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Indian Journal of Clinical Anaesthesia

Journal homepage: www.ijca.in

Case Report

Anaesthetic management in a sarcoidosis patient undergoing bilateral styloidectomy: A case report

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Abstract

Sarcoidosis is a complex, multi-organ systemic granulomatous disease that presents significant challenges in anaesthetic management. This case report describes the anaesthetic approach for a 32-year-old female patient with sarcoidosis undergoing bilateral styloidectomy. Preoperative pulmonary function tests revealed mild restrictive lung impairment, and there was a potential for difficult intubation, necessitating careful preoperative preparation and monitoring. During the procedure, inflammatory responses were managed with intravenous dexamethasone and magnesium sulphate, which contributed to maintaining cardiac stability. The surgery was completed without complications, and the postoperative recovery was uneventful. This case emphasizes the need for personalized anaesthetic planning and teamwork in treating patients with sarcoidosis. It also highlights the importance of further research to improve perioperative care and develop standard guidelines for managing these patients.

Keywords: Sarcoidosis, Anaesthesia, Bilateral styloidectomy, Airway management, Multidisciplinary approach.

Received: 28-11-2024; **Accepted:** 02-04-2025; **Available Online**: 31-10-2025

1. Introduction

Sarcoidosis, a complex systemic granulomatous disease characterised by formation of non-caseating granulomas primarily in the lung, but the disease can affect other organs such as the heart, eyes, lymph nodes, skin, liver, kidney, pancreas, thyroid gland, and central and peripheral nervous system. Environmental factors, as well as genetic predisposition, are thought to be key contributors to the disease's prevalence.[1,2] Sarcoidosis has a varied global prevalence, however, its incidence is notably higher in Northern Europeans and African Americans. Such data are extremely restricted in the area of epidemiology and the condition is relatively rare in India.[3]

The clinical manifestation of sarcoidosis is highly variable, involving pulmonary, constitutional and lymphadenopathy. This variability can make diagnosis and management challenging, especially in cases where surgical interventions are needed.[4] These individuals frequently show heightened sensitivity to anaesthetics, particularly those that may impair pulmonary and cardiac function. This case report shows the challenges in administering anaesthesia in patients with systemic granulomatous disease, emphasising the complexity of management and the need for individualised treatment.

Anaesthesia may be required for various reasons in patients with sarcoidosis, including diagnostic procedures such as mediastinoscopy or lung biopsy, management of disease-related airway complications or incidental surgeries related to the chronic nature of the condition. Anaesthetic management of these patients is effective only if the systemic involvement of the disease and possible perioperative complications are well considered.

2. Case Presentation

A 32-year-old female with elongated styloid process was scheduled for bilateral styloidectomy. Her clinical

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https://doi.org/10.18231/j.ijca.56593.1761906699

presentation included persistent dry cough, dysphagia for solids, dyspepsia and progressive weight loss. On evaluation, she was diagnosed to have stage I pulmonary sarcoidosis (**Table 1**). The patient had hypothyroidism, for which she is on levothyroxine 100mcg/day. Her past surgical history included two uncomplicated caesarean sections under spinal anaesthesia. She was not on any treatment for sarcoidosis.

The patient presented with stable cardiovascular parameters, including a blood pressure of 110/70 mm Hg and a heart rate of 64 bpm. Airway assessment revealed a Mallampati Class 3, indicating potential intubation difficulty. Respiratory findings included normal breath sounds, a respiratory rate of 18 breaths per minute, and a breath-holding time of 30 seconds. Her chest X-ray was normal. Preoperative pulmonary function tests indicated a mild restrictive pattern (TLC <80% FVC < 80% FEV1/FVC ratio-normal). Laboratory investigations, including haematology and biochemistry, were normal. Her thyroid stimulating hormone (TSH) was 2.203 MUI/L. Her Electrocardiogram (ECG) and echocardiographic findings (ECHO) were normal showing no evidence of cardiac sarcoidosis. Contrast-enhanced CT (CECT) of the chest identified multiple enlarged mediastinal and hilar lymph nodes, with the largest measuring 16 mm in the right hilar region (**Figure 1**). It did not show any atelectasis. “Endoscopic ultrasound-guided fine-needle aspiration cytology" (EUS-FNAC) confirmed the diagnosis of granulomatous lymphadenitis.

Figure 1: CT chest image showing multiple enlarged mediastinal and bilateral hilar lymph nodes

The patient was premedicated with Alprazolam 0.25 mg and Ranitidine 150 mg and Metoclopramide 10 mg in the morning of the operation. In the operating room, the patient's vital signs were monitored using pulse oximetry, non-invasive blood pressure, and ECG. The patient received Ondansetron 4mg to prevent nausea and vomiting and glycopyrrolate 0.2 mg was administered as an antisialagogue as the styloid process was approached intraorally through the tonsillar fossa. The patient was preoxygenated with 100% oxygen for three minutes. Anaesthesia was induced with propofol 2 mg/kg and fentanyl 2 mcg/kg, followed by intubation using a 7 mm internal diameter south-pole endotracheal tube facilitated by vecuronium 0.1 mg/kg as a muscle relaxant. Anaesthesia was maintained with 1% sevoflurane in a mixture of air and oxygen, with inspired concentrations adjusted to keep the BIS within 40-60%. Intraoperative medications included dexamethasone injection for long-acting glucocorticoid therapy. To ensure cardiac stability, magnesium sulphate 50% injection in 100 ml NS was infused over an hour to maintain an adequate plane of anaesthesia so as to reduce muscle relaxant doses.

The surgery lasted for 2 hours, and the procedure was accomplished without any complications. After confirming spontaneous respiratory efforts, the patient was reversed with an injection of Myopyrrolate and extubated. Pain management was achieved with a 25 µg/hr transdermal fentanyl patch along with 1 gram of paracetamol injection at 8 hr intervals.

3. Discussion

Sarcoidosis is an idiopathic multisystem granulomatous disorder, primarily affecting individuals aged 20-40, with some preponderance towards females involving various organs, most commonly the lungs.[2] The staging of the disease is based on the radiological features (**Table 1**), which help in determining the course of treatment and follow up protocol.[5]

Table 1: Radiological staging of pulmonary sarcoidosis

Stage	Result	Follow up timelines
I	Bilateral hilar lymphadenopathy	Every six months at first, then annually if stable
II	Bilateral hilar lymphadenopathy and pulmonary infiltrates	Indefinitely, every three to six months for stages II, III and IV
III	Pulmonary infiltrates without bilateral lymphadenopathy	
IV	Pulmonary fibrosis	

Corticosteroids are the first-line treatment for sarcoidosis. In individuals with symptomatic pulmonary sarcoidosis, they have shown to improve overall disease control, symptoms, and standard of life. Additionally, they can also slow the progression of the illness. Biologic agents and antimetabolites are the other two types of medications that have an anti-inflammatory effect used for sarcoidosis[6] (**Table 2**).

Table 2: Common drug therapy for sarcoidosis

Drug	Dosage	Adverse effects	Anesthetic consideration
Prednisolone	20 to 40 mg per day initially 5 to 10 mg per day for long-term therapy	Weight gain Hypertension Osteoporosis Fragile skin Impaired glucose tolerance Immunosuppression Electrolyte abnormalities	Preoperative electrolyte correction. Impaired stress response replacement is required. Sliding scale if diabetic RSI if significant reflux. Careful intra-operative positioning
Methotrexate	10-15 mg weekly	Nausea; low white blood cell count; liver toxicity; lung inflammation	Complete blood count (CBC); liver (LFT)and renal function tests (RFT)
Leflunomide	10 - 20 mg daily	Nausea; low white blood cell count; liver toxicity; lung inflammation	CBC; LFT; RFT; blood pressure check
Azathioprine	50-250 mg daily	Nausea; low white blood cell count; increased infection risk; cancer risk	CBC; LFT;RFT
Mycophenolate Mofetil	500-1500 mg twice daily	diarrhoea; low white blood cell count; infection risk; cancer risk	CBC; LFT
Mycophenolic acid	360-1080 mg twice daily	Diarrhoea; low white blood cell count; infection risk; cancer risk	CBC; LFT
Infliximab or Bioimilars	3-5 mg/kg initially, then 2 weeks later, followed by every 4-6 weeks	Infection risk; allergic reactions; cancer risk	TB screening; Hepatitis B and C screening; avoid in severe CHF, demyelinating diseases, active TB, deep fungal infections
Adalimumab	40 mg every 1-2 weeks	Infection risk; cancer risk	TB screening; Hepatitis B and C screening; avoid in severe CHF, demyelinating diseases, active TB, deep fungal infections
Rituximab	500-1000 mg every 1-6 months	Infection risk; allergic reactions	Hepatitis B and C screening; monitor IgG levels with long-term use
RCI(ACT Har Gel)	40-80 units twice weekly	Diabetes; high blood pressure; fluid retention; anxiety	Glucose levels; blood pressure monitoring
Hydroxychloroquine	200 mg daily or twice daily or weight-based	Eye damage; prolonged QTc interval	Regular eye exam

This case discusses a 32-year-old female diagnosed with stage I pulmonary sarcoidosis who underwent bilateral styloidectomy. Preoperative pulmonary function tests revealed a mild restrictive pattern (TLC <80%, FVC <80%, FEV1/FVC ratio-normal), necessitating vigilant respiratory monitoring, consistent with literature noting similar pulmonary abnormalities in sarcoidosis.[7] The Mallampati Class 3 airway indicated potentially difficult intubation, supported by studies suggesting a higher risk of difficult intubation in sarcoidosis patients.[8] Additionally, sarcoidosis can involve the larynx and cause tracheal stenosis, mediastinal compression, which interferes with the passage of the endotracheal tube.[9] Therefore, adequate preparation and availability of advanced airway management tools are essential. In this case, we successfully performed intubation with a South Pole tube of size 7 mm internal diameter without any complications.

Anaesthesia induction involved injection midazolam, fentanyl, propofol, and vecuronium for maintenance of sedation and muscle relaxation effectively. The administration of dexamethasone managed the inflammatory response, aligning with perioperative care guidelines for sarcoidosis patients as highlighted by Sanders et al.[7]

Continuous monitoring of vital signs, including pulse oximetry, non-invasive blood pressure, and ECG, was crucial. Intraoperative stability was maintained without significant hemodynamic fluctuations, showcasing the importance of meticulous monitoring and prompt intervention.

Postoperative pain management with a transdermal fentanyl patch and paracetamol was effective, preventing stress-induced disease exacerbations. The patient recovered well postoperatively, with no complications observed.

Calaras D et al., highlighted the application of intraoperative inhalational bronchodilators to increase the airway patency and lower the risk of bronchospasm. Patients with restrictive lung disease should maintain adequate tidal volume and continuous oxygen saturation monitoring using non-invasive ventilation along with other oxygen therapy.[10]

Non-caseating granuloma's infiltration into heart tissue, specifically damaging myocardium, is a characteristic of cardiac sarcoidosis (CS). Heart block, cardiomyopathy, and arrhythmias are some of the causes of sudden cardiac death.[11] Anaesthetists must precisely evaluate the patients with CS by monitoring cardiac functions and rhythm, and avoid drugs that could worsen arrhythmias. Comprehensive cardiac assessments should be undertaken from preoperative evaluations. Intrusive monitoring measures may be required intraoperatively to ensure effective cardiovascular treatment in cases where hemodynamic instability is predicted.

Renal sarcoidosis has significant challenges in anaesthesia management due to its potential risk of renal impairment and associated complications. Nephrocalcinosis and renal scarring can result from hypercalcemia. Therefore, comprehensive preoperative assessment should be performed, which should include electrolyte values, especially calcium, and renal function tests.[12] Avoid or use the drugs with caution that are mostly eliminated via the kidneys. Avoiding fluid overload requires careful fluid management, particularly in patients with impaired renal function.

Second most frequently affected organ in sarcoidosis is the skin, which includes lupus pernio, erythema nodosum, and different kinds of papules.[13] Patients may be using immunosuppressants or corticosteroids, which can influence the choice of anaesthetic and increase the risk of infection.

Significant side effects from ocular sarcoidosis might include inflammation and blindness as a result of granulomas developing in the eyes.[14] Anaesthetists must avoid using drugs like succinylcholine that might raise intraocular pressure, particularly in glaucoma patients.

Managing anaesthesia in sarcoidosis patients, particularly those with mediastinal and hilar lymphadenopathy, is challenging. It highlights the importance of preoperative optimization, including corticosteroid administration, to reduce the risk of inflammatory responses.

4. Conclusion

This case highlights the importance of an individualized anaesthetic approach for sarcoidosis patients, emphasizing the need for thorough preoperative assessment, precise intraoperative management, and effective postoperative care. Future research should focus on developing standardised anaesthetic guidelines to improve perioperative outcomes for this patient population.

5. Ethical Considerations

Informed consent was obtained from the patient for the publication of this case report. This adherence to ethical standards ensures that the patient's rights and privacy are respected throughout the reporting process.

6. Declaration of Patient Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

7. Source of Funding

None.

8. Conflict of Interest

None.

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Cite this article: Sankar I, Gayathri B, Chandrasekar G. Anaesthetic management in a sarcoidosis patient undergoing bilateral styloidectomy: A case report. *Indian J Clin Anaesth*. 2025;12(4):737–741.