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Case 3: Stridor and cough in a young child

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A five-year-old boy presented to a paediatric emergency department three times over a one-month period with symptoms consistent with croup. Each time, he was treated with dexamethasone and discharged with instructions to return if he experienced increased respiratory distress.

Two weeks later, he presented again because he had awoken from sleep with significant breathing difficulty, which was noisy. He then appeared to "turn black" and was confused for a few minutes. The parents believed his symptoms were worse when supine. On examination, the patient was in moderate respiratory distress. He was afebrile, with a heart rate of 127 beats/min, respiratory rate of 28 breaths/min, oxygen saturation of 98% and blood pressure of 96/62 mmHg. He had inspiratory stridor with increased work of breathing consisting of nasal flaring, a tracheal tug and moderate intercostal retractions. The remainder of the examination was unremarkable.

He was given nebulized epinephrine and was treated with oral dexamethasone. Soon, he was in no distress and speaking in full sentences. A soft-tissue lateral neck radiograph was obtained and was interpreted to be normal (Figure 1). The ear, nose and throat (ENT) service was consulted and performed a nasopharygoscopy (normal). The patient was discharged with a diagnosis of croup, but was called back when the original film was reviewed by radiology. A subsequent chest radiograph helped to reveal the final diagnosis (Figure 2).

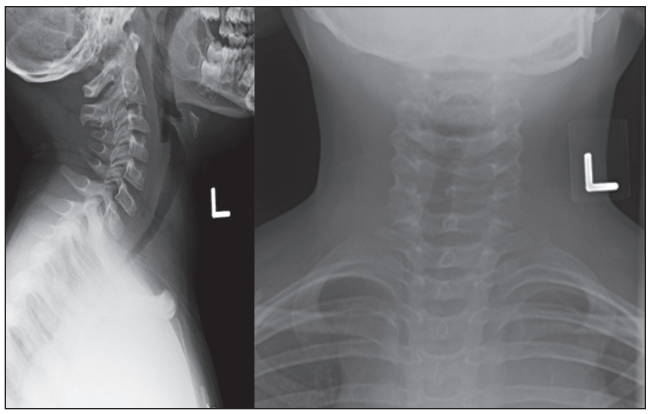

Figure 1) *Soft tissue lateral and posterior-anterior neck radiograph*

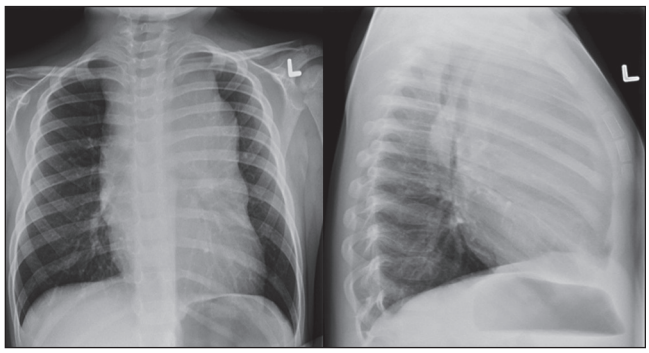

Figure 2) *Chest radiograph*

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CASE 3 DIAGNOSIS: MEDIASTINAL MASS FROM ACUTE LYMPHOBLASTIC LEUKEMIA

The soft-tissue lateral neck radiograph identified precipitous narrowing of the trachea, beginning at the C5 level with superior mediastinum widening and a concern for a mass extending superiorly through the thoracic inlet. The chest radiograph confirmed a mass in the anterior and superior mediastinum with displacement of the trachea right of the midline and suspected trachea narrowing. Subsequent blood work revealed a white blood cell count of 38.0×10^9/L (22.0×10^9/L lymphoblasts), hemoglobin level of 108 g/L and platelet count 139×10^9/L. Computed tomography of the head and neck further delineated the mass, confirming extension above the clavicles, with a mass effect on the trachea and esophagus to the right. The bone-marrow aspirate confirmed the diagnosis of acute lymphoblastic leukemia.

A child presenting and subsequently being diagnosed with multiple croup episodes or 'recurrent croup' may warrant further investigation. Diagnostic challenge occurs when the patient with stridor and nonspecific viral symptoms responds well to conventional therapy for croup. Clinicians should consider a broader differential diagnosis for 'recurrent croup' that results in stridor (Table 1). A retrospective study found that of 53 pediatric patients referred to ENT surgeons for 'recurrent croup', 28 (53%) were diagnosed with an intrinsic laryngotracheal disease, such as acquired laryngotracheal stenosis, and nine (17%) were diagnosed with extrinsic laryngotracheal disases, of whom two (4%) were found to have a mediastinal mass (1).

An interesting twist to our case was that the mediastinal mass was incidentally noted on the posteroanterior view of the soft-tissue lateral neck radiograph. Croup is typically a clinical diagnosis. Lateral neck radiographs have not been shown to be predictive of the severity of croup, but are helpful if the clinician considers other etiologies for upper airway obstruction. Interestingly, approximately one-third of mediastinal masses in childhood have been reported incidentally.

In a case series by Saraswatula et al (2), the authors stress the importance of consideration of age when a child presents with croup-like symptoms. Children >4 years of age tend not to present with croup, and teenagers rarely have this disease. Furthermore, mediastinal masses are significantly more common among older children and teenagers than among children <5 years of age.

Our patient also had a history of orthopnea, which is an unusual symptom for croup. Orthopnea as a symptom is highly correlated with risk of airway occlusion on induction of anesthesia. Orthopnea has also been a symptom of mediastinal masses that turn out to be leukemia/lymphoma (2).

Several case reports and case series involving malignant mediastinal neoplasms suggest that admission chest radiographs and severity of pulmonary symptoms may not be a reliable indicator of the degree of airway compromise, due to the observation that cardiorespiratory complications may occur abruptly. This was investigated by Lam et al (3), who found that 87.5% of patients with acute airway compromise presented with airway compression or displacement on radiological imaging, although the degree of compression on imaging was not associated with degree of airway compromise. As such, identification of risk factors associated with cardiorespiratory failure before any procedure requiring general anesthetic or sedation needs to be assessed. Risk factors include superior vena cava syndrome, pleural effusion, pericardial effusion, stridor, orthopnea, computed tomography findings of >50% cross-sectional compression of the trachea and peak expiratory flow rate <50% of the predicted value.

TABLE 1
Differential diagnosis for 'recurrent croup'

Infectious
Viral laryngotracheobronchitis (croup)
Bacterial tracheitis
Paraesophageal abscess
Congenital
Subglottic stenosis
Tracheal stenosis
Tracheobronchomalacia
Laryngotracheoesophageal cleft
Vocal cord paralysis or paresis
Tracheoesophageal fistula
Esophageal duplication cyst
Vascular ring
Congenital goiter
Inflammatory
Allergic reaction/anaphylaxis
Gastroesophageal reflux
Traumatic
Acquired subglottic stenosis (ie, prolonged intubation)
Foreign body (airway or esophageal)
Vocal cord paresis or paralysis
Thermal inhalation injury
Tumour
Subglottic or mediastinal hemangioma/hemangioendothelioma
Lymphangioma
Aberrant thyroid tissue
Thyroid neoplasm
Thymoma
Teratoma
Leukemia or lymphoma (mediastinal)
Neurofibroma or neuroblastoma
Granulomas (mediastinal)
Miscellaneous
Psychogenic

Adapted from reference 1

Clinicians should be aware of when a referral to an ENT surgeon for endoscopy should be considered based on specific risk factors (Table 2) (1).

Our patient initially presented with common respiratory symptoms consistent with croup and responded appropriately to treatment. However, over time, his presentation became atypical, with symptoms of orthopnea and recurrent stridor. The present case demonstrates the need for clinicians to further investigate a child presenting with multiple episodes of croup or upper airway obstruction with atypical symptoms; other etiologies, such as mediastinal masses, should be considered. Another reason for considering other diagnoses is that upper airway obstruction secondary to mediastinal mass can lead to life-threatening situations of critical upper airway occlusion, as the current literature reveals. Our patient proceeded to respond well to chemotherapy treatment for acute lymphoblastic leukemia.

CLINICAL PEARLS

- Clinicians should have a low threshold to investigate children presenting with multiple episodes of croup or upper airway obstruction with atypical symptoms.
- Special attention should be devoted to children >4 years of age or <6 months of age diagnosed with croup; failure to

TABLE 2
Risk factors for referral to an ear, nose and throat surgeon for endoscopy

Demographic characteristics
Children >4 years of age or <6 months of age diagnosed with croup
Frequency
>3 episodes of croup in one year
Severity
Failure to improve despite medical intervention
Multiple episodes of severe croup requiring emergent outpatient management for airway obstruction
Any inpatient management of croup
Presentation
Persistent stridor in a well-appearing patient
Medical history
History of neck trauma
Craniofacial abnormalities
Neurological disorders
History of prolonged neonatal intubation

Adapted from reference 1

improve despite medical intervention; and persistent stridor in a well-appearing patient.

- Orthopnea is an unusual symptom in children and anterior mediastinal masses should be considered

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