

CASE REPORT

Subglottic hemangioma: not all stridor is laryngitis

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Abstract

Stridor is the sound produced during respiration due to irregular airflow in a narrowed airway and can be due to a wide variety of conditions. In infants with persistent stridor, it is essential to include congenital upper airway malformations in the differential diagnosis.

Here we present the case of an almost two-month-old infant with persistent stridor due to subglottic hemangioma. This congenital anomaly of the airway is uncommon but important to recognize, as it can grow considerably and potentially compromise the patient's life. In addition, there is a specific therapy with systemic propranolol, which has proven effective in remission of growth and reduction of the size of these lesions.

HEMANGIOMA SUBGLÓTICO: NO TODO ESTRIDOR ES LARINGITIS

Resumen

El estridor es el sonido producido durante la respiración provocado por el paso de un flujo de aire que se vuelve turbulento al atravesar la vía aérea con un calibre estrechado, pudiendo deberse a una gran variedad de patologías. En los lactantes con estridor persistente, es imprescindible incluir en el diagnóstico diferencial las malformaciones congénitas de la vía aérea superior.

Se presenta el caso de una lactante de casi dos meses con estridor persistente debido a hemangioma subglótico. Esta anomalía congénita de la vía aérea es poco frecuente, pero de vital importancia, dado que en su evolución natural puede alcanzar un tamaño considerable llegando a comprometer la vida del paciente. Además, cuenta con una terapia específica con propranolol sistémico, la cual ha demostrado ser eficaz en la remisión del crecimiento y reducción del tamaño de estas lesiones.

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INTRODUCTION

Stridor is the sound produced during respiration due to irregular airflow in a narrowed airway. The most common cause in childhood is acute laryngitis of viral origin⁽¹⁾, although there are other infectious, inflammatory, anatomical, and functional causes, which may be congenital or acquired.

The differential diagnosis of stridor includes airway malformations, especially in children under 6 months of age, where the incidence of laryngitis is low and laryngomalacia is more common⁽¹⁾. These malformations include subglottic hemangiomas, proliferative vascular tumors that present with symptoms of airway obstruction starting at around two months of life⁽²⁾. They are characterized by obstructive dyspnea and biphasic stridor⁽²⁾. The diagnosis is made by observation of the lesion on laryngoscopy⁽²⁾, and it is treated with propranolol^(3,4).

CASE REPORT

The patient was an infant aged 1 month and 25 days (gestational age 41 weeks and 3 days), with no significant perinatal history, evaluated in the emergency department due to respiratory distress. The infant presented with a 2-week history of catarrhal symptoms, associated in the last 24 hours with respiratory distress and a metallic cough. No infectious exposures at home or family history were reported.

On arrival she presented with an unstable Pediatric Evaluation Triangle (PET) indicating respiratory distress. The airway was patent, with inspiratory stridor at rest, subcostal, intercostal, and supraclavicular retractions, polypnea up to 45 breaths per minute, oxygen saturation at 95%, and pulmonary auscultation revealing good bilateral ventilation without added noises. The heart rate was 165 bpm, blood pressure was 77/49 mmHg, and temperature 36.5°C. Capillary blood gases showed the following values: pH 7.38, pCO₂ 46.4 mmHg, HCO₃ 27.3 mmol/L, EB -1.6 mmol/L. Chest X-ray showed no abnormalities or tracheal stenosis; a nasopharyngeal swab was positive for rhinovirus.

Acute laryngitis of moderate intensity according to the Westley scale (4 points) was suspected, therefore nebulized adrenaline and oral dexamethasone were administered with a favorable response, decreasing the respiratory difficulty and stridor, which did not completely subside. She remained hospitalized for 4 days, received two additional doses of corticosteroids, and at discharge persisted with mild intermittent stridor during crying.

After 48 hours, the patient was readmitted to the emergency department due to increased stridor, associated with metallic cough and significant respiratory distress. Again, PET was altered with breathing difficulties, inspiratory stridor, and three-level retractions. Given the unfavorable course despite optimized treatment of laryngitis, and since the patient was younger than 6 months with persistent stridor after resolution of catarrhal symptoms, it was considered necessary to rule out a congenital malformation of the upper airway. Therefore, imaging tests and evaluation by otolaryngology were requested.

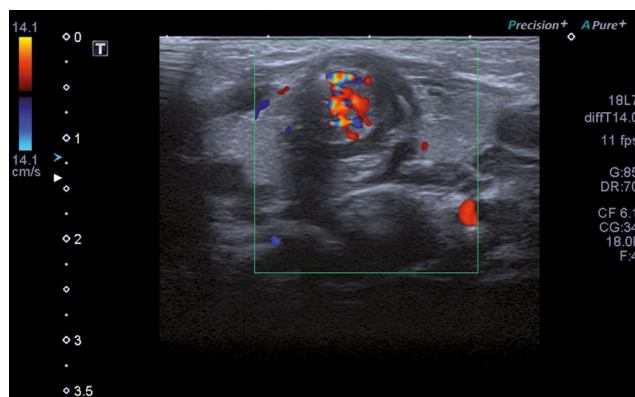


FIGURE 1. Color Doppler ultrasonography of the neck. Ultrasound imaging shows anterior displacement of the upper airway with lobular narrowing of the subglottic trachea. A measuring 7 x 7 mm is identified within the thyroid cartilage in the transverse plane, which appears markedly hypervascular on color Doppler.

Doppler ultrasound of the neck (Figure 1) showed anterior displacement of the upper airway with lobulated narrowing of the subglottic trachea due to the presence of a markedly hypervascular mass, findings suggestive of subglottic hemangioma. The study was completed with neck computed tomography (CT) (Figure 2), showing a focal mass with homogeneous enhancement after contrast administration, located in the left subglottic region with significant secondary laryngeal obstruction. A fibrolaryngoscopy was performed by otolaryngology, showing bilateral subglottic edema, more marked on the left side and in the posterior region.

As subglottic hemangioma was suspected, treatment was started with oral propranolol at 1 mg/kg/day, which was progressively increased to 3 mg/kg/day. The stridor disappeared completely in 48 hours and there were no adverse effects. After 5 days, an ultrasound revealed that the diameter of the lesion had decreased from 7 mm at diagnosis to 0.9 mm. After 3 months of treatment, a repeat fibrolaryngoscopy showed a small lesion remaining in the left subglottic region, occupying 10% of the lumen. By 10 months, complete remission was observed. At 12 months, propranolol was gradually reduced and discontinued one month later.

DISCUSSION

The presence of stridor suggests significant obstruction of the larger airways⁽⁵⁾. It is a common reason for consultation in pediatrics⁽⁶⁾ and is an important finding that requires rapid evaluation and sometimes emergency intervention.

Stridor is the form of presentation of a wide variety of upper airway disorders, and its characteristics may help guide the etiological diagnosis and indicate the level of the obstruction. A high volume suggests significant airflow obstruction, inspiratory stridor usually reflects obstruction in the supraglottic region, while expiratory stridor is associated with obstruction at the intrathoracic level. If the stridor is similar

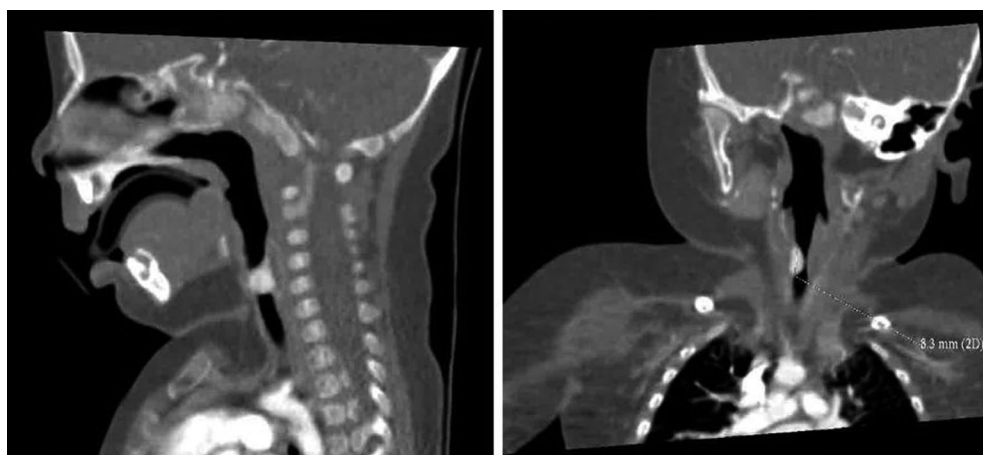


FIGURE 2. Computerized tomography scan of the neck. The CT scan of the neck reveals a focal mass measuring 8.3 mm, enhancing homogeneously after intravenous contrast administration. The lesion is located in the left subglottic region and is associated with significant secondary laryngeal obstruction.

in both phases of respiration, the lesion is likely located at the laryngeal level⁽¹⁾.

In children older than 6 months, the most common cause is acute laryngitis of viral origin⁽¹⁾. In infants younger than 6 months, the most common cause is laryngomalacia, although the differential diagnosis should include congenital upper airway malformations such as subglottic hemangioma, a rare but potentially fatal infantile hemangioma if it reaches a size that completely obstructs the tracheal lumen⁽³⁾.

Infantile hemangiomas are proliferative vascular tumors composed of endothelial cells that can be located anywhere in the body, including the respiratory tract. One percent of children with cutaneous infantile hemangiomas have subglottic hemangiomas, while 50% of children with subglottic hemangiomas have associated cutaneous hemangiomas, usually in the cervicofacial region^(2,5).

Subglottic hemangiomas predominate in females (2:1 ratio)⁽²⁾, and their most common location is at the left posterolateral subglottic level^(2,7). They are not present at birth and proliferate during the first 15-18 months of life. Symptoms appear at around 2 months, and their intensity depends on the degree of obstruction. The presence of dysphonic cough and recurrent croup is frequent, and biphasic stridor, with greater intensity during inspiration, is characteristic⁽²⁾. Diagnosis is made by direct laryngoscopy^(2,3). Plain radiography may show asymmetric narrowing of the subglottis, and CT with contrast may be useful to delineate large hemangiomas or those extending beyond the larynx.

Systemic therapy with propranolol is the first-line treatment and it is believed to act through a mechanism of inhibition of angiogenesis⁸. The dose is 1 to 3 mg/kg/day, divided into 2 or 3 doses, and should be maintained until 15 months^(5,7-10), at which point the proliferative phase of the hemangioma ends and involution begins according to the natural progression of the condition. The incidence of complications is low⁽²⁻⁴⁾, the failure rate is 0.9%⁽⁹⁾, and the recurrence rate after removal is 19-25%⁽¹⁰⁾. Other treatments classically used include systemic or locally injected corticosteroids and open surgical or laser resection. However, these are less effective compared to propranolol and are currently reserved as second-line treatments^(2,3,7).

COMMENTS

There is a wide variety of conditions that can cause stridor in infancy, of which laryngitis is the most common. The differential diagnosis of stridor should include congenital upper airway malformations, especially in infants with persistent or recurrent stridor.

Subglottic hemangioma is rare but potentially fatal; therefore, it should be suspected in infants younger than 6 months of age with persistent stridor and no other identifiable cause.

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