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Case Report

Rare disorders of the immune system: Kimura's disease and Evans syndrome – anaesthetic implications

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Abstract

Background: Kimura's disease is a rare, chronic eosinophilic inflammatory disorder that typically affects the salivary glands, skin, lymph nodes and kidneys. Evans syndrome is an autoimmune condition characterised by the simultaneous or sequential occurrence of idiopathic thrombocytopenic purpura and autoimmune haemolytic anaemia.
Case Presentations: We present two distinct cases highlighting the anaesthetic considerations in patients with rare immune disorders. The first case involves a 35-year-old male with Kimura's disease, presenting with a post-auricular mass scheduled for surgical excision. The second case features an 18-year-old female diagnosed with Evans syndrome, who was scheduled for pan-retinal photocoagulation under topical anaesthesia due to visual disturbances.
Conclusions: Anaesthetic management of Kimura's disease (KD) requires vigilance due to potential complications such as increased intraoperative bleeding, difficult airway management from mass effect and renal impairment associated with nephrotic syndrome. In patients with Evans syndrome, key concerns include thrombocytopenia, risk of airway bleeding, systemic involvement, and the need for perioperative corticosteroid supplementation.

Keywords: Kimura's disease, Evans syndrome, Anaesthesia, Immune disorders, Airway management, Perioperative care.

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1. Introduction

The immune system is a complex network of cells, tissues, and organs that work collectively to protect the body from "foreign" invaders, including bacteria, viruses, fungi and parasites. Disorders of the immune system can be broadly classified into allergic diseases, autoimmune diseases, immune complex diseases or immunodeficiency disorders.[1] These conditions can significantly influence perioperative outcomes and pose unique challenges during anaesthetic management.

Immune function may be altered by various perioperative factors. Beyond the physiological stress response to surgery, anaesthetic agents and intraoperative analgesics can directly modulate immune cell activity. Dysregulation of immune function increases susceptibility to infections, potentially leading to complications such as impaired wound healing, systemic infections and sepsis. Therefore, thorough preoperative evaluation, individualised anaesthesia planning, and vigilant intraoperative and postoperative monitoring are essential for patients with immune system disorders. Among these, Kimura's disease and Evans syndrome are rare but clinically significant conditions that demand a multidisciplinary approach and careful anaesthetic considerations. In this report, we present two cases managed under anaesthesia, highlighting the specific challenges and implications encountered.

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2. Case Report

2.1. Case 1

A 35-year-old male presented to the outpatient department with a history of multiple subcutaneous swellings in the head and neck region. He had undergone surgical excision of a left post-auricular mass in the past and now reported a gradually enlarging mass in the right post-auricular area over the past two years. The patient also had a history of bronchial asthma for the last 10 years and was on budesonide metered dose inhaler (200 µg twice daily). On clinical examination, a lesion measuring 3.5 × 2.5 × 2.5 cm was observed in the left post-auricular region. It was non-adherent to the skin or underlying structures and the overlying skin appeared normal without redness or warmth. Local ultrasonography showed cystic lesions with increased internal vascularity. Histopathological evaluation revealed lymphoid follicle formation with prominent germinal centres, eosinophilic infiltration, increased post-capillary venules and vascular proliferation, suggestive of Kimura's disease. The patient was diagnosed accordingly and scheduled for surgical excision of the post-auricular swelling under general anaesthesia.

Airway examination revealed a Mallampati Class II with normal thyromental distance and unrestricted neck movements. A preoperative videolaryngoscopy did not reveal any airway involvement. Laboratory investigations showed hemoglobin of 14.2 g/dL, total leukocyte count of 6,403/mm³ with 13.9% eosinophils and an absolute eosinophil count of 824. Serum IgE was significantly elevated at 620 IU/mL (normal: 0–14 IU/mL). Liver and renal function tests were within normal limits. Abdominal ultrasound showed both kidneys were normal in size and echotexture. Chest X-ray revealed an opacity in the right upper lobe.

Preoperatively, the patient received tablet diazepam 10 mg the night before surgery along with salbutamol and budesonide nebulization, which was repeated on the morning of surgery. In the operating room, intravenous access was established and standard monitors were applied. Intravenous hydrocortisone 100 mg and dexamethasone 8 mg were administered to minimize airway inflammation and prevent bronchospasm. The patient was premedicated with ondansetron and fentanyl 100 µg IV. Anaesthesia was induced with propofol 200 mg IV, and muscle relaxation was achieved using Cisatracurium 12 mg, selected to minimize histamine-induced bronchospasm. Orotracheal intubation was performed smoothly using a video laryngoscope and mechanical ventilation was initiated. The patient was placed in the prone position with adequate padding of all pressure points. Anaesthesia was maintained with nitrous oxide, isoflurane and intermittent doses of Cisatracurium. The surgical procedure involved complete excision of the swelling and scalp flap creation to cover the mastoid defect. The total duration of surgery was one hour, with a blood loss of approximately 200 mL, managed with crystalloids. Urine output during the procedure was 220mL. The patient remained hemodynamically stable throughout. At the end of the surgery, neuromuscular blockade was reversed, and the patient was extubated and shifted to the recovery room for observation. There were no postoperative complications, and the patient was discharged on the seventh postoperative day in stable condition.

2.2. Case 2

An 18-year-old female presented with complaints of blurred vision for one month, along with a two-year history of menorrhagia and petechiae over her extremities. She had received multiple transfusions of packed red blood cells and platelets in the past. She was a known case of hypothyroidism, well-controlled with tablet thyroxine 25 µg once daily. Based on clinical evaluation and laboratory findings, she was diagnosed with steroid-responsive Evans syndrome associated with hypothyroidism. Her regular medications included prednisolone 5 mg once daily, azathioprine 50 mg twice daily, and tranexamic acid 500 mg as needed. On general examination, the patient appeared pale and complained of fatigue and generalized weakness. Petechial spots were noted over her trunk and limbs. Neck examination was normal with no signs of goitre. Her vital signs were within normal limits.

Initial blood investigations revealed hemoglobin of 7 g/dL, total leukocyte count of 4,000/mm³, and a critically low platelet count of 10,000/mm³. She was transfused with 4 units of packed red blood cells and 8 units of random donor platelets over five days. Post-transfusion, her hemoglobin improved to 10 g/dL, total leukocyte counts to 5,000/mm³, and platelet count to 90,000/mm³. Thyroid function tests and other labs were within normal ranges. Once stabilised, the patient was posted for pan-retinal photocoagulation under topical anaesthesia.

The night before the procedure, the patient was given oral diazepam 10 mg. Intravenous hydration with 500 mL of DNS was administered overnight. On the day of surgery, she received hydrocortisone 100 mg and dexamethasone 8 mg IV. Intraoperatively, she was administered ondansetron 4 mg and midazolam 1 mg IV for mild sedation. Topical anaesthesia was achieved using 0.5% proparacaine hydrochloride eye drops. The procedure lasted for 30 minutes, during which the patient's vital parameters remained stable. Postoperatively, hydrocortisone 50 mg IV was continued every 6 hours for 24 hours. The platelet count remained stable at 88,000/mm³, and there was no evidence of active bleeding. The patient had an uneventful recovery and was discharged on the fifth postoperative day.

Table 1: A comparative overview of anaesthetic concerns in Kimura's disease and Evans syndrome

	Kimura's disease	**Evans syndrome**
Airway management	Potential subglottic stenosis or lymphoid infiltration requiring careful airway assessment and management	No specific airway concerns but caution with intubation due to thrombocytopenia.
Bleeding risk	Eosinophilia and vascular proliferation increase bleeding risk; careful management of coagulation and haemostasis required	Thrombocytopenia increases bleeding risk; platelet transfusions may be necessary.
Allergic reactions	Potential for perioperative allergic reactions due to eosinophilia and mast cell activation	No specific concerns for allergic reactions.
Hemodynamic stability	Potential for hemodynamic instability due to vascular proliferation	Potential for hemodynamic instability due to anaemia and thrombocytopenia.
Fluid management	Careful fluid management required due to potential for vascular leakage and edema	Careful fluid management required due to potential for bleeding and anaemia.
Blood product management	No specific concerns for blood products management	Platelet transfusions may be necessary due to thrombocytopenia; careful management of blood products required.
Postoperative Care	Close monitoring for respiratory complications, bleeding and allergic reactions.	Close monitoring for bleeding, anaemia and thrombocytopenia; careful management of pain and hemodynamic required.

3. Discussion

Kimura's disease (KD) is a rare, chronic inflammatory disorder predominantly found in the Asian population, particularly affecting young males between the ages of 20 and 40, with a male-to-female ratio of 3:1.[2] It is characterised by eosinophilic infiltration and vascular proliferation involving subcutaneous tissues, lymph nodes—commonly in the head and neck (peri-auricular, cervical, axillary and lingual areas)—as well as salivary glands like the parotid and submandibular.[3] Renal involvement, seen in about 60% of patients, typically presents as membranous glomerulonephritis or nephrotic syndrome,[4] necessitating preoperative evaluation with urine microscopy and renal function tests. Intraoperative urine output monitoring is also essential.

KD is often misdiagnosed as malignancy and confirmed only through histopathological examination.[5] Differential diagnoses include angiolymphoid hyperplasia with eosinophilia (ALHE), Hodgkin's lymphoma, tuberculosis, Kaposi's sarcoma and eosinophilic granuloma.[6] Optimal treatment remains unclear, with current options including conservative medical management (steroids, radiotherapy, cryotherapy, laser therapy) and surgical excision.[7]

Associated features such as raised IgE and peripheral eosinophilia are common, and bronchial asthma is frequently observed.[8] These patients often respond better to steroids than bronchodilators during exacerbations, which justifies the use of preoperative corticosteroids. For anaesthesiologists, KD presents unique challenges. Detailed airway examination is vital, as rare localizations in the epiglottic, laryngeal or mediastinal regions may lead to airway compromise. Large cervical masses can compress airways, potentially resulting in a "cannot ventilate, cannot intubate" scenario. Difficult airway equipment should always be readily available, and awake fibreoptic intubation may be required in selected cases (**Table 1**).

Cisatracurium was chosen as the neuromuscular blocker to minimize histamine release and avoid bronchospasm. Given the lesion's vascularity, intraoperative blood loss is a concern. Balanced anaesthesia with close hemodynamic monitoring is recommended, with mean arterial pressure maintained between 75–95 mmHg. Renal protection involves avoiding nephrotoxic drugs and ensuring adequate hydration. Perioperative steroid coverage is necessary in chronically treated patients to prevent adrenal insufficiency due to hypothalamic-pituitary-adrenal axis suppression.[9]

Evans syndrome is an autoimmune disorder characterised by the simultaneous or sequential presence of direct Coombs-positive autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP) without an identifiable underlying cause.[10] Patients may present with purpura, petechiae, menorrhagia, and symptoms of anemia. Complications include bleeding due to thrombocytopenia and infections due to leukopenia or neutropenia. Preoperative planning should include readiness for transfusions and avoidance of trauma, especially during airway management (**Table 1**).

Medical management is the cornerstone of treatment. First-line therapy includes corticosteroids or intravenous immunoglobulin, with steroids dosed at 1–2 mg/kg and tapered depending on the condition. Immunosuppressants may be added if there is poor response. Anaesthetic considerations include continuation of immunosuppressants and steroids, supplemented by perioperative corticosteroids.

Due to the high risk of infection, strict asepsis is critical. In patients with coexisting autoimmune conditions such as systemic lupus erythematosus (SLE), difficult airway should be anticipated due to potential subglottic or laryngeal edema. Equipment for managing a difficult airway, such as smaller endotracheal tubes, supraglottic airway devices and bougies, should be kept ready. Laryngeal involvement may range from mild edema to acute obstruction.

Deep vein thrombosis prophylaxis and early mobilization should be considered, especially in patients with vasculitis. In our case, the main perioperative concerns included ensuring adequate platelet count, avoiding trauma and maintaining a sterile environment. The choice of anaesthesia should be tailored based on the coagulation status, the type of surgery and overall clinical condition. In this case, the procedure was safely conducted under topical anaesthesia with careful postoperative monitoring for bleeding and transfusion needs.

The anaesthetic challenges associated with Kimura's disease and Evans syndrome differ significantly, necessitating individualised approaches. Awareness of the pathophysiology and tailored perioperative management are critical for successful outcomes. In both cases presented, multidisciplinary planning and attention to specific immune-related considerations led to safe and effective perioperative care.

4. Conclusion

Kimura's disease and Evans syndrome pose distinct anaesthetic challenges due to airway involvement, renal impairment, and haematological abnormalities. Careful airway and renal assessment in Kimura's disease, and optimization of anaemia and thrombocytopenia in Evans syndrome, are essential. Anaesthetic plans should be tailored to address these unique challenges with multidisciplinary collaboration for optimal perioperative outcomes.

5. Source of Funding

None.

6. Conflict of Interest

None.

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