A Repeatedly Barking Baby

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Background:

Respiratory complaints are a frequent cause of parents bringing their children to the ED. They can range from the simple viral infection to life threatening emergencies. Often it is difficult to distinguish a potentially dangerous diagnosis from a simple case of croup. The Emergency physician is thus tasked with making difficult decisions with very little information. The easy patients are the ones who clearly look toxic or we have already intubated. Where the art of Emergency Medicine plays its most important role are with those bright-eyed healthy appearing children that may be harboring some unforeseen respiratory disaster.

Patient Presentation:

A 3 month old female presents with parents with 1 day history of "noisy breathing." The child is a 36 week single gestation female delivered via C-section without complications. There had been normal pre-natal care including regular ultrasounds. The patient has no tobacco exposure, and attends daycare.

During the patient's first emergency department (ED) visit, she is noted to have a cough, nasal congestion, and wheezing, but with an intact appetite, feeding without difficulty, and remaining interactive. There is no fever. Vital signs are: HR 127, RR 28, Temp 99.2°F rectally, O2 sat 100% on RA. On physical examination, the infant appears active and non-toxic with audible stridor and wheezing at rest. Chest radiograph shows possible focal narrowing of the thoracic trachea and a perihilar streaky density consistent with viral infection. The patient is treated with 4mg of oral dexamethasone solution and a racemic epinephrine nebulizer with complete resolution of symptoms. She is discharged with normal vital signs to follow up with her pediatrician.

Two days later the patient returns with her parents to the ED with the abrupt onset of stridor, wheezing and barking cough. The patient continues to appear active, playful and nontoxic; she feeds without difficulty. Vital signs at this time: HR 124, RR 42, Temp 98.4°F rectally, O2 sat 89% on RA taken on the left great toe and 97% on RA taken on the right thumb. Repeat chest radiography is normal. Soft tissue neck radiographs show mild subglottic narrowing. The patient is treated with an albuterol nebulizer treatment as well as a racemic epinephrine nebulizer treatment with dramatic improvement.

The patient presents for a third ED visit the next day following a visit to the pediatrician as her symptoms have returned. She is admitted to pediatrics for further diagnostic testing.

Differential Diagnosis:

- Croup
- Laryngotracheomalacia
- Vocal cord paralysis
- Subglottic stenosis
- Vascular ring

Conclusion:

While on the pediatrics floor, the patient continues to require frequent racemic epinephrine and albuterol nebulizer treatments. She progresses to having constant stridor refractory to racemic epi, steroids and humidified oxygen. She also came to require oxygen via nasal cannula to maintain oxygen saturations greater than 92%. A barium swallow shows what appeared to be fixed lesion leading to compression of the anterior portion of the esophagus. After discussion with the parents, a CT of the chest with contrast is performed which reveals a double aortic arch with an aortic diverticula encircling and compressing the trachea. The patient also has an anomalous left subclavian artery arising from the aorta itself. The patient is transferred to another institution that evening for emergency vascular surgery the next morning. The patient made a complete recovery since and is developing normally.

Images

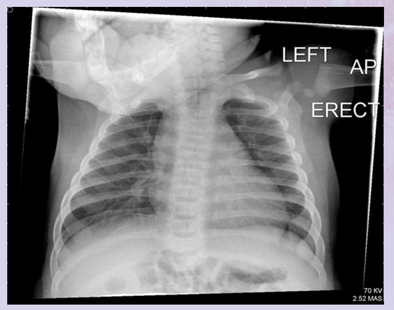

Image 1: A/P view of chest from visit 1.

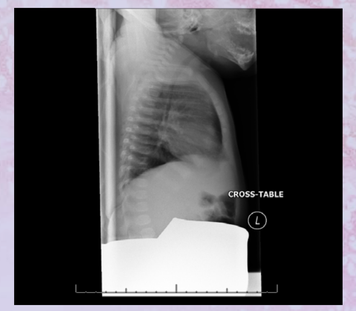

Image 2: CXR showing compression of the thoracic trachea.

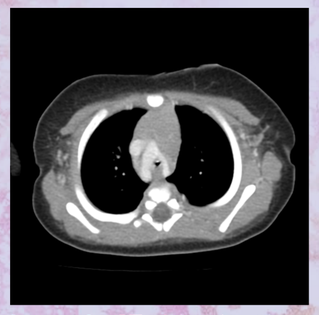

Image 3: CT of the chest showing double aortic arch encircling the trachea and esophagus.

Discussion:

Vascular rings are rare congenital anomalies of the aortic arch. They can completely encircle and compress the trachea, esophagus or even both. These lesions typically present in infancy; however, the variety of presenting symptoms makes timely diagnosis difficult. In several studies stridor, wheezing, recurrent upper respiratory infections, or feeding difficulties were among the most common presenting symptoms[1,2]. A diagnosis of a vascular ring must be entertained when infants appear with recurrent respiratory or gastrointestinal symptoms, especially in situations in which a viral illness is unlikely. Other clues can be found on radiography including abnormal or right-sided location of the aortic arch, indentation of the thoracic trachea (such as was described in this patient), and several others[3]. A careful history and physical must be done in order to not miss this potentially life threatening emergency. Repair of symptomatic lesions should be undertaken in most cases and all cases with severe airway compromise[4]. Most children have complete resolution of their symptoms after surgery. Factors influencing outcome include the degree of tracheomalacia prior to operative intervention, the degree to which respiratory function was optimized prior to surgery and the specific type of vascular abnormalitiy[4,5].

Resources:

1. Rosen's Emergency Medicine. Pediatric Respiratory Emergencies. pgs 2104-2113. 9th Edition. 2010.
2. Tintinalli's Emergency Medicine. Judith Tintinalli, et al Section 12 The Pediatric patient. 7th edition.2011.
3. Common and uncommon vascular rings and slings: a multi-modality review. Jonathan R Dillman, Anil K Attili, Prachi P Agarwal, Adam L Dorfman, Ramiro J Hernandez, Peter J Strouse, Pediatric Radiology. 2011.
4. Vascular ring abnormalities: A retrospective study of 62 patients. Bonnard et al. Journal of Pediatric Surgery. Voulme 38, Issue 4 pgs 539-543. April 2003.
5. Double Aortic Arch causing esophageal compression. Mao-Tang et al. The American Journal of Surgery. Vol. 165 Issue 5, pgs 628-631. May 1993.