Subglottic cyst: a rare cause of laryngeal stridor

Cisto subglótico: uma causa rara de estridor laríngeo

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To the Editor:

Subglottic cysts (SGCs) are a rare cause of airway obstruction in children, and only a small number of cases have been reported.[1-4] The first was published in 1968, when Wigger & Tang reported the case of an infant who died from airway obstruction caused by SGCs.[1] Medical advances, higher survival rates for preterm infants, and improved diagnostic equipment have increased the number of reported cases of SGCs over the last three decades, the majority occurring in infants who had been extremely premature neonates and had suffered from respiratory distress, therefore having been intubated and managed in neonatal ICUs.[3,5] Symptoms of laryngeal cysts depend on the size and the location of the cyst and include a change in the tone of voice, dysphonia, hoarseness, dysphagia, laryngeal stridor, and dyspnea.[6] This condition is often misdiagnosed as laryngomalacia, asthma, croup, or other diseases, due to the fact that it manifests as recurring respiratory infections, stridor, and wheezing. Death can occur in severe cases that are not treated.

We recently treated a 16-month-old female infant who had been born prematurely (29 weeks) and, shortly after birth, developed respiratory distress, requiring intubation. The infant remained intubated for 30 days in the neonatal ICU of our hospital. There were no episodes of accidental extubation or extubation failure. She was extubated, progressed well, and remained in the hospital to gain weight for another two months, being discharged with no respiratory complaints and under pediatric follow-up. Within the first 6 months of life, she developed rhinorrhea, cough, wheezing, and slight dyspnea. The infant was brought to the emergency room, where she was diagnosed with croup. She was treated with oral prednisolone and nebulized epinephrine. The patient showed partial improvement of symptoms and, despite presenting mild stridor, was discharged to home care. The mild stridor persisted, occurring daily, until she was 8 months of age, when she presented with worsening of dyspnea and respiratory distress accompanied by laryngeal stridor. In the same emergency room, the patient was again treated for croup and discharged to home care. She continued to have stridor on a daily basis until reaching the age of 11 months, when she presented severe respiratory distress with dyspnea, laryngeal stridor, hoarseness, and cough. She was admitted for treatment and further investigation of the stridor, undergoing fiberoptic laryngoscopy due to the suspicion of congenital laryngeal disease. This procedure revealed narrowing of the subglottis, and the patient underwent rigid bronchoscopy to confirm the diagnosis and to treat the condition. Prior to the procedure, the infant had biphasic stridor, hoarse cry, weak cough, and dyspnea. Her SpO_2 was 95% on room air. Rigid bronchoscopy, with the patient under general anesthesia, revealed a cystic lesion, consistent with an SGC, in the right lateral wall. The obstruction was reducing her airflow by 80%. The cyst was ruptured by the passage of the rigid bronchoscope, and the infant showed improvement in the symptoms. However, 1 month later, the stridor and dyspnea recurred. After the recurrence of the SGC had been confirmed, a second endoscopic procedure—suspension laryngoscopy and marsupialization with cup forceps—was performed. Subsequently, there was complete resolution of the symptoms. As of this writing (6 months after the last treatment), there had been no recurrence.

Although SGCs are rare, they can cause significant airway obstruction, which is reversible if the diagnosis is established early.[5] Recently, SGCs have become a more common finding in previously intubated infants. Among infants presenting with stridor and undergoing direct laryngoscopy or bronchoscopy, SGCs are diagnosed in 7%.[3,4] In addition, SGCs are an intubation-related process and represent an uncommon cause of stridor in infants.[7] When

managing preterm infants discharged from ICUs who present with airway obstruction, physicians should consider SGCs.[3] In children with recurrent stridor, laryngoscopy is the gold standard for the diagnosis of SGCs, because it allows early recognition of laryngeal diseases, including rare cases of cysts. Intubation remains the main means of ventilatory support in extremely premature neonates, and it can induce local subglottic trauma, which is the logical explanation for the genesis of SGCs. The sequence of events from subglottic injury to SGC formation has been described, and it is believed to involve acute mucosal tearing and ulceration followed by necrosis, granulation, and healing of the affected tissue. This healing process, in some cases, results in subepithelial fibrosis and squamous metaplasia, with subsequent obstruction of mucous glands and cyst formation.[8-10] Among the treatments for SGC, endoscopic marsupialization has been found to be the most efficacious and the treatment that most reduces the potential for recurrence. Topical application of mitomycin C might play a special role in the management of these cysts, because it can reduce post-marsupialization scarring and cyst recurrence. Other methods of treating SGCs include observation (for very small cysts) and bronchoscopic cyst puncture. Even when treated appropriately (surgically of clinically), SGCs recur in 19-60% of cases, especially in the first 4 months after the initial procedure.[2,3,5] A history of intubation, prematurity, and stridor should raise the clinical suspicion of SGCs.[3] All patients with recurrent stridor should be submitted to laryngoscopy in order to hasten the diagnosis and treatment.

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