Pregnancy complicated with pulmonary lymphangioleiomyomatosis: Case report

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Summary

A case of a 30-year-old primiparous woman with pulmonary lymphangioleiomyomatosis is described. The patient had experienced six episodes of spontaneous pneumothorax at the age of 27 years and had been diagnosed as having pulmonary lymphangioleiomyomatosis based on histological findings of specimens obtained by transbronchial biopsy. She had undertaken open lung surgery and thoracoscopy. Thereafter, she became pregnant spontaneously. Her antenatal course was uneventful with no exacerbation of respiratory status. At 38 weeks of gestation, she underwent a selective cesarean section and myomectomy under combined spinal and epidural anesthesia. Her postoperative course was uneventful. No remarkable changes in computed tomographic findings of the lung were noted on the 20th day of postoperation compared with those before pregnancy. She has been followed-up in the pulmonary outpatient clinic with no deterioration of the disease.

Key words: Anesthesia; Pulmonary lymphangioleiomyomatosis; Pregnancy.

Introduction

Lymphangioleiomyomatosis (LAM) is an uncommon disease that primarily affects women in the reproductive ages [1]. LAM is characterized by diffuse interstitial proliferation of bundles of immature smooth muscle cells in the wall of air cavities [2]. LAM can occur in association with tuberous sclerosis complex (TSC) or without evidence of other disease (sporadic LAM) [3]. A recent study has demonstrated the role of somatic mutations in the TSC2 gene in the pathogenesis of sporadic LAM [4]. The clinical manifestations include exertional dyspnea, recurrent pneumothorax, cough, hemoptysis, and chylothorax [2]. Renal angiomyolipomas and retroperitoneal lymphangiomas can occur in patients with LAM [2]. The clinical course of LAM is slowly progressive, but eventually leads to respiratory failure and death [2]. LAM is believed to be sex steroid hormone-related disorder because exogenous estrogen [5] and pregnancy [6-12] have been reported to exacerbate pulmonary LAM. In addition, some LAM cells are shown to express estrogen and progesterone receptor, and these receptors are down-regulated by hormonal therapy [13]. The optimal treatment of LAM during pregnancy has not been established. We describe a case of pulmonary LAM in a 30-year-old primiparous woman.

Case report

The patient was a 30-year-old primiparous woman with a medical history of pulmonary LAM. She had experienced six episodes of bilateral spontaneous pneumothorax at the age of 27 years. At that time, a computed tomography of the lung had shown diffuse distribution of multiple well-defined, thin-walled cysts throughout both lungs. She had been diagnosed as having pulmonary LAM based on the histological findings of specimens obtained by transbronchial biopsy. LAM cells were found to be negative for estrogen and progesterone receptors immunohistochemically. She had undertaken open lung surgery and thoracoscopy at the age of 27 and 28 years, respectively.

Thereafter, she became pregnant spontaneously. Her antenatal course was uneventful with no exacerbation of respiratory status. Mode of delivery was discussed with the patient at 36 weeks of gestation. Considering the risk of spontaneous pneumothorax during labor, she desired a cesarean section. In consultation with an anesthetist, we decided to perform surgery under combined spinal and epidural anesthesia. She was admitted to the hospital at 38 weeks of gestation. On admission, respiratory sounds were normal with no rales. Chest X-ray did not show any signs of diffuse reticular shadows or pleural effusion. The spirometric tests were normal with a vital capacity (VC) of 2.70 l, %VC of 92.4%, a forced expiratory volume in one second ($FEV_{1.0}$) of 2.13 l, and % $FEV_{1.0}$ of 83.2%. SpO_2 was 97% in room air. The following day she underwent a cesarean section and myomectomy under combined spinal and epidural anesthesia. She delivered a male baby with a birth weight of 2,812 g. Apgar scores were nine at one minute and nine at five minutes, respectively. Pathological examination showed leiomyoma with no findings of LAM. Her postoperative course was uneventful. No remarkable changes in computed tomographic findings of the lung were noted on the 20th day of postoperation compared with those before pregnancy. She has been followed-up in the pulmonary outpatient clinic with no deterioration of the disease.

Discussion

Several authors [6-12] have demonstrated an association of pregnancy with an increased risk of complications in patients with LAM. It has been reported that an exacerbation of LAM was observed in 14% of 69 cases during pregnancy [8] and that the overall incidence of complications in patients with LAM was 11 times higher during pregnancy than at other times [7]. The complications

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during pregnancy include pneumothorax [7, 10, 11], chylothorax [7, 12], mediastinal LAM and chylothorax [9], and worsening renal function secondary to angiomyolipomas [6].

On the other hand, Yigla *et al.* [14] reported a case of LAM in whom the disease did not deteriorate to respiratory failure despite the experience of multiple pregnancies. In our patient, the respiratory function remained normal during pregnancy. The reason for no exacerbation of LAM during pregnancy in our patient is unknown. However, it seems likely that the absence of estrogen and progesterone receptors in LAM cells may be attributable to the unresponsiveness to sex steroid hormones excessively secreted during pregnancy.

Mode of pregnancy should be dictated by obstetric considerations and the medical condition of the patient with LAM [15]. The risk of pneumothorax may be increased during labor because intraalveolar and intrathoracic pressure rises along with uterine contractions. In our patient, a cesarean section was performed under combined spinal and epidural anesthesia with no respiratory complications. McLoughlin *et al.* [15] suggest that epidural anesthesia be offered for labor to reduce hyperventilation and excessive changes in intrathoracic pressure during contractions. In contrast, general anesthesia with intermittent positive pressure ventilation for cesarean section constitutes a risk for pneumothorax [16].

In conclusion, the present case with pulmonary LAM had an uneventful course during pregnancy and postpartum with no recurrence of pneumothorax. Careful monitoring for respiratory function is mandatory to avoid LAM-related complications during pregnancy.

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