SAGE Open Medical Case Reports
2: 2050313X14553864

DOI: 10.1177/2050313X14553864
sco.sagepub.com

All that wheezes: A young infant with a mediastinal mass

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Abstract

Wheezing infants are frequently encountered in the emergency department. Bronchiolitis is the most commonly seen cause. Radiographs are not recommended in the routine management of bronchiolitis. We present the case of a young wheezing infant with a mildly abnormal chest x-ray whose cystic hygroma caused life-threatening respiratory distress soon after he was admitted to the hospital.

Keywords

Cystic hygroma, lymphangioma, mediastinal mass, bronchiolitis

Date received: 11 June 2014; accepted: 4 September 2014

Introduction

Bronchiolitis is "a constellation of clinical symptoms and signs including a viral upper respiratory prodrome followed by increased respiratory effort and wheezing in children less than 2 years of age."[1] Wheezing in infancy may, however, have other causes. We present the case of a 5-week-old infant who presented with wheezing and respiratory distress, rapidly progressive after admission with an ultimate diagnosis of cervicomediastinal cystic hygroma.

Case

A 41-day-old male infant presented to the emergency department (ED) via ambulance from his physician's office. He was taken to the doctor's office for cough, fussiness, and decreased feeding since the previous day, accompanied by wheezing for several hours. There had been no fever. At the office, he received an albuterol treatment for respiratory distress with oxygen saturations of 88%–89% in room air on presentation. He was given another albuterol treatment by emergency medical services (EMS). In the ED, he was noted to be in moderate respiratory distress with retractions, wheezing, and a respiratory rate of 44 breaths per minute. No masses were noted on neck or chest examination. He was given nebulized racemic epinephrine with some improvement. His venous pH was 7.25, and his pCO2 was 43 mm Hg. Initial hemoglobin was 9.3 g/dL and hematocrit 26.9%. Rapid tests for respiratory syncytial virus (RSV) and influenza were negative. Since respiratory distress continued, arrangements were made to admit him to the pediatric intensive care unit (PICU). He developed a fever of 100.6°F just prior to transfer. A chest radiograph was suspicious for a mediastinal mass with mass effect on the trachea, displacing it to the left (Figure 1).

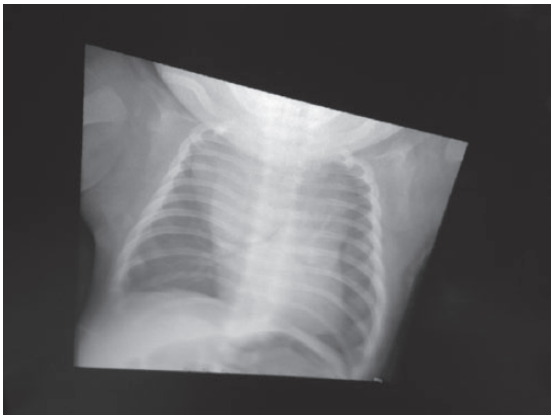

Figure 1. Radiograph obtained in the emergency department was concerning for a mediastinal mass.

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Past medical history was remarkable for a 9 lb 8 oz birth weight after a 36-week gestation to a diabetic mother. He was hospitalized for 3 weeks after delivery for rapid breathing not otherwise classified in his discharge summary from an outside hospital. The mother reported he had done well at home until the previous day.

On admission to the PICU, he had moderate respiratory distress, decreased air entry, bilateral expiratory wheezing, and stridor with agitation. He was continued on nebulized racemic epinephrine every 2 h. The computerized tomography (CT) scan of his chest revealed a large infiltrative mass involving the right cervical region, prevertebral space, and anterior and middle mediastinum. Narrowing of the trachea with leftward deviation was noted from the level of the arch to the carina, with the cervical trachea deviated anteriorly. The mass encased the right carotid and subclavian arteries and innominate vein, with displacement of the superior vena cava into the right chest. The right internal jugular vein was not visualized. An ultrasound confirmed that the mass was cystic. Over the next 12 h, he developed respiratory failure, tachypnea, and an increasing oxygen requirement. His capillary blood gas showed a pH of 6.9 and a pCO2 of 89.3 mm Hg. His hemoglobin had decreased to 6.5 g/dL. He was taken emergently to the operating room (OR) with a surgical team that included otolaryngology (ENT), pediatric surgery, and cardiothoracic surgery. The extracorporeal membrane oxygenation (ECMO) team was on standby with a primed circuit. Airway evaluation by ENT included bronchoscopy and intubation to the carina with a 3.0 mm endotracheal tube (ETT). His airway had extrinsic compression on the right. Following initial observation in the OR with airway pressures remaining markedly elevated, the decision was made to proceed with mediastinal decompression. The child underwent resection of the mass via a midline sternotomy with dissection around multiple blood vessels. Phrenic and vagal nerves were spared. A significant amount of blood was contained within the cystic tissue. A small amount of hygroma remained in the right posterior neck at the end of the procedure, and he was easily reintubated with a 3.5 mm cuffed ETT, with markedly improved airway pressures. The mediastinal tube was removed on postoperative day (POD) #1, and he remained intubated for 48 h, followed by elective extubation in the OR. Mild respiratory distress on POD #3 resolved quickly with high flow nasal cannula. He was discharged on POD #7 with normal breathing on room air, and tolerating full oral feedings. He had a magnetic resonance imaging (MRI) 5 months after his surgery which revealed small areas of residual lymphatic malformation in the right neck and mediastinum. All areas were less than 2 cm, and there was no mass effect. In the setting of scattered remnants, it is likely the remaining areas had disruption/bleeding/fibrosis sufficient to render them harmless. The patient himself was described as happy and healthy. He had grown well. He did have persistent stridor related to laryngomalacia.

The Organizational Ethics Committee stated that approval by the CHRISTUS Santa Rosa Ethics Committee and patient/guardian consent are not required to report individual cases. The mother did give written consent to report this case.

Discussion

Infants presenting to the ED with wheezing in the fall and winter frequently have bronchiolitis. These babies commonly have poor response to bronchodilators. They may require admission for supportive care including oxygen and nutritional support. Our patient had no focal findings on his pulmonary examination, but his continued respiratory distress, negative RSV test, and absence of significant rhinorrhea led to a chest radiograph being ordered.

Chest radiographs are not routinely recommended by the American Academy of Pediatrics (AAP) for the diagnosis and management of bronchiolitis.[2] A study of children 2–23 months of age with bronchiolitis showed that only 2/235 patients had x-ray findings inconsistent with bronchiolitis. These findings did not change management except for a tendency to prescribe more antibiotics. This treatment was probably not indicated.[3]

Besides bronchiolitis, the differential diagnosis of wheezing in infancy includes other respiratory infections, laryngotracheomalacia, foreign body (esophageal or respiratory tract), gastroesophageal reflux, congestive heart failure, vascular ring, allergic reaction, cystic fibrosis, mediastinal mass, bronchogenic cyst, and tracheoesophageal fistula.[2] Mediastinal masses in neonates include lymphangioma, teratoma, thymic enlargement, lymphadenopathy, and lipoma or lipothymoma.[4] Review of 320 children with mediastinal cysts and tumors yielded only 9 with lymphangiomas.[5]

Cystic hygromas or lymphangiomas are congenital malformations of the lymphatic system characterized by single or multiple fluid-filled lesions that occur at sites of lymphatic–venous connection, especially in the posterior neck (90%).[6] Cervical lymphangiomas may extend into the mediastinum. Gallagher and Mahoney[6] reported that up to 10% of neck lymphangiomas extended to the chest or mediastinum, but Glasson and Taylor[7] found this only 4% of the time. Glasson had no patients with purely mediastinal lymphangiomas, while Alqahtani et al.[8] found this in 2/186 patients over a 25-year period. All of Glasson's patients with cervical or cervicomediastinal cystic hygromas had a mass present in the neck.[7] Ninh and Ninh reported 126 pediatric cases of cystic hygroma (none cervical), noting that 16/126 cases had hemorrhage found at surgery. They noted that cystic hygromas rarely caused difficulty breathing, identifying only two cases of mild respiratory distress.[9]

Airway compromise has been reported with both neck and mediastinal lymphangiomas. Glasson reported respiratory symptoms in 11/52 patients with cervical or cervicomediastinal lymphangiomas. Those symptoms caused by mediastinal extensions were usually less dramatic. In his series, the most severe presentations were in patients with infiltrative lymphangioma involving the neck, tongue, pharynx, epiglottis, and larynx which presented as neonatal emergencies.[7]

Cystic hygromas rarely regress. Surgical excision is the treatment of choice. Rapid enlargement may occur from hemorrhage, trauma, or infection. Aspiration may be performed for

emergency decompression. Recurrences after surgical excision have been reported in 5%–15% of cases, but seem less frequent in internal lymphangiomas (including thorax).[8–10]

Cystic hygromas may be detected on prenatal ultrasound. Up to 60% of these fetuses have abnormal chromosomes, most commonly 45,X or trisomy 21. Karyotyping should be done when a fetus has a cystic hygroma, especially a posterior nuchal one. These pregnancies may be complicated by hydrops and fetal loss. Patients with the diagnosis of cystic hygroma after 30 weeks gestation have a good prognosis, and the karyotype of the fetus is usually normal.[6]

Pham et al. reported an infant who was noted to have a cystic mass adjacent to the right atrium at 20 weeks gestation. The lesion regressed on subsequent ultrasounds. Echocardiogram and chest radiograph were normal at birth. At 2 months of age, he had acute respiratory distress with a right subpulmonic fluid collection. This was found to be a lymphangioma which was successfully resected.[11] Muraskas et al. had an infant with a mediastinal cystic hygroma noted at 22 weeks gestation. An in utero decompression was performed at 24 weeks due to hydrops with cardiac compromise and mediastinal shift. The baby was delivered at 37 weeks without respiratory distress. Excision of a large thoracic cystic hygroma was performed shortly thereafter. At 19 months, unilateral wheezing developed. Surgery confirmed a recurrent cystic hygroma.[12] In 1970, Bratu et al. reported a 2-month-old girl with respiratory distress and anterior mediastinal opacity on chest radiograph that was thought to be a staphylococcal pneumonia. She was treated with thoracentesis three times and discharged from the hospital with a diagnosis of possible mediastinal cyst. She was readmitted with respiratory distress 2 weeks later. The cystic hygroma was surgically excised during that admission. The chest radiograph was reported as normal 1 year later.[13]

Our patient had respiratory distress initially felt to be bronchiolitis. A chest radiograph concerning for mediastinal mass led to the diagnosis of lymphangioma by CT and ultrasound. This patient's rapid deterioration was secondary to hemorrhage into the lymphatic malformation. Clinically, he progressed over a 12-h period from mild–moderate respiratory distress to acute respiratory failure. He underwent surgical resection and was discharged from the hospital in good condition. Especially in the midst of bronchiolitis season, clinicians need to consider other causes of infant wheezing.

Conclusion

Wheezing in infants is common. Response to bronchodilators may not differentiate between bronchiolitis and other causes. Chest radiographs are not routinely recommended for bronchiolitis. Our infant's lack of response led us to obtain a chest radiograph which suggested a mediastinal mass. His subsequent deterioration would have been difficult to manage had he not undergone evaluation for this mass. His cystic hygroma was unusual in lacking the cervical component which usually suggests the diagnosis of a mediastinal lymphangioma. Clinicians need to avoid complacency in caring for wheezing infants in the midst of bronchiolitis season.

Declaration of conflicting interests

The authors report no conflict of interest.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

References

1. American Academy of Pediatrics, Subcommittee on Diagnosis and Management of Bronchiolitis. Diagnosis and management of bronchiolitis. *Pediatrics* 2006; 118(4): 1774–1793.
2. Zorc JJ and Hall CB. Bronchiolitis: recent evidence on diagnosis and management. *Pediatrics* 2010; 125(2): 342–349.
3. Schuh S, Lalani A, Allen U, et al. Evaluation of the utility of radiography in acute bronchiolitis. *J Pediatr* 2007; 150(4): 429–433.
4. Bees NR, Richards SW, Fearne C, et al. Neonatal thymic haemorrhage. *Br J Radiol* 1997; 70: 210–212.
5. Takamatsu H, Noguchi H, Tahara H, et al. Giant mediastinal lymphangioma in a neonate: report of a case. *Surg Today* 1996; 26: 527–531.
6. Gallagher PG and Mahoney M. Cystic hygroma in the fetus and newborn. *Semin Perinatol* 1999; 23(4): 341–356.
7. Glasson MJ and Taylor SF. Cervical, cervicomediastinal and intrathoracic lymphangioma. *Prog Pediatr Surg* 1991; 27: 62–83.
8. Alqahtani A, Nguyen LT, Flageole H, et al. 25 years' experience with lymphangiomas in children. *J Pediatr Surg* 1999; 34(7): 1164–1168.
9. Ninh TN and Ninh TX. Cystic hygroma in children: a report of 126 cases. *J Pediatr Surg* 1974; 9(2): 191–195.
10. Miller R and Nieder E. Neck masses in childhood. In: Kliegman RM (ed.) *Practical strategies*. Philadelphia, PA: W.B. Saunders, 1996, pp. 831–832.
11. Pham N, Alexander P, Chow CW, et al. Anterior mediastinal lymphangioma in an infant: diagnosis and surgical management. *Heart Lung Circ* 2012; 21: 289–291.
12. Muraskas JK, Gianopoulos JG, Husain A, et al. Mediastinal cystic hygroma: prenatal decompression with neonatal resection and recurrence at 19 months of age. *J Perinatol* 1993; 13: 381–384.
13. Bratu M, Brown M, Carter M, et al. Cystic hygroma of the mediastinum in children. *Am J Dis Child* 1970; 119: 348–351.