of bilateral large pneumatoceles. Endotracheal intubation with IPPV in patients with tension pneumatoceles is a high-risk procedure because patients can develop pneumothorax progressing to hemodynamic instability. If unrecognized and untreated, this can result in respiratory failure and even cardiac arrest. Nitrous oxide should be avoided because of risk of pneumothorax following the rupture of pneumatoceles. In our case, the risk of contralateral pneumothorax was very high, hence vigilant monitoring of hemodynamic (IBP, CVP) and respiratory parameters (SpO2, ETCO2, airway pressures, stethoscope under contralateral hemithorax) were vital. Selective bronchial intubation and isolation allowed effective ventilation in our case.

Considering the immunodeficiency, perioperative prophylactic antibiotics, and strict asepsis for all cutaneous punctures (including venous access and regional anesthesia) is mandatory. Regional anesthesia is not contraindicated; however, the benefit of regional anesthesia should be considered against the risk of abscess formation in an immunocompromised patient. Formation of epidural abscess and late detection has been reported, hence we avoided epidural analgesia. These patients also exhibit various allergies. Hence, cross-sensitivity of propofol with egg or soya allergy must be kept in mind.

To summarize, successful anesthesia management of such patients includes perioperative chest optimization, strict asepsis, prophylactic antibiotics, careful positioning, lung isolation, avoiding N2O, a high index of suspicion for contralateral pneumothorax, vigilant monitoring with adequate systemic analgesia. Repeated surgeries and anesthesia exposure of these ‘frequent fliers’ should be kept in mind along with recurrence of pneumatoceles.

References