Adult congenital lobar emphysema in pregnancy

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Abstract
A young woman presented with left sided chest pain. Chest radiography revealed a hyperexpanded left upper lobe and the rare diagnosis of congenital lobar emphysema was made. She was then found to be pregnant. Thoracotomy and left upper lobectomy were performed during the pregnancy without adverse effects to the mother or fetus. The implications of pregnancy on the surgical management of this rare condition are discussed.

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Keywords: congenital lobar emphysema, pregnancy.

An 18 year old woman presented to her family doctor with left sided chest pain. This followed a minor coughing episode, although inhalation was denied. She had previously been fit and well. The chest radiograph revealed hyperexpansion of the left upper lobe with mediastinal shift (fig 1). Computed tomographic scanning showed an emphysematous left upper lobe (fig 2). Respiratory function tests revealed a forced expiratory volume in one second (FEV₁) of 1.71 and a forced vital capacity (FVC) of 2.11, both 50% of her predicted values. She was therefore referred to our thoracic unit for bronchoscopy in view of the possibility of an obstructing foreign body. At this stage she was found to be six weeks into her first pregnancy. Subsequent bronchoscopy failed to show an obstructing endobronchial lesion and diagnosis of congenital lobar emphysema was made. As the pregnancy progressed she became increasingly dyspnoeic and the chest radiograph showed further mediastinal shift.

At 26 weeks gestation a thoracotomy and left upper lobectomy were performed. The operative procedure consisted of a standard
posterirolateral approach to the left hemithorax. A grossly emphysematous upper lobe was resected; the remaining lung tissue was macroscopically normal. She made a routine postoperative recovery and 13 days later was discharged home.

Follow up at six weeks was satisfactory. The chest radiograph showed resolution of the mediastinal shift. Respiratory function tests showed an improvement in both FEV₁ to 2.51 and FVC to 3.01. The patient went on to deliver a healthy full term baby without difficulty. At one year both mother and baby were well.

Histological examination of the resected lobe showed widely distended alveolar air spaces with minimal destruction of lung tissue. This was consistent with the clinical and radiological diagnosis of congenital lobar emphysema.

**Discussion**

Congenital lobar emphysema is an uncommon bronchopulmonary malformation which nearly always presents in the first months of life. Most cases affect the left upper lobe. Symptoms usually include respiratory distress and/or recurrent pulmonary infections. In congenital lobar emphysema lobectomy is usually the treatment of choice. Respiratory function is expected to improve, especially when mediastinal shift has occurred.

There are few reported cases presented in adult life. This case is unusual in that, upon detection, pregnancy was diagnosed, so the management decisions become more complicated because of risks to both mother and fetus.

The optimal timing of surgery is controversial. Intrapartum general anaesthesia and thoracotomy are well tolerated without excessive risk to the fetus, provided they are performed after the period during which organogenesis occurs — namely, the first trimester. In the third trimester there is increased cardiopulmonary demand as the fetus enlarges. The exact timing of thoracotomy should, of course, be dictated by the patient's clinical course. In this case the elected time of operation was towards the end of the second trimester when the risks to both mother and fetus were thought to be least. This coincided with the progressive dyspnoea and worsening chest radiograph.

In conclusion, the optimal time to operate on pregnant patients with congenital lobar emphysema is currently uncertain. We have presented a case of an adult undergoing left thoracotomy and pulmonary resection at 26 weeks into her pregnancy with a satisfactory outcome for both mother and fetus.

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