

Severe anaphylactic shock after anesthesia induction: An unusual initial manifestation of Churg-Strauss syndrome

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Abstract

Churg-Strauss syndrome or eosinophilic granulomatosis with polyangiitis (EGPA) is a rare multisystem disorder. A case of anaphylactic shock after induction of anesthesia, as the initial clinical presentation of Churg-Strauss syndrome in a 15-year-old girl is reported. It is extremely rare to see pediatric patients with previous perioperative anaphylaxis receiving future anesthesia; a multidisciplinary approach including allergist, rheumatologist, anesthesiologist, and surgeon is necessary in order to provide a better future anesthetic plan.

Keywords

anaphylaxis, anaphylactic shock, Churg-Strauss, eosinophilic granulomatosis with polyangiitis (EGPA), pediatric anesthesia

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Introduction

Churg-Strauss syndrome or eosinophilic granulomatosis with polyangiitis (EGPA) is a rare multisystem disorder affecting only three people per million.¹ Perioperative allergic reactions can be life-threatening but the frequency of anaphylaxis during general anesthesia is reported to be low, with a frequency between 1:5000 and 1:20,000 cases.^{2,3} Statistical data on perioperative anaphylaxis in pediatric patients are rare and the overall incidence in children is usually underreported, compared to adults.² Only a few case reports of patients with Churg-Strauss syndrome receiving anesthesia have been reported.^{4,5}

A case of anaphylactic shock after induction of anesthesia, as the initial clinical presentation of Churg-Strauss syndrome in a 15-year-old girl is presented.

Case report

A 15-year-old girl (height: 158 cm, weight: 60 kg) with achalasia was scheduled for Heller's esophagomyotomy. Her medical history included asthma,

neonatal autoimmune neutropenia at the age of 10 months, recurrent otitis media treated with myringotomy and mesenteric lymphadenitis at 7 years old. The patient also suffered recurrent upper respiratory tract infections. On the preoperative evaluation, the patient reported nocturnal regurgitation of undigested food and cough during the previous 2 months; however, she had normal lung function on spirometry and her asthma was well controlled with inhaled budesonide/formoterol (160 + 4.5 mcg/dose) twice daily and montelukast sodium 10 mg per os, once per day. Preoperative chest x-ray, pulmonary and cardiac auscultation were unremarkable. Mild paranasal sinusitis was attributed to allergic, seasonal rhinitis. Abnormal laboratory findings included a total

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IgE of 1289 IU/ml (normal range: 11–210 IU/ml). Peripheral eosinophil count was normal but toward the upper limit. Gastric and duodenal biopsies revealed moderately intense eosinophilic infiltration of the stomach and the duodenum (microscopic views with more than 93 eosinophils/high-power field, H&E stain $\times 400$).

The patient underwent rapid sequence induction anesthesia with propofol 180 mg (3 mg/kg), rocuronium 60 mg (1 mg/kg) and 100% oxygen. Endotracheal intubation was easily achieved with a cuffed tube of 6.0 mm with no visible signs of regurgitation or aspiration. Immediately after intubation, the patient developed respiratory compromise and hypoxemia; manual and mechanical ventilation were impossible. SpO₂ decreased to 70% with rapidly progressing bronchospasm and stridor. Simultaneously, the patient developed whole body maculopapular rash starting from the chest. Blood pressure decreased to 60/30 mmHg and heart rate increased to 115 beats/min. Rapid crystalloid infusion was started, 0.04 mg of adrenaline, 500 mg of hydrocortisone and 240 mg of aminophylline were administered. Mechanical ventilation under positive end expiratory pressure ameliorated oxygenation and SpO₂ rapidly recovered to 98%. Blood collection very shortly after the onset of symptoms did not show any increase in plasma tryptase.

Surgery was suspended and the patient remained intubated in the operative room for 2 h in order to be sure that the cutaneous rash improved and that the patient was hemodynamically stable. The patient was then transferred to the ICU intubated and sedated because of concerns of possible transient anaphylaxis. Chest x-ray on the ICU revealed diffuse pulmonary infiltrates; therefore, she was extubated on the third post-operative day, in the operative room. Recurrent cutaneous rash was observed during the 3 days of her ICU stay despite regular dimetindene and hydrocortisone administration.

Discussion

Churg-Strauss syndrome is a systemic small and medium vessel necrotizing vasculitis, characterized by extravascular granulomas and tissue infiltration by eosinophils.⁶ Mean age at onset is 48. Table 1 summarizes clinical criteria for classification of Churg-Strauss syndrome from the American

Table 1. Classification criteria for Churg-Strauss syndrome (adopted from the American College of Rheumatology).

Asthma
Eosinophilia of >10% in peripheral blood
Paranasal sinusitis
Pulmonary infiltrates, sometimes transient
Histologic evidence of vasculitis with extravascular eosinophils
Multiple mononeuropathy or polyneuropathy

College of Rheumatology. If 4 \geq criteria are present, sensitivity is 85% and specificity is 99.7%.^{7,8} Diagnosis is suggested by clinical findings and routine laboratory and serologic tests are not specific or sensitive for Churg-Strauss syndrome.^{7,9} In the above-mentioned patient asthma, paranasal sinusitis, pulmonary infiltrates, and extravascular eosinophilia co-existed. Follow-up of the patient was negative for antineutrophil cytoplasmic autoantibodies (ANCA) and C-reactive protein levels were within normal range. Subsequent chest x-rays showed transient patchy pulmonary infiltrates and a 2D echocardiogram of the heart revealed no signs of heart failure. The patient refused to undergo lung biopsy. IgE levels remained elevated, suggesting activation of T lymphocytes which is highly associated with eosinophilic inflammation.¹⁰ Skin tests were negative for most drugs used in anesthesia, including propofol, rocuronium, cis-atracurium, fentanyl, and midazolam.

Churg-Strauss syndrome was diagnosed by exclusion. A multidisciplinary team consisting of anesthesiologist, surgeon, intensivist, and allergist concluded that Churg-Strauss syndrome might have this atypical presentation in the pediatric patient. Cases of the syndrome in the pediatric population are only scarcely reported. Intraoperative bronchospasm and pulmonary compromise were attributed to possible bronchoconstriction and vasculitis of the vascular bed of the lung parenchyma. Cutaneous manifestations are also present in about one-half of patients. Up to one-third of patients present with gastrointestinal symptoms such as abdominal pain, diarrhea due to eosinophilic gastroenteritis or mesenteric ischemia due to vasculitis.^{11–13}

It is worth mentioning that the same patient, a year later, underwent Heller's myotomy under general anesthesia with exactly the same anesthetic regimen. Meanwhile, the patient had been assessed by an allergist and treated with two doses of omalizumab 150 mg. The risk of perioperative anaphylaxis was

reduced by administration of prednisolone 60mg per os, once daily, for 3 days before surgery and methylprednisolone 125 mg and dimetindene 4mg an hour before surgery. No anaphylactic or anaphylactoid reaction was observed.

Treatment of Churg-Strauss syndrome is based on corticosteroids and other immunosuppressants (e.g. cyclophosphamide, methotrexate, azathioprine),¹³ but none can guarantee maintenance of remission.

As far as this patient is concerned, it should be stated that it is rare to see pediatric patients with previous perioperative anaphylaxis receiving future anesthesia; a multidisciplinary approach would provide a meticulous analysis of previous suspected anaphylactic reactions and a better future anesthetic plan.

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Informed consent

Written informed consent was obtained from a legally authorized representative(s) for anonymized patient information to be published in this article.

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