CASE REPORT

ANESTHESIA FOR CESAREAN DELIVERY IN A PATIENT WITH LYMPHANGIOLEIOMYOMATOSIS: A CASE REPORT

ANESTESIA PARA CESARIANA EM PACIENTE COM LINFANGIOLEIOMIOMATOSE: RELATO DE CASO

Andresa Cavalcante Rodrigues[1/+], Jayme Marques dos Santos Neto[1], Raphaella Amanda Maria Leite Fernandes[2], Manoel Ananias da Silva Neto[3]

[1] MD, Anesthesiologist at the Hospital das Clínicas da Universidade Federal de Pernambuco-HC-UFPE, [2] MD. TSA-SBA. Anesthesiologist. Professor at the UFPE e da Faculdade de Medicina de Olinda-FMO, [3] Anesthesiologist

ABSTRACT

Introduction: Lymphangioleiomyomatosis (LAM) is a rare disease of unknown etiology, classically described in reproductive age women and, occasionally, reported in postmenopausal. Gestation in these patients is high risk, since the physiological changes of gestation add to those of LAM, worsening maternal cardiorespiratory status. **Case Report:** A 29-year-old pregnant woman with LAM and collapsing segmental and focal glomerulosclerosis (ESRF) diagnosed two years before had an indication of interruption of pregnancy during the 35th gestational week, due to worsening renal function. Spirometer examination revealed significant restrictive disorder. She was hospitalized during the 34th week of gestation and underwent steroid therapy for fetal lung maturation, when worsening of renal function and cesarean section were indicated. Combined regional anesthesia was chosen. **Comments:** LAM has a prevalence of 1 to 2.6 / 1,000,000 women. It is associated with Tuberous Sclerosis or idiopathic one.. Combined regional anesthesia was adopted because of the diffculty in managing the airway. Hormonal factors appear to play a role in the initiation and progression of LAM.

Keywords: Lymphangioleiomyomatosis. Obstetric anesthesia. Regional anesthesia.

RESUMO

Introdução: A linfangioleiomiomatose (LAM) é uma doença rara de etiologia desconhecida, classicamente descrita em mulheres em idade reprodutiva e, ocasionalmente, na pós-menopausa. A gestação nessas pacientes é de alto risco, pois as alterações fisiológicas somam-se às da LAM, agravando o status cardiorrespiratório materno. **Relato de caso:** Gestante, 29 anos, portadora de LAM e glomeruloesclerose segmentar e focal colapsante diagnosticadas há dois anos, teve indicação de interrupção da gestação no decorrer da 35ª semana por piora da função renal. Espirometria revelou importante distúrbio restritivo. Internada no curso da 34ª semana de gestação, submeteu-se a corticoterapia para a maturação pulmonar fetal, quando apresentou piora da função renal e foi indicada a cesárea. Optou-se pela anestesia regional combinada. **Comentários:** A prevalência da LAM em mulheres varia de 1 a 2,6/1.000.000 de mulheres e surge associada à esclerose tuberosa ou de forma idiopática. No caso, adotou-se a anestesia regional combinada em virtude da dificuldade no manejo da via aérea na gestante. Fatores hormonais parecem ter papel na iniciação e progressão da LAM.

Palavras-chave: Linfangioleiomiomatose; Anestesia obstétrica; Anestesia regional

INTRODUCTION

Lymphangioleiomyomatosis (LAM) is a rare disease of unknown etiology, often described in women of reproductive age and less common in postmenopausal women. This condition is characterized by the hamartomatous proliferation of smooth muscles in the bronchioles, arterioles, and pulmonary lymphatic vessels, leading to airway narrowing, obstruction, and air trapping. Associated alveolar damage results in the development of pulmonary cystic lesions and lymphatic vessels involvement (lymphangioleiomyomas) over time1,2. LAM may also be associated with an extrapulmonary lesion, such as a renal angiomyolipoma2.

Pregnancy in patients with LAM is a high-risk condition because of the physiological cardiorespi-

ratory changes of gestation, added to the existing structural alterations, and is poorly tolerated3.

This study is a case report of a patient with LAM who underwent a cesarean delivery under combined regional anesthesia (epidural and spinal).

CASE REPORT

A 29-year-old pregnant woman at 35 weeks of gestation was indicated for pregnancy termination due to clinical complications. At the age of 27, she was admitted to the nephrology department of the Hospital das Clínicas de Pernambuco, presenting with nephrotic syndrome and progressive worsening of renal function. A renal biopsy revealed collapsing focal segmental glomerulosclerosis. Her condition improved after treatment with prednisone (1 mg/kg) and furosemide. After one month, the patient experienced two episodes of pleuritic pain and sudden dyspnea secondary to pneumothorax, occurring 15 days apart and involving both lungs, as shown on chest X-ray. She underwent closed chest drainage, and due to recurrence, a computed tomography scan of the chest was performed, which showed multiple thin-walled cysts in both lungs and a clear left pneumothorax (Figure 1). Another drainage procedure was performed, and the pulmonary condition was further investigated. The previous medical history of the patient included the use of oral contraceptives for 10 years and intermittent asthma with the use of a beta-2 agonist inhaler about twice per month.

The patient underwent pleurodesis and left pulmonary segmentectomy by video-assisted thoracoscopic surgery for diagnostic biopsy in April 2012. Histopathological findings confirmed LAM, and a spirometry showed a substantial restrictive disorder.

At age 28, the patient became pregnant and remained under follow-up in the nephrology outpatient clinic, with stable renal function and a reduction of prednisone dosage (50 mg/day). An ultrasound of the urinary tract highlighted findings consistent with parenchymal nephropathy. At 34 weeks of pregnancy, the patient was admitted for corticosteroid therapy to accelerate fetal lung development. During this period, elevated nitrogenous waste levels were observed, and pregnancy termination was indicated.

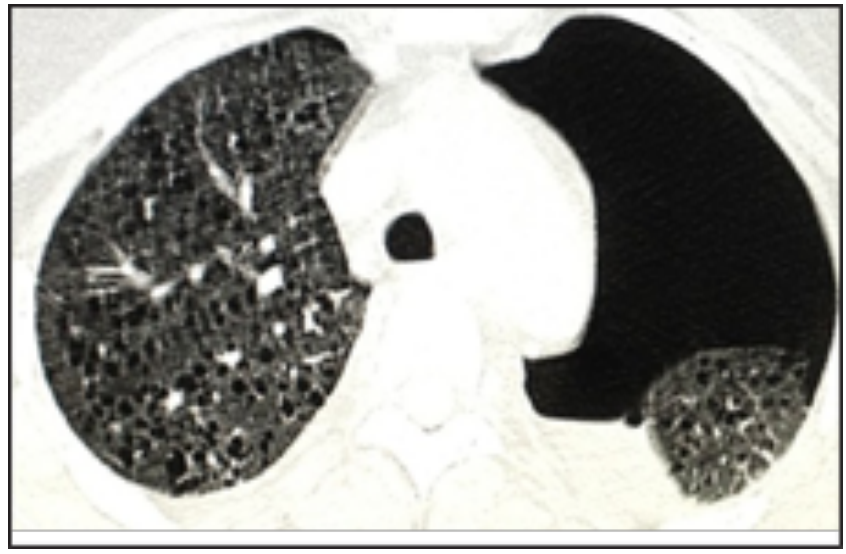

Figure 1. Computed tomography of the chest showing pneumothorax on the left and multiple cysts in both lungs.

During the pre-anesthetic evaluation, no findings were suggestive of a difficult airway, and the patient was classified as Mallampati class II. Preoperative arterial blood gas analysis showed pH = 7.37; pO2 = 93 mmHg; pCO2 = 29 mmHg; HCO3- = 19.3 mmol/L; BE = -7.3 mmol/L; SatO2 = 97%; and lactate = 1.2 mmol/L.

The anesthetic technique chosen was epidural at the T9-T10 interspace combined with spinal anesthesia at the L3-L4 level, with the administration of 8 mg of hyperbaric bupivacaine and 80 mcg of morphine into the subarachnoid space. Sensory block reached the T4 dermatome within three minutes. The surgery lasted 30 minutes without complications, and arterial hypotension was corrected with titrated doses of 50 mg of ephedrine and increased infusion of crystalloid solution. Postoperative arterial blood gas analysis showed pH = 7.29; pO2 = 124 mmHg; pCO2 = 32 mmHg; HCO3- = 17.1 mmol/L; BE = -9.6 mmol/L; SatO2 = 98%; and lactate = 2.1

mmol/L.

The newborn had an Apgar score of 7 at one minute and 8 at five minutes. After surgery, the patient was referred to the intensive care unit. No hemodynamic alterations or signs of respiratory distress were observed. The patient was discharged on the eighth postoperative day.

COMMENTS

LAM in women has a prevalence of 2.6 per 1,000,000. However, the limited awareness of the disease causes often misdiagnosis as asthma, chronic obstructive pulmonary disease, or bronchitis[1].

The most common symptoms are exertional dyspnea and pneumothorax. The latter is frequently recurrent, even in patients with a normal chest X-ray, with a reported incidence of 40% to 80% [1].

The patient presented with episodic dyspnea that has been diagnosed as intermittent asthma. However, after three spontaneous pneumothoraces (two on the left lung), the possibility of another primary pulmonary disease was considered.

The choice of the anesthetic technique must be guided by weighing risks and benefits for each patient. In this case, the severe pulmonary condition and the physiological changes of pregnancy were considered. The anesthetic blockade at a high spinal level was concerning, as it could hinder the lung function of a patient who was already dependent on accessory muscles. General anesthesia was also considered, as it offers more precise ventilatory control regardless of patient effort. Nevertheless, airway management in pregnant patients is challenging due to worsening Mallampati classification, soft tissue edema (e.g., pharynx and larynx), friable mucosa, reduced glottic area, and enlarged breasts[4].

A combined blockade technique was chosen to allow the administration of low doses of anesthetic into the subarachnoid space and the insertion of an epidural catheter for titration. The aim was to avoid airway manipulation with positive-pressure ventilation, given the substantial pulmonary restrictive disease and multiple cysts of the patient, which increased her susceptibility to pneumothorax.

Hormonal factors may contribute to the initiation and progression of LAM, which is more prevalent in women, as hormone receptors are found in some LAM cells. Exogenous estrogen use and pregnancy may worsen the disease[5,6]. In this case, a possible risk factor for the patient was the 10-year use of oral contraceptives. These patients should be advised against pregnancy due to maternal and fetal risks[5-7]. A study by Urban et al. (1999)8 described that 23% of patients presented their first pulmonary event during pregnancy, although only two showed clinical worsening related to pregnancy.

REFERENCES

1. Hohman DW, Noghrehkar D, Ratnayake S. Lymphangioleiomyomatosis: A review. Europ J of Inter Medic. 2008; 19(5): 319-24
2. Zhan Y, Shen L, Xu W, Wu X, Zhang W, Ang J, Li X, Yang Y, Tian X, Xu KF. Functional improvements in patients with lymphangioleiomyomatosis after sirolimus: an observational study. Orphanet J Rare Dis. 2018;13(1):34.
3. Iserte PP, Nadal JS, Folch BF, Ferrero IS, Almela VD, Marín AP. Linfangioleiomiomatosis pulmonar y gestación. Prog Obstet Ginecol. 2013; 56(1): 29-31
4. Boutonnet M, Faitot V, Salomon L, Keita H, Katz A. Mallampati class changes during pregnancy, labour, and after delivery: can these be predicted? Br J Anaesth. 2010;104(1):67-70.
5. Pais F, Fayed M, Evans T. Lymphangioleiomyomatosis: an explosive presentation of a rare disease. Oxford Medic Case Repor. 2017;2017(6): 92-4
6. Lu C, Lee HS, Pappas GP, Dilling DF, Burger CD, Shaifren A, Veeraraghavan S, Chapman JT, Paranbil J, Ruoss SJ, Yong LR, Hamm SR, Kopas EJ, Roabs T, Krischer JP, McCormack FX; Trial of and Aromatase Inhibitor in lymphangioleiomyomatosis Group. A Phase II Clinical Trial of an Aromatase Inhibitor for post menopausal women with lymphangio-leiomyomatosis. Ann Am Thorac Soc 2017;14(6):919-28.
7. Harari S, Casandro R, Chiodiny J, Taveira-Da Silva AM, Moss J. Effect of a gonadotrophin-releasing hormone analogue on lung function in lymphangioleiomyomatosis. Chest. 2008; 133(2): 448-54.
8. Urban T, Lazor R, Lacronique J, Murris M, Labrune S, Valeyre D, Cordir J F. Pulmonary lymphangioleiomyomatosis – a study of 69 patients. Medicine. 1999;78(5): 321-27.