

Perioperative Management Challenges and Anaesthetic Considerations for a HIV Positive Patient of Multiple Symmetric Lipomatosis (Madelung Disease): A Case Report

Kapil A Kulkarni¹, Vipin V Nair², Anuj Kumar¹

Abstract

A 60-year-old man with obstructive sleep apnea and human immunodeficiency virus positive status presented to this hospital with a huge mass at the base of his neck which caused him to sleep on his left side for the past 6 years; he was eventually diagnosed with Madelung's disease. He was scheduled for the removal of the tumor through reconstructive surgery. Madelung's disease is a rare disorder characterized by the development of benign fatty tumors in a symmetrical pattern primarily in the upper body. Considering the patient's American Society of Anesthesiologists grade, co-morbidities, and specific challenges such as difficult airway and prone positioning, careful planning for anaesthesia was crucial. Therefore, the decision was made to use general anaesthesia with awake fiberoptic intubation to reduce the possible dangers of difficult airway complications during the initial stages of the procedure. Management of Madelung's disease requires a multidisciplinary approach and thorough pre-operative assessment, patient counseling, meticulous monitoring and airway planning to achieve patient comfort and safety. Continuous monitoring, pain treatment and adequate follow-up after surgery all help to ensure a smooth recovery.

Keywords: Multiple Symmetric Lipomatosis, General Anaesthesia, HIV infection, Awake fiberoptic intubation

Introduction

Madelung's disease or multiple symmetrical lipomatosis (MSL) is a rare disorder characterized by prominent, symmetrical, non-encapsulated fat masses involving the face, neck, occipital region, and supraclavicular fossa causing serious cosmetic deformity and neck immobility. Fat can also penetrate deeper, affecting arteries, nerves, and muscles, compressing the larynx, trachea, and esophagus and producing dyspnea [1]. The etiology of MSL remains unclear, but chronic alcoholism, metabolic disturbances, and hormonal imbalances have been proposed as potential contributing factors. MSL is often associated with significant functional impairment, cosmetic concerns, and the presence of co-existing medical conditions [2]. The characteristics of Madelung's disease impose a careful anaesthetic approach because it makes airway management very challenging and peripheral or spinal block difficult, depending on where the masses are located. Here, we present a case of MSL in a patient with human immunodeficiency virus (HIV) infection on anti-retroviral therapy (ART), highlighting the clinical presentation, anaesthetic considerations and perioperative challenges faced during operative procedure.

Case Report

A 60-year-old male with documented HIV infection on ART presented with complaints of swelling over the nape and front of his neck, upper back and both flanks for the past 6 years. He was planned

to undergo excision of the mass over the nape of his neck by the reconstructive surgery team. Sulpha medications rendered him allergic. The swelling began as a small 2 × 2 cm enlargement and grew over time to its present size of 15 × 15 cm. In addition, he indicated restricted neck movement, hoarseness of voice since 2 years and signs of obstructive sleep apnea (Fig. 1a).

He was diagnosed with HIV in 2007 and was on regular anti-retroviral therapy. He was also treated for pulmonary tuberculosis in 2007 with anti-tubercular therapy.

Past medical or surgical history was insignificant. He was morbidly obese and had mixed diet with normal bowel and bladder habits. He was a chronic smoker, consuming 30–40 cigarettes per day for 10 years but stopped in 2004. He used to consume about 60 mL of alcohol daily for 15 years but has left since 2004.

General physical examination was within normal limits. His blood pressure was 132/84 mm of Hg. He had firm dentition, fair venous access, and good exercise capacity with a METS score <4. His breath-holding time was 18 s. He was 168 cm tall and weighed 79 kg, with BMI of 28 kg/m². His STOP-BANG score was six out of eight, indicating severe obstructive sleep apnea. Airway examination revealed a mouth opening of more than three finger breadths, Mallampati Class 3, restricted neck extension due to a huge fat pad over the nape of his neck, mobile temporomandibular joint, and a thyromental distance of approximately 6.5 cm (Fig. 1b).

Systemic examination was essentially normal. Spine examination

¹Department of Anaesthesiology & Critical Care, Armed Forces Medical College, Wanowrie, Pune, Maharashtra, India.

²Department of Surgery, Armed Forces Medical College, Wanowrie, Pune, Maharashtra, India.

Address of Correspondence

Dr. Kapil A. Kulkarni,

Assistant Professor, Department of Anaesthesiology & Critical Care, Armed Forces Medical College, Wanowrie, Pune, Maharashtra, India.

E-mail: kapsguy07@gmail.com

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Figure 1: preoperative evaluation 1(a),1(b)

revealed no abnormalities. On local examination, the swelling on the nape of the neck was approximately 35 × 25 cm in size, oval-shaped, non-pulsatile, non-tender, and without any cough impulse, superficial skin changes, or palpable pulsation.

The pre-operative investigations of the patient are detailed in Table 1. Notably, chest X-ray and electrocardiogram were normal indicating healthy cardiac function. The 2D echocardiography unveiled a left ventricular ejection fraction of 60%, along with concentric left ventricular hypertrophy and trivial tricuspid and mitral regurgitation. A tailored management plan was devised to address the patient's specific concerns and optimize outcome.

Anaesthesia Protocols

Pre-operative assessment

A comprehensive evaluation of the patient's medical history and examination including special emphasis on airway was conducted, with particular attention to the severity and extent of Madelung's disease. The patient was classified as American Society of Anesthesiologists (ASA) physical status Grade III. A detailed pre-operative counseling was provided and written informed consent was obtained.

Fasting guidelines and Pre-operative room assessment

The patient was instructed to follow appropriate fasting guidelines, minimizing the risk of aspiration during the procedure. Peripheral venous access was established with an 18 G catheter in both the right and left arms.

Patient positioning and monitoring

The patient was positioned comfortably by placing two pillows below his occiput as he could not rest his head onto the bed due to the huge mass on the neck and standard ASA monitors, including

Table 1. Blood Haematology & Biochemistry Investigations			
Investigation	Values	Normal Range	
Haemoglobin	13.6 g/dL	M 13-17 g/dL	
Total leukocyte count	7,100/mm ³	4000 – 11000 mm ³	
Differential leukocyte count	50% neutrophils 44% lymphocytes 6% monocytes 1% eosinophils	40-75% neutrophils 20-45% lymphocytes 2-10% monocytes 0-0-6% eosinophils	
Platelet count	2.74 lakh/mm ³	150,000-400,000 mm ³	
Blood Urea	6.4 mg/dL	10 – 50 Mg/dL	
Serum Creatinine	0.7 mg/dL	0.7 – 1.3 Mg/dL	
Serum Total Bilirubin	0.34/ mg/dL	0.2 – 1.0 Mg/dL	
Sodium/Potassium Levels	135/5.1 mmR/L	B6-M5 /3-5-5-/mmR/L	
Total Protein	8.12 g/dL	6.4 – 8.2 g/dL	
AST and ALT	15.8 U/L 23.2 U/L	16-63 U/L	
CD4 viral counts	1234 cells/mm ³	500 – 1400/mm ³	

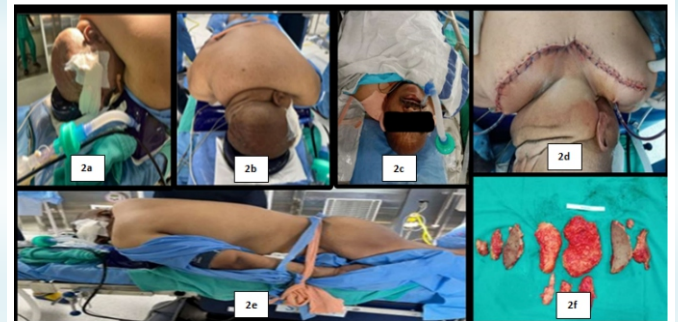


Figure 2: Intraoperative management 2(a), 2(b), 2(c), 2(d), 2(e), 2(f)

electrocardiography, non-invasive blood pressure, and pulse oximetry were applied. Supplemental oxygen was administered through a nasal cannula to optimize oxygenation (Fig. 1 c).

Airway management and conduct of Anaesthesia

A general anaesthesia technique with awake fiberoptic intubation was planned.

To desensitize the upper airway for awake intubation, topical anaesthesia with 10% lignocaine spray was applied. Nebulization was done with 4 mL of 4% lidocaine over 20 min in the pre-operative room. The following airway blocks were given by landmark technique: Glossopharyngeal nerve block – 2 mL of 2% lidocaine on either side, Superior laryngeal nerve block – 2 mL of 2% lidocaine on either side, transtracheal block – 4% lidocaine directly into the tracheal lumen using a 22 G intravenous (IV) Cannula. Additional interventions included administration of injection glycopyrrolate (0.2 mg IV). Conscious sedation was induced using IV dexmedetomidine at a dose of 1 mcg/kg IV bolus over 10 min through an infusion pump to achieve a state of reduced anxiety and cooperation. The sedation level was carefully titrated to maintain patient comfort and cooperation while ensuring responsiveness and preservation of protective airway reflexes.

Once the patient was adequately sedated, we performed an awake fiberoptic intubation with a flexible fiberoptic bronchoscope (AMBU). The scope was inserted through the mouth with a bite block in situ and was advanced gently until the trachea was reached. Once the trachea was visualized, an ID 7.5 mm flexometallic endotracheal tube (ETT) was carefully threaded over the bronchoscope and advanced into the trachea. The bronchoscope was then removed, leaving the ETT which was secured after ensuring confirmation of correct placement by auscultation and capnography.

Following successful intubation, anaesthesia was induced with IV propofol at 2 mg/kg and IV vecuronium at 0.1 mg/kg. Anaesthesia was maintained with sevoflurane and IV analgesics; muscle relaxant was administered as required.

The biggest challenge, in this case, was turning the patient prone. Hence, we secured the airway while the patient was on trolley and then turned prone onto the OT table. All the pressure points including eyes were adequately padded with soft bolsters (Fig. 2).

The actual procedure took 3 h. Ringer's lactate (RL) solution, 2200 mL, was given without transfusion of blood products. Perioperative blood loss was about 500 mL, including the resected soft-tissue mass (Fig. 2b). Notably, no adverse events occurred during the anaesthetic course.

Extubation was successfully performed after the patient was turned supine again onto the trolley with smooth, uneventful recovery. All vital signs and parameters were normal following the operation.

Throughout the procedure, SpO₂ levels were maintained above 95%, and ET CO₂ was maintained between 35 and 40 mmHg, ensuring optimal ventilation. The patient was ventilated in pressure-controlled mode to target a tidal volume of 6–8 mL/kg IBW with a peak airway pressure ranged from 22 to 24 cm of H₂O. Antibiotic prophylaxis was administered through injection cefotaxime 1 g, and additional medications included injection paracetamol 1 g IV, injection dexamethasone 8 mg IV, injection ondansetron 8 mg IV, and injection tramadol 50 mg IV. Mean arterial pressure was maintained between 70 and 90 mmHg to ensure adequate tissue perfusion.

Post-operative instructions involved maintaining a head-up position at a 30-° angle and implementing a nil per oral regimen for the initial 6 h after surgery. Vitals were continuously monitored for the first 24 h. RL at a rate of 120 mL/h was recommended until the resumption of oral intake. For post-operative analgesia, the patient received injection paracetamol 1 g IV and injection tramadol 50 mg IV every 8 h for 3 days. Vigilance was maintained for any signs of airway compromise, such as obstruction or swelling, and prompt intervention was planned if necessary.

Discussion

Airway management challenges arising from anatomical anomalies associated with Madelung's disease pose inherent obstacles necessitating the use of sophisticated intubation techniques. Infrequent occurrence of macroglossia in Madelung's disease accentuates the difficulties with airway management, necessitating the implementation of specialized procedures, such as awake fiberoptic intubation and video laryngoscopy to attain optimal control and visualization. Associated lipomas exhibit heightened vascularity and susceptibility to hemorrhagic complications, underscoring scrupulous attention to hemostatic measures.

Rarity of Madelung's disease renders a paucity of standardized anaesthesia protocols, necessitating the adoption of individualized

approaches tailored to accommodate unique patient attributes. Associated comorbidities, such as HIV infection and pulmonary, give rise to complexities mandating a nuanced approach to anaesthetic management due to potential interactions and health implications.

ART therapy in HIV-infected Madelung patients demands consideration of potential drug interactions with anaesthetic drugs. Furthermore, the impact of HIV on immunological function heightens the susceptibility of these patients to infections and exerting influence on the post-operative recovery.

Post-operative complications in Madelung's disease include the reoccurrence of lipomas and predisposition to adverse events such as infections or hematoma formation, necessitating vigilant post-operative surveillance.

Surgical procedures that necessitate the prone positioning of patients introduce additional difficulties in airway management, monitoring, and ventilation.

Most importantly, due to the intricate nature of Madelung's disease and its potential complications, comprehensive pre-operative counseling becomes imperative to manage patient expectations and address potential concerns effectively.

Conclusion

Anaesthesia management in a patient with severe Madelung's disease posted for surgery requires a systematic and well-coordinated approach. The utilization of awake fiberoptic intubation adds a layer of complexity and necessitates meticulous planning and execution. Through pre-operative assessment for associated comorbidities, adequate patient counseling, meticulous monitoring and planned airway management can be achieved while ensuring patient comfort and safety. Postoperatively, continuous monitoring, pain management, and follow-up contribute to a smooth recovery. The multidisciplinary collaboration between anesthesiologists, surgeons, and other specialists plays a key role in achieving optimal outcomes in patients with severe Madelung's disease undergoing surgical intervention.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the Journal. The patient understands that his/her name and initials will not be published, and due efforts will be made to conceal his/her identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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