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CASE REPORT

Stridor in infant caused by subglottic hemangiome: case report

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Abstract

Subglottic hemagioma is a rare cause of stridor, but it is one of the most common vascular neoplasms of the airways in childhood. If the treatment is not promptly instituted, it becomes a life-threatening condition. The diagnosis should be suspected when infants outside the age range for acute laryngitis present with stridor associated with severe respiratory effort, without viral prodromes, with a condition that is not responsive to initial therapeutic measures considering the main diagnostic hypothesis. Infantile hemangiomas begin to proliferate during the first year of life (between the 1st and 2nd month of life). Involution usually occurs between 6 months and 12 months of life (most involution until 4 years). The case is a 5-month-old female infant, with sudden stridor associated with respiratory distress without viral prodromes or fever, with little response to inhaled short-acting beta-agonist, inhaled adrenaline, as well as corticosteroids inhalation/parenteral. Bronchoscopy showed a bulging of the submucosa to the right of the subglottis with slight vascularization, suggestive of subglottic hemangioma. Treatment with propranolol was initiated orally with the aim of regressing the hemangioma and after clinical stability, the infant was discharged with outpatient follow-up.

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INTRODUCTION

Stridor is a suggestive sign of upper airway obstruction, being an inspiratory, discontinuous and monotonic sound. Several etiologies can be responsible for stridor, such as congenital anomalies (laryngomalacia, vocal cord paralysis, laryngeal membrane, subglottic stenosis, laryngocele, vascular ring, choanal atresia) or acquired (infantile hemangioma, foreign body, infectious and inflammatory processes). Subglottic hemangioma is a vascular neoplasia, usually discovered in the first months of life. It manifests by cough, hoarseness and stridor, it usually is inspiratory. Direct laryngoscopy makes the diagnosis. The conduct can be from expectant in cases with minimal symptoms to surgical, including tracheostomy to protect the airway. Corticosteroids are a therapeutic option, but today propranolol is considered a first-line treatment.

CASE REPORT

Female infant, 5 months old, with a history of gastroschisis and intestinal atresia of the apple peel type, corrected in the neonatal period, presenting with sudden stridor and respiratory discomfort with one day of progress, without viral prodromes or fever. The child was taken to the emergency department, where she remained hospitalized for 24 hours. During this period, she received nebulization with short-acting beta 2 agonist and regular adrenaline, in addition to intramuscular hydrocortisone, but with no initial clinical response. The family then sought a federal hospital in Rio de Janeiro, where the child was born. She progressed with significant respiratory effort, worsening of the stridor and a drop in oxygen saturation, being admitted to the intensive care unit (ICU). Chest radiography upon admission showed no changes. The infant was submitted to a flexible transnasal laryngoscopy, showing subglottic and arytenoid edema. In the ICU, she remained in an oxygen hood, with regular nebulization of adrenaline, inhaled budesonide and antibiotic therapy. On the second day of hospitalization, her breathing pattern improved, and she was transferred to the pediatric ward in room air. Bronchoscopy performed during hospitalization revealed a submucosal bulging to the right of the subglottis, with slight vascularization, typical of a subglottic hemangioma. In addition, there was also an increase in vascularization in the nasal dorsum, suggestive of a flat hemangioma. During hospitalization, we opted to start propranolol orally for the treatment of hemangioma. The infant remained clinically stable and was discharged with propranolol to use at home.

DISCUSSION

Stridor is a common symptom in children and requires immediate medical evaluation, since its presence may indicate an underlying pathology with a potential risk of severity[1,2]. It is characterized by a respiratory noise caused by the whirling of the air that occurs due to a partially obstructed airway (laryngeal or tracheal)[2]. There are several causes of stridor, which can be divided according to the anatomical location: supraglottic (laryngomalacia, tumors, cysts, infection); glottic (laryngeal membranes, vocal fold paralysis, glottic stenosis, papillomatosis); subglottic (subglottic cysts, hemangiomas, strictures, croup or acute laryngitis, tracheal rings, bacterial tracheitis)[2].

Subglottic hemangioma is a rare cause of stridor, but it is one of the most common vascular neoplasms of the airways in infancy[3]. The incidence of infantile hemangiomas (IH) is higher in preterm infants, and low birth weight seems to represent a higher risk[3]. Hemangiomas, in general, affect 4 to 5% of Caucasian infants, are 4 to 5 times more common in females, and 1.4% of affected children will present concomitant airway injury[1]. Cutaneous IH in "beard distribution" are associated with greater airway involvement. About 50% of children with subglottic hemangioma have associated skin lesions[2]. A subgroup of patients with IH exhibits an association of ocular, brain, cerebrovascular, cardiovascular and thoracic abnormalities, all of which are included in a disorder called the PHACE syndrome[3,7].

IH begins to proliferate during the first year of life (usually between the 1st and 2nd months of life), completing their growth around the 5th month of life. Involution usually occurs between 6 months and 12 months. Most of them resolve by the 4th year of age[3]. Classically, subglottic hemangiomas manifest around 4 to 6 weeks of life, and can progress rapidly to respiratory failure[1]. A subglottic hemangioma should be suspected when infants outside the age group present with stridor associated with severe respiratory effort, with a condition that is not responsive to therapeutic measures if the hypothesis of acute (stridulous) laryngitis is considered[1,4]. In the case reported, the symptoms started later, at 5 months of age, with sudden onset biphasic stridor, with rapid progression to severe respiratory distress, without other symptoms. At first, the proposed diagnosis was acute laryngitis, despite not belonging to the age group of the disease. The infant did not respond to the treatment of the main diagnostic hypothesis: nebulization with adrenaline and systemic corticosteroids. Then, the possibility of a foreign body aspiration was suggested, due to the sudden clinical picture, and a chest X-ray was requested.

Flexible laryngoscopy can initially be important both to define the etiology and to exclude other differential diagnoses of stridor of congenital etiology, such as vocal fold paralysis, laryngeal cysts, subglottic stenosis, laryngomalacia, subglottic stenosis. It allows assessing the degree of airway obstruction, extent and location of the lesion, as well as possible complications, such as bleeding and ulcerations[1]. In the case report, due to the severity of the condition, laryngoscopy was first performed, which revealed subglottic and arytenoid fold edema, and the diagnosis of foreign body aspiration was ruled out. After clinical stability, the investigation continued with bronchoscopy for a better assessment of the airway, which showed increased vascularization in the nasal dorsum,

suggestive of a flat hemangioma and subglottic submucosal bulging to the right with discrete vascularization - image of a possible subglottic hemangioma.

Currently, propranolol is considered the first-line treatment. It is a low-cost, well-tolerated drug, being a non-invasive method of treatment. The proposed mode of action consists of vasoconstriction and inhibition of angiogenesis, mainly. Inducing endothelial cells apoptosis is questionable[3,6,9]. When used early, during the proliferative phase, it presents better results[5]. The recommended dose of propranolol is 2 to 3 mg/kg/day, with a response within 24 hours of starting therapy. Start at 1 mg/kg/day and increase the dose by 0.5 mg/kg/day per week[10]. Adverse effects include bradycardia, hypotension, bronchospasm and hypoglycemia[1,6]. Other therapeutic options include corticosteroids, antiangiogenic agents (alfa interferon), vincristine, laser therapy, surgical resection and even tracheostomy in cases of severe airway obstruction with impossibility of resection[8].

The patient in the case was discharged after two days of using propranolol at a dose of 2mg/kg/day, with clinical improvement, without respiratory distress, without stridor and had no side effects from the medication. She was followed up on an outpatient basis in dermatology and general pediatrics to assess lesion regression.

CONCLUSION

Subglottic hemangioma is a rare cause of stridor in children, but it should be suspected when children outside the typical age range for acute laryngitis present with stridor and severe respiratory distress, especially when there are no associated viral prodromes. Bronchoscopy is essential in the complete assessment of the airway, excluding differential diagnoses and, therefore, making its diagnosis. Propranolol is currently the first-line treatment, yielding good outcomes when started as early as possible.

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