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 hyper-expansion 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

only when the patient is cooperative and has no bleeding diathesis. It is usually an easy, efficient and safe procedure.

Mediastinal amyloidosis occurs infrequently and may be confused with other tumours more usually found in this region. The diagnosis should be considered in the evaluation of a large mediastinal mass. Percutaneous needle biopsy may be sufficient to provide a definitive diagnosis.

posterolateral 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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