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Dr. Rakshitha R
Department of Anaesthesiology, Dr Prabhakar Kore Hospital, JNMC KLE University Campus, Belagavi, Karnataka, India

Dr. Raghavendra H Kalal
Department of Anaesthesiology, Dr Prabhakar Kore Hospital, JNMC KLE University Campus, Belagavi, Karnataka, India

Corresponding Author:
Dr. Rakshitha R
Department of Anaesthesiology, Dr Prabhakar Kore Hospital, JNMC KLE University Campus, Belagavi, Karnataka, India

A case report: Anaesthetic management in a child with Rosai-Dorfman disease undergoing cervical lymph node excisional biopsy

Rakshitha R and Raghavendra H Kalal

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Abstract
Rosai-Dorfman disease (RDD) is a benign, self-resolving disorder often characterized by cervical lymphadenopathy. This raises complications for anaesthesiologists given the multiorgan involvement, particularly with the airway, which complicates mask breathing and intubation. A 14-year-old girl diagnosed with Rosai-Dorfman disease received an excisional biopsy for bilateral cervical oedema that had persisted for one year. She had received treatment with dexamethasone and sirolimus and had a recent history of mumps infection. The PET-CT revealed significant lymphadenopathy and hypermetabolic lesions. A complicated airway cart was constructed in the operation room. Standard monitors were used, and the patient underwent preoxygenation. Mask ventilation was difficult owing to oedema; nevertheless, sufficient breathing was accomplished using a size 1 oropharyngeal airway. Anaesthesia was initiated with ketamine after the achievement of sufficient anaesthetic depth. Succinylcholine was administered as a muscle relaxant, and intubation was successfully performed with a 5.5 mm uncuffed tube via video laryngoscopy. Anaesthesia was sustained using oxygen, nitrous oxide, isoflurane, and muscle relaxants. The procedure carried out without any further challenges. and the patient was extubated after complete recovery from anaesthesia. RDD may complicate airway care due to lymphadenopathy and soft tissue tumours. In this instance, achieving an adequate mask fit proved difficult; yet, intubation was executed successfully without difficulties, underscoring the need of readiness for tough airway situations.
This case highlights the importance of thorough pre-anaesthetic assessment and careful intraoperative management in patients with RDD undergoing surgical procedures.4

Keywords: Rosai-Dorfman Disease, airway management, sinus-histiocytosis with massive lymphadenopathy

Introduction
Rosai-Dorfman disease is a histiocytic proliferative idiopathic disorder characterized by the presence of sinus histiocytosis with mass lymphadenopathy, with benign haematopoietic and fibrous proliferation in head and neck. Extra-nodal involvement is rare and unusual in central nervous system (CNS) affecting 75% to brain leptomeninges. Surgery is indicated to relieve compressive symptoms or biopsy studies. Possible anaesthetic problems include acute airway obstruction, superior cava vena syndrome (SCVS), cardiac or mediastinal compression on the pulmonary artery, acute pulmonary oedema or cardiopulmonary collapse.

Case report
A 14-year-old girl, weighing 40 kg, with a documented case of Rosai-Dorfman illness, arrived for excisional biopsy of bilateral cervical enlargement that had persisted for one year. The diagnosis of Rosai-Dorfman disease was established one year ago by lymph node biopsy, followed by a 6-month course of Dexamethasone and a subsequent 6-month regimen of Sirolimus. A recent history of mumps infection, which is believed to exacerbate the swelling, now involves the administration of Tablet Lenalidomide 10mg once daily in the morning for 21 days as part of a two-month regimen. The patient had a history of snoring, indicative of obstructive sleep apnoea. The patient has had obstructed breathing when laying down due to swollen palatine tonsils for the last month. The neck swelling was painless, rubbery in texture, and non-tender, located in the contralateral submandibular area. Her airway was sufficient, with no indications of constriction. The oral cavity examination

indicated significantly enlarged tonsils bilaterally, accompanied by a modified Mallampati classification of 4. Patients' whole body 18 F-FDG Positron Emission Computed tomography (PET-CT) revealed metabolically active cervical, supraclavicular, infraclavicular, mediastinal, pulmonary hilar, abdominal, and inguinal lymph nodes. Hypermetabolic lesion affecting the nasal cavities, nasopharyngeal tonsil, oropharyngeal tonsil, and skeletal structures. A difficult airway cart, equipped with a laryngeal mask airway, various sizes of masks, oropharyngeal airways, nasopharyngeal airways, a bougie, and a stylet, was maintained on standby in the operating room. Intravenous (IV) access established using a 20 G cannula in the right upper leg. The patient was transferred to the operating theater. All ASA standardized monitors, including the pulse oximeter, blood pressure cuff, and three-lead electrocardiography, were attached. The patient received preoxygenation with 100% oxygen. The patient received premedication with Injection Glycopyrrolate at a dosage of 0.005 mg/kg and Injection Fentanyl at a dosage of 2 mcg/kg. The patient was induced with 40 mg of Injection Ketamine, 40 mg of Injection Propofol, and 1.5% Isoflurane. During preoxygenation, challenges in maintaining the mask were experienced due to bilateral submandibular oedema. Effective bag and mask ventilation were achieved using a size 1 oropharyngeal airway. Succinylcholine 1.5 mg/kg was delivered upon the confirmation of sufficient mask breathing. The patient was intubated using a 5.5 mm ID uncuffed tube, utilizing video laryngoscopy to meticulously navigate the laryngoscope blade and endotracheal tube inside the oral cavity to prevent unintentional harm to the tonsils. Anaesthesia was sustained using oxygen, nitrous oxide, and isoflurane as the inhalational agent, with an injection of Atracurium at a maintenance dosage of 0.1 mg per kg administered to provide sufficient muscular relaxation. The patient was placed on pressure-controlled mechanical breathing with a tidal volume of 5 to 7 ml/kg of body weight. Injection of fentanyl at a dosage of 1 mcg/kg was used to sustain intraoperative analgesia and hemodynamic stability. Heart rate and mean arterial pressure within 20% of baseline levels. No episodes of hypoxia or hypercarbia occurred. The surgical procedure proceeded without complications. At the conclusion of the procedure, neuromuscular blockade was antagonized with Glycopyrrolate

Discussion

Rosai-Dorfman disease typically presents with extensive cervical lymphadenopathy, most often bilateral and painless (87%). At first lymph nodes are mobile and discrete, but over time they become adherent and tend to develop into a large multinodular mass. The axillary (23.7%), inguinal (25.7%) and mediastinal (14.5%) regions may be affected, though not as severely as the cervical region. The diagnosis of RDD is based on the clinical history and confirmed by histopathological examination. Specimens may be obtained by open surgical biopsy or fine needle aspiration[3]. Our patient presented painless unilateral cervical lymphadenopathy with compromised lymph nodes in the mediastinum (pretracheal, prevascular, aortopulmonary window, right hilar and subcarinal), retroperitoneum and mesentery. It is essential, in preoperative evaluation of Rosai-Dorfman disease, to establish the degree of organ involvement. Classical clinical presentation, along with painless cervical lymphadenopathy, associates with nasal obstruction, adenoidal hypertrophy, fever, neutrophilia, and elevated erythrocyte sedimentation rate, with polyclonal hypergammaglobulinemia [1].

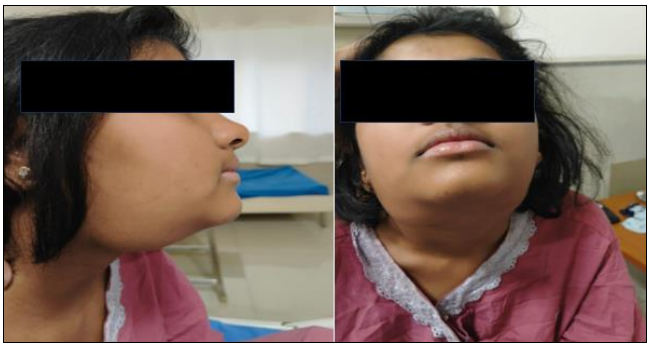
Rosai-Dorfman disease encompasses most of the features that can be encountered in difficult airway like difficult mask fit, bag mask ventilation and difficult intubation can also occur because of distorted airway anatomy due the presence of lymphadenopathy and soft tissue mass.[4] In this case, the patient demonstrated classical RDD features, including bilateral submandibular lymphadenopathy and enlarged tonsils, which are known to compromise airway patency. A significant concern in patients with extensive head and neck involvement is the potential for airway obstruction both during sedation and after extubation. Preoperative evaluation revealed features suggestive of obstructive sleep apnoea, further increasing the perioperative risk. The presence of nasopharyngeal and oropharyngeal hypermetabolic lesions, along with bilateral tonsillar hypertrophy, necessitated avoidance of nasal airway adjuncts to prevent trauma and bleeding.

In pre-anaesthetic evaluation of RDD, evaluation of the degree of organ involvement and the location of intracranial occupation is essential. An assessment of the respiratory system should include checking for extrinsic airway compression and an obstructive or restrictive pattern on lung function tests because RDD with cervical lymphadenopathy is commonly present. Tracheal intubation should be performed under video laryngoscopy for the preparation of difficult airway intubation [5].

Our anaesthetic plan prioritized airway security while minimizing the risk of airway collapse. Ketamine was chosen for induction due to its ability to preserve airway reflexes and maintain respiratory drive, especially beneficial in cases of potential airway compromise. Succinylcholine was administered after ensuring adequate mask ventilation, allowing for rapid sequence induction while minimizing apnoea-related risks. The use of video laryngoscopy allowed for careful navigation around the hypertrophied tonsillar tissue and facilitated a successful intubation.

Maintaining anaesthesia with a combination of isoflurane, nitrous oxide, and controlled ventilation helped provide adequate depth while ensuring stable hemodynamics. Atracurium was selected as the muscle relaxant due to its predictable metabolism and ease of reversal. Intraoperative analgesia was managed with fentanyl to blunt sympathetic responses. The patient's perioperative course remained uneventful, owing to meticulous planning, including preparation of a difficult airway cart and adherence to difficult airway algorithms.

This case underscores the importance of individualized anaesthetic management and preparedness when dealing with rare diseases like RDD. Anaesthesiologists must be vigilant about airway complications, especially when lymphadenopathy is extensive or when extra-nodal masses involve the airway structures. Postoperative care should include monitoring for delayed airway obstruction or respiratory compromise, particularly in patients with obstructive sleep symptoms or recent upper respiratory infections.



Conclusion

Rosai-Dorfman disease encompasses most of the features that can be encountered in difficult airway like difficult mask fit, bag mask ventilation and difficult intubation can also occur because of distorted airway anatomy due the presence of lymphadenopathy and soft tissue mass. Mask fit was difficult in our patient, as she had massive bilateral submandibular and post auricular lymph node enlargement. Difficult intubation can also be because of intratracheal extension of mass though such problem was not encountered in our case. Nasal and oral airways were avoided in our patient to avoid trauma and bleeding because mass was extending into nasal cavities, soft palate.

Conflict of Interest

Not available

Financial Support

Not available

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