

## Anesthesia in a Patient with Lady Windermere Syndrome

Varlik K.Erel\*, Güler S

<sup>1</sup>Department of Anaesthesiology and Reanimation, Adnan Menderes University Faculty of Medicine, Aydin, Turkey

<sup>2</sup>Anesthesiologist, Department of Anesthesiology and Reanimation, Ardahan Public Hospital, Kars, Turkey

### Abstract

Lady Windermere Syndrome (LWS), which was inspired by character of Oscar Wilde's the Lady Windermere's Fan novel, describes Mycobacterium Avium Complex (MAC) infection, following middle lobe, lingua bronchiectasis and painless, with high morbidity and mortality rate, especially in middle-aged women. Patients can undergo surgery by neurosurgery due to spinal deformities associated infection. In addition, due to some features of this syndrome, it is necessary to keep in mind during anesthesia and operation some complications, such as intubation difficulty, extreme bleeding, cardiac and respiratory complications and may occur as well. The patients have high risk of extubation and postoperative respiratory complications as they cannot easily cough or extract secretions. Anesthesiologists should be careful in terms of infectious, immune, respiratory, skeletal and cardiac system complications, because no literature data could be obtained about this syndrome's anesthesia management.

**Keywords:** Anesthesia; Lady Windermere Syndrome; Rotoscoliosis; Spine Surgery; Mycobacterium Avium

### Introduction

Lady Windermere Syndrome (LWS) defines the presence of Mycobacterium Avium Complex (MAC) infection in the lung middle lobe or lingus of non-immunosuppressive, non-smokers and, was first described by Reich and Johnson in six adult women in 1992. In this syndrome, which was inspired by character of Oscar Wilde's the Lady Windermere's Fan novel, these patients who suppress their coughs like Lady Windermere. It is overly rigorous, characterized painless MAC infection following middle lobe or lingua bronchiectasis in middle-aged women, with high morbidity and mortality rate [1]. Its prevalence is not known precisely and is usually observed in thin, white, middle-aged women [2]. LWS is more common in white women than in women of African or African-American background. It may be more common in Asian women too, and genetics may be a predisposing factor [2]. There is neither smoking nor an underlying pulmonary disease in the patient's history [1]. Patients suppress cough reflex as a result of this anatomical features and lack of collateral ventilation. It is thought to cause nidus for chronic inflammation and infection in the middle lobe with lingual secretions [3]. Voluntary suppression of coughing reflexes and psychological factors such as extreme rigorous may be responsible for the pathogenesis [3,4]. In the vast majority of these patients, pectusexcavatum, scoliosis, straight back, chest wall and vertebra disorders are observed [1,5]. Skeletal deformities are thought to have a role in infections and secretions. Connective tissue diseases can be accompanied by syndrome and mitral valve prolapse (MVP) is frequent [3]. This syndrome can be seen both in immune-suppressed individuals and normal population [6].

Oral and written informed consent was obtained for both anesthesia and publication during consultation by anesthetist.

### Case Description

A 65 year old, non-immunosuppressive, female patient (weight: 65kg, length: 165cm) who had low back pain while walking, was planned both laminectomy and posterior stabilization by neurosurgeon due to detected rotoscolyosis, lumber disc hernia (Figure 1). The patient being treated for LWS for one year, had history of migraines, asthma, and tuberculosis. There was no cigarette-smoking history. The patient had no finding other than lumber disc hernia operation a year ago. On physical examination, pulmonary sounds and cardiovascular system examination were normal. There was no clue or known connective tissue or cardiac disease and in the laboratory values, no abnormality except eosinopenia (0.3%). Increased infiltration was observed in the right anterior mid-lower zone of the chest X-ray (Figure 2). Computerized Tomography of the chest revealed

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#### \*Correspondence:

Erel Varlik K, Department of Anaesthesiology and Reanimation, Adnan Menderes University Faculty of Medicine 09100 Aytepe/Aydin, Turkey.

**E-mail:** varlik.erel@gmail.com

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Figure 1: Spine X-ray.



Figure 2: Lung X-ray.

middle lobe bronchiectasis on the right side (Figure 3). Respiratory function tests were normal. It was stated that the patient was scheduled to undergo general anesthesia. Possible risks were explained to the patient and her relatives and informed consent was approved.

Preoperatively, the patient had blood pressure of 140/80mmHg, body temperature of 36.3, pulse rate of 90 and Spo2 value of room air of 98. Induction of the anesthesia after appropriate preoxygenization was performed with propofol, 1mg/kg lidocaine, 1mg/kg fentanyl and 0.6mg/kg rocuronium at a dose of 2mg/kg without problems. There was no problem during intubation. Anesthesia was administered with 35% O<sub>2</sub> - 65% N<sub>2</sub>O and 2.5% sevoflurane. The patient, who had intra-operative blood loss of approximately 900 ml, was given 2U erythrocyte suspensions. There was no problem during the operation, but severe hypotension (70/40mmHg) was observed. Total of 4500cc fluid was delivered to the patient for three hours. At the end of the operation, the patient was awakened smoothly by muscle relaxant antagonist and delivered to intensive care unit.

## Conclusion

LWS is a rare MAC infection developed after the middle lobe or lingual bronchiectasis and affects other systems. LWS should be kept in mind as it can affect both gender and in terms of skeletal deformities and mitral valve prolapse. Patients often can undergo surgery due to lung infection or associated skeletal deformities. In addition, due to these features in this syndrome, it is obvious that anesthetists should be careful before, during and after anesthesia in terms of intubation difficulty, cardiac and respiratory complications. The patients have a high risk of extubation and postoperative respiratory complications as they cannot easily cough or extract secretions. We experienced no problem, probably due to successful long-term treatment of

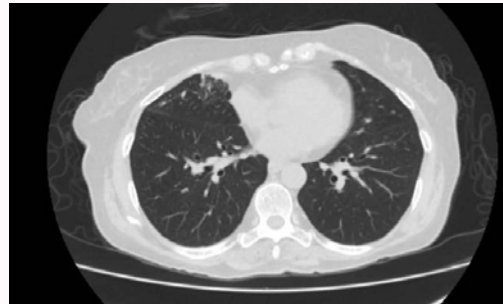


Figure 3: Lung Computerized Tomography.

tuberculosis. It is also important to protect the anesthesia device in the patient groups who have not been treated, who have active disease or whose treatment is ongoing. Eventually, anesthesiologists should be careful in terms of complications of the infectious, immune, respiratory, skeletal and cardiac system, because no literature data could be obtained about anesthesia management of this syndrome.

**Ethical approval:** All procedures performed in studies involving human participants were in accordance with the ethical standards of the Adnan Menderes University Research Committee and with the 1964 Helsinki declaration and its later amendment or comparable ethical standards.

## Statements

1. The work described has not been published previously (except in the form of an abstract or as part of a published lecture or academic thesis or as an electronic preprint, see; that it is not under consideration for publication elsewhere; that it does not duplicate or overlap other published work; and, if accepted, that it will not be published elsewhere in the same form, in English or in any other language, including electronically without the written consent of the copyright-holder. To verify originality, the article may be checked by the originality detection service CrossCheck.

2. They will take public responsibility for the contents, have contributed substantially to the drafting, and have approved the final version; and that publication is approved, tacitly or explicitly by the responsible authorities where the work was carried out.

3. For Research and Original Articles only, the lead author affirms that the manuscript is an honest, accurate, and transparent account of the study being reported; that no important aspects of the study have been omitted; and that any discrepancies from the study as planned (and, if relevant, registered) have been explained. The authors certify that they have disclosed any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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