Rosai-Dorfman Disease – perianaesthetic concerns

P. KHANNA, R. GARG and R. A. SUNDER

Abstract: Rosai-Dorfman Disease (RDD) may be challenging for anesthesiologist in view of its multisystem involvement, specially the airway. We present a patient with RDD scheduled for bilateral rhinotomy and bilateral neck dissection for nasal obstruction and gross bilateral lymphadenopathy. Care should be taken in airway management because of the distortion of airway anatomy by the soft tissue mass with possible intracheal extension of mass. Central venous catheterization should be guided by ultrasonography.

Key words: Rosai-Dorfman Disease, anesthesia, difficult airway, airway distortion, cervical lymphadenopathy.

INTRODUCTION

Rosai-Dorfman Disease (RDD), historically called as Jaun Ron Fever, is an uncommon benign self limiting disorder, most commonly presenting as bilateral cervical lymphadenopathy. RDD is the term now commonly used to describe cases of sinus histiocytosis with massive lymphadenopathy (SHML) and was first described by Rosai and Dorfman in 1969. It is a disease more commonly found among children and young adults, with a slight male preponderance. They present with massive painless cervical lymphadenopathy, fever, raised elevated erythrocyte sedimentation rate, weight loss, hypergammaglobulinemia, and neutrophilia. These patients may be challenging for the anesthesiologist in view of multisystem involvement, specially the airway. We present a patient with RDD for excision of mass.

CASE REPORT

A 30-year-old 60 kg male, known case of Rosai-Dorfman disease was posted for bilateral rhinotomy and bilateral neck dissection. He also had a history of frequent episodes of epiphora and epistaxis since 17 years, bilateral nasal obstruction and neck swelling since 15 years. He also had a history of mouth breathing, snoring and apneic episodes during sleep. On examination, gross bilateral lymphadenopathy involving cervical, supra clavicular and submandibular glands was present. Airway examination revealed restricted mouth opening, limited neck extension and with a modified Mallampati class 3. His neck circumference was 44 cm. The electrocardiogram and chest x-ray was normal. Contrast enhanced computed tomography (CECT) revealed bilateral supraclavicular lymphadenopathy with a right paratracheal lymph node measuring 1.5 cm. Bilateral cervical, postauricular, submandibular and submental nodes were also involved. Both internal jugular veins were compressed by enlarged nodes. CECT of paranasal sinuses showed enhancing soft tissue mass in the nasopharynx and oropharynx extending into the nasal cavities, pterygoid fossae and soft palate.

He was premedicated with ranitidine 150 mg in night and morning. Glycopyrrolate (0.2 mg) was given intramuscularly 45 minutes before shifting to the operation room. He was explained about awake fiberoptic guided tracheal intubation. In the operation room, routine monitoring included electrocardiogram, pulse oximeter and automated non-invasive blood pressure measurement. The airway was topically anesthetized by gargling 4% viscous lidocaine, 15% lidocaine spray (3 puffs) and with 4 mL of 2% lidocaine with ‘spray and go’ technique using an epidural catheter inserted via the fiberoptic suction port. Midazolam 2 mg was given intravenously. Fiberoptic guided tracheal intubation was accomplished orally with a 8.0 mm ID endotracheal tube. Propofol (100 mg) and fentanyl (100 µg) was administered intravenously. Lungs were ventilated with 1.2% isoflurane in oxygen and nitrous oxide (50:50). Neuromuscular blockade was achieved with vecuronium 6 mg. A peripherally inserted
central venous catheter (PICC) for central venous pressure and an arterial cannula in radial artery for invasive blood pressure monitoring was secured. An oral pack was placed. Capnography and temperature monitoring was also initiated. Anesthesia was maintained with 1% isoflurane in oxygen and nitrous oxide (50:50). Analgesia was provided with 6 mg morphine intravenously. Controlled hypotension was provided with titrated volatile agent (1-2%) and esmolol intraoperatively. There was a blood loss of 850 mL that was replaced with 2 packed red blood cells.

Total duration of the surgery was 3 hrs 30 minutes. At the end of surgery, residual neuromuscular blockade was reversed with neostigmine and glycopyrrolate. The trachea was extubated and patient was shifted to the post anesthesia care unit for observation. Patient remained stable and was shifted to the ward the next day and discharged on the 7th postoperative day uneventfully.

**DISCUSSION**

Little is known regarding the pathogenesis of Rosai-Dorfman disease. Most authors have suggested that it represents either an autoimmune disease or a reaction to an infectious agent that has yet to be identified. Currently it is best considered as a benign, idiopathic histiocytosis.

Though reports regarding medical and surgical considerations are mentioned in the literature, the anesthetic considerations in RDD patients have not been described. The multisystem involvement in RDD can be of concern to the anesthesiologist in the perioperative period.

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 he 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.

In view of difficult airway we planned awake fiberoptic guided tracheal intubation in our patient. Our patient had paratracheal lymph node enlargement and a difficulty in surgical approach was anticipated owing to massive bilateral lymphadenopathy and paratracheal lesion. Presence of lymphadenopathy precluded us from giving glossopharyngeal nerve block (external approach was not possible because of lymphadenopathy and internal approach was not done because of tonsillar enlargement). For the same reason, blind nasal intubation should also be avoided. Nasal and oral airways were avoided in our patient to avoid trauma and bleeding because mass was extending into nasal cavities, soft palate and the patient had history of repeated epistaxis. In a small study it was concluded that maximum inspiratory flow is impeded by the effects of topical local anaesthetic in the upper airway (2). But tracheal intubation under general anesthesia was not an option due to presence of difficult airway. In case of airway obstruction with topicalization or sedation, we were ready with our difficult airway cart including bag and mask, laryngeal mask airway and needle cricothyroidotomy apart from surgical tracheostomy. We were successful in achieving tracheal intubation in one attempt using fiberoptic bronchoscope though the airway anatomy was slightly distorted due to presence of lymphadenopathy. Our patient had a history of mouth breathing, snoring and episodes of fragmented sleep during night suggestive of obstructed sleep apnea probably because of tonsillar and lymph node enlargement. So sedative premedication should be cautiously used.

Common sites of extranodal involvement include skin, eye and adnexae, the upper airway, salivary glands, paranasal sinuses, genitourinary system, central nervous system (CNS) – intracranial and spinal involvement, bone, breast, soft tissues and thyroid (Table 1) (3, 4).

Immune disorders occur in approximately 13% of patients with RDD, with antineutrophil cytoplasmic antibodies and joint disease being the most common findings (4). Patients with extranodal RDD however should have a diagnostic workup for immunologic abnormalities, since almost half of these patients with extranodal head and neck manifestations have immune dysfunction.

Treatment of RDD is based on clinical manifestations. Many lesions are asymptomatic, heal spontaneously, and do not require treatment. When destructive or disseminated disease is present, radiotherapy, surgical excision, systemic glucocorticoids and alkylating agents may be used (12). Radiotherapy may lead to stiffness in the neck tissue and reduced submandibular space. The side effects pertaining to long term steroids and alkylating agents should be kept in mind during perioperative period.

Care should be taken during placement of central venous catheter as the internal jugular vein.
Anatomical location may be distorted by enlarged lymph nodes as happened in our case too.

We conclude that patients with Rosai-Dorfman Disease needs to be evaluated extensively for multi organ involvement. Care should be taken in airway management because of the distortion of airway anatomy by the soft tissue mass with possible intrachel extension of mass involvement and their functional consequences (OSA, tracheal or great vessels compression etc). Central venous catheter placement should be executed cautiously.

References


Table 1
Organ involvement, its symptomatology and perioperative concerns of RDD (5, 6, 7, 8, 9, 10, 11)

<table>
<thead>
<tr>
<th>Organ Involvement</th>
<th>Symptoms</th>
<th>Concerns</th>
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<tbody>
<tr>
<td>Airway Distortion</td>
<td>obstructive breathing</td>
<td>obstructed airway, difficult ventilation and intubation</td>
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<tr>
<td>Eye and Orbital structures</td>
<td>exophthalmos, blepharoptosis, conjunctivitis, keratitis, diplopia, dry eyes, and photophobia</td>
<td>mask placement, risk of injury to eye</td>
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<tr>
<td>Skeletal System</td>
<td>osteolytic lesion</td>
<td>evaluation of bony pain, care during positioning</td>
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<tr>
<td>Thyroid</td>
<td>enlargement with lymph node enlargement</td>
<td>differential with thyroid malignancy</td>
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<tr>
<td>Central Nervous System (intra cranial and spinal)</td>
<td>headache, seizures, cranial nerve deficits, numbness and paraplegia</td>
<td>neurological assessment and specific perioperative care</td>
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<tr>
<td>Immune System</td>
<td>autoimmune hemolytic anaemia and polyclonal gammopathy</td>
<td>haematologic monitoring, strict asepsis</td>
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