Spontaneous rupture of the diaphragm

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Abstract:

A 2.5 year old girl with metachromatic leukodystrophy presented with acute respiratory distress and was initially wrongly diagnosed with pneumothorax. Barium meal showed bowel loops in the left hemithorax, which prompted surgical intervention; spontaneous rupture of the diaphragm was diagnosed at surgery.

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Spontaneous rupture of the diaphragm, which is often confused with a pneumothorax, is seen mainly in adults as a result of an increase in the abdominothoracic pressure gradient, such as during physical exertion, parturition, and coughing. It has previously been reported in only two children, both with pertussis.^{1 2}

Case report

A 2.5 year old girl with profound neurodevelopmental delay secondary to metachromatic leukodystrophy, presented to the emergency department with sudden onset of dyspnoea, floppiness, and pallor. A chest x ray two months previously had been normal. There was no history of trauma or preceding respiratory symptoms. Figure 1 shows the chest x ray at this presentation. There was immediate improvement after needling the left chest and aspiration of 120 ml of air. A subsequent x ray suggested a multiloculated air and fluid collection, which was aspirated under ultrasound guidance and yielded 140 ml of "pus". Microscopy showed many different organisms and pus cells. An intercostal drain was inserted and intravenous fluids and antibiotics administered. The drain was removed after three days as no fluid was drained.

Figure 1 Chest x ray at presentation.

The child continued to have recurrent episodes of respiratory distress. Computed tomography of the chest appeared to show a large hydropneumothorax in the left hemithorax with mediastinal shift to the right and medial collapse of the left lung. As these episodes were associated with enteral feeding, a barium meal was performed, which showed diaphragmatic hernia. Microbiological culture of the original fluid aspirate grew mixed organisms.

At surgery the whole of stomach, greater omentum, and spleen were lying in the left hemithorax. There was a 6 cm linear tear in the diaphragm extending anterolaterally towards the costophrenic angle. There was no pus or inflammation in the left hemithorax and the lungs were entirely normal with no suggestion of pneumonia. The contents of the left hemithorax were reduced and the diaphragmatic tear repaired. Oesophagoscopy and gastroscopy did not reveal any perforations. The postoperative period was uneventful with a very rapid recovery. She was discharged home a week later.

Discussion

There are no pathognomonic symptoms or signs of spontaneous rupture of the diaphragm and diagnosis is often delayed. The non-specific symptoms are nausea and vomiting, epigastric pain, and dyspnoea. Bisgaard *et al* reported an adult case with a four month delay in diagnosis.³ A chest x ray may be difficult to interpret as it can easily be confused with pneumothorax, pleural effusion or pneumatocele.

The presenting chest x ray in our patient was thought to show a left sided tension pneumothorax. Pneumothorax is uncommon in childhood but may result from trauma or in association with mechanical ventilation, asthma, and cystic fibrosis. The incidence is relatively high in infants with staphylococcal pneumonia. Spontaneous pneumothorax may occur in teenagers on exertion with Valsalva manoeuvre and is more common in patients with connective tissue disorders. Our patient had none of these features.

A retrospective examination of the presenting x ray showed the left upper lobe compressed upwards with no discernible left hemidiaphragm (fig 1). In the presence of a pneumothorax, the lung tends to collapse medially towards the hilum and the diaphragm is conspicuous. After needling the "pneumothorax" the left upper lobe did partially expand but more of the gut was seen in the left hemithorax. This was falsely interpreted as a multiloculated air and fluid collection that was aspirated. The microscopy and culture result should have alerted us to the possibility of the fluid being of gastric origin. Gastric perforation

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has previously been reported in three cases as a result of thoracentesis.^{4,5} Fortunately, the initial needling, fluid aspiration, and chest drain insertion in our case did not cause a gastric leak or perforation. Barium meal showed bowel loops in the left hemithorax and prompted surgical intervention. Computed tomography of the chest was unhelpful as the findings were misinterpreted. The final diagnosis in our case was made on thoracotomy. We did not identify a contributory factor in our patient; an association with metachromatic leukodystrophy is unknown.

In summary, the diagnosis of pneumothorax should not be too readily accepted in the absence of an explanation. Spontaneous rupture of the diaphragm in children is very rare

but should be considered in such circumstances. A barium meal will lead to the correct diagnosis but a plain chest x ray with a nasogastric tube in situ may alert one to the possibility of this diagnosis.

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Vagal nerve stimulation for epilepsy

Children with intractable epilepsy are a therapeutic challenge. In meeting that challenge it is important to have constantly in mind what it is hoped to achieve. The aim must be significant improvements in quality of life for the child and the family. Fairly minor reductions in seizure frequency may not achieve that aim.

By mechanisms that are not known, stimulation of the left vagus nerve reduces seizure frequency in experimental animals. Studies in adults have led to this procedure being approved in the USA for patients over the age of 12. Now the paediatric data (19 patients) from two multinational trials, which mostly contained adults, have been reported together with data on 41 children treated in an uncontrolled fashion (Jerome V Murphy and the Pediatric VNS Study group. *Journal of Pediatrics* 1999;134:563–6; see also editorial, *Ibid*: 532–3). A stimulator is implanted under the skin of the chest wall with an electrode on the left vagus nerve in the neck. Standard practice is 30 seconds on and 5 minutes off continuously. In children aged 3–18 with various types of seizure there was a reduction in seizure frequency by 23% at 3 months, rising to 42% by 18 months. No correlation was found between seizure type or cause and response. In some patients stimulation caused cough, fever, headache, or voice change. The device eroded through the skin in one patient.

This report serves to emphasise the importance of paediatric trials, performed on children and designed to provide the answers that patients, their families, and their doctors want and need. It does not answer the most important question; were they better off for the intervention? It seems unclear whether vagal stimulation might provide real benefit for some children or whether, like the cerebellar stimulation briefly in vogue some 25 years or so ago, it will prove just another fad.

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