Intrathoracic cystic hygroma with sudden respiratory distress mimicking pneumonia

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Abstract

Benign cystic lesions such as cystic hygroma commonly manifest as progressively increasing swelling in the neck with or without compression effects. Rarely, they present with sudden respiratory distress in instances such as infection or haematoma resulting in a sudden increase in the size of the tumour. We present a seven month old child with sudden onset respiratory distress without any obvious neck swelling. The chest X ray findings correlated with the history and were suggestive of right upper lobe pneumonia that leads to a wrong diagnosis of aspiration pneumonia. However, presence of a deviated trachea in the neck raised a suspicion of possible mass. Computed tomogram showed a large cystic mass in the right upper mediastinum with tracheal collapse. We caution intensivists and paediatricians that sudden respiratory distress in infants in the absence of obvious neck swelling does not rule out possibility of intrathoracic tumour.

Keywords: Cystic hygroma, intrathoracic tumour, pneumonia, respiratory distress

Introduction

Cystic hygroma is a benign tumour that mostly manifests in the first two years of life as a progressively increasing neck mass with or without compression effects on the trachea or oesophagus. However, rapid increase in the size of the previously small and asymptomatic tumour may contribute to sudden onset of respiratory distress and stridor. Patients in these reports were associated with an obvious visible neck swelling. In this article, we present cystic hygroma in an infant that manifested with sudden respiratory distress and stridor without a visible neck swelling that was mistakenly diagnosed and initially treated as aspiration pneumonia.

Case Report

A seven month old infant was brought to the emergency room with suspected aspiration following breast feeding. The child was cyanosed and in severe respiratory distress resulting in respiratory arrest within minutes of hospitalisation. Child’s trachea was intubated with a 4.0mm ID uncuffed tube and mechanical ventilation instituted. Bilateral rhonchi were auscultated and bronchodilator therapy was initiated. Past history of the child was unremarkable with term vaginal delivery, uncomplicated perinatal course, the milestones and vaccination were appropriate for age. The child was asymptomatic until this event.

A chest X ray (CXR) and arterial blood gas (ABG) were obtained. The ABG revealed combined respiratory and metabolic acidosis that was corrected with adjustments in ventilator settings and intravenous fluids. The CXR revealed a picture suggestive of right upper lobe pneumonia [Figure 1] for which antibiotics were started. However, intermittent rhonchi persisted for > 48h despite multiple bronchodilator therapy. There was no evidence of severe infection such as fever or increasing leucocyte counts. The child intermittently had disappearance of rhonchi at which time, we could easily wean to low pressure support ventilation. A trial of extubation resulted in rapid deterioration
in the respiratory condition necessitating emergent re-intubation. In view of recurring rhonchi, unresolving pneumonia like radiograph picture and failed extubation, opinion of the intensivists was sought. A close look at the CXR revealed that the trachea was considerably deviated to left side of the neck [Figure 1]. In the light of these findings, a computed tomogram of the neck and chest was obtained that confirmed presence of a mass in the right upper thorax the upper limit of which was extending to the lower neck. The tumour was found not only to cause a shift of the trachea, but also contributed to lateral compression with the narrowest portion of the trachea being about half a cm above the carina [Figure 2].

Subsequently, the tracheal tube was deliberately made endobronchial and then withdrawn gently till breath sounds were audible bilaterally. This was done to ensure the tracheal tube tip bypassed the narrowest portion of the trachea following which the rhonchi disappeared. Next day, the child underwent surgery. The cyst was approached through an incision at the right neck base, adhesions were removed, fluid was aspirated and the cyst was excised. The pathology report confirmed it to be a benign cystic hygroma. In view of possible tracheomalacia, the trachea was extubated 48h later under fibreoptic evaluation which confirmed absence of tracheal wall collapse at extubation. The child remained asymptomatic post-extubation and is doing well at one year follow up.

Discussion

Cystic hygroma is any cystic mass lined by endothelium. Inability of the peripheral lymph spaces to join the central drainage system is supposed to be the mechanism.²,¹⁴ Gradual accumulation of fluid in these tiny closed spaces contributes to further growth. As they grow, competition for space in the neck and thorax may result in compression of the surrounding structures such as the trachea and the oesophagus. Therefore, they often present as painless swelling in the neck that gradually increase in size and may subsequently manifest with progressively increasing difficulty in breathing or swallowing. However, rarely they may also present as sudden respiratory distress secondary to infection or haemorrhage into a pre-existent asymptomatic cyst.¹¹-¹⁵ In our case, sudden respiratory distress was not associated with a visible neck swelling as the mass was mostly intrathoracic. It appears that the intrathoracic part of the trachea was continuously being compressed by this mass which probably resulted in thinning of the right side of the tracheal wall that was in contact with the tumour. However, the child remained asymptomatic since the tumour was supporting the tracheal wall. Possible aspiration during breast feed must have resulted in development of significant negative intrathoracic pressures during coughing attempts contributing to collapse of the thinned out tracheal wall which was followed by stridor and respiratory distress which appears to be the mechanism for sudden respiratory distress in our case. Persistent rhonchi were pointers to possible collapse of the tracheal lumen distal to the tracheal tube tip as evidenced by their disappearance after repositioning of the tube beyond obstruction. Although the CXR finding correlated well with the history suggestive of aspiration pneumonia, failure to notice the deviation of trachea and the tracheal tube lead to delay in establishing the correct diagnosis and prolongation of mechanical ventilation.

Intrathoracic cystic hygroma without visible swelling in the neck is extremely rare. Such tumours usually present several months to years after birth following gradual expansion in their size. They may be identified as incidentalomas during radiological screening or may present with compressive manifestations.²,¹⁵ Our
case shows that intrathoracic cystic hygroma may present with sudden respiratory distress in a previously asymptomatic child. This is because a softened tracheal wall (as a result of compression by the tumour) is susceptible to sudden collapse when it is subjected to significant negative intrathoracic pressures. It is easy to misdiagnose such condition as pneumonia as was in this case.

Conclusions

In a child with sudden onset respiratory distress, absence of a visible neck swelling does not rule out intrathoracic mass as a contributing factor. Persistent rhonchi and respiratory distress in children despite institution of appropriate measures such as invasive mechanical ventilation, bronchodilator therapy and antibiotic therapy must raise suspicion of tracheal collapse. In such cases, even when the CXR findings are well substantiated by the history suggestive of aspiration pneumonia, further evaluation for possible underlying tumour must be considered.