Case Report

Subglottic hemangioma masquerading as croup and treated successfully with oral propranolol

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ABSTRACT

Subglottic hemangioma (SGH) is a rare and potentially life-threatening condition of the airway. A 3-month-old girl presented with croup which improved only partially with nebulized adrenaline and intramuscular dexamethasone. An upper airway endoscopy revealed the diagnosis of SGH. Oral propranolol was started, and following that, there was a dramatic response within 48-h of therapy, and complete remission after 1 year of therapy.

KEY WORDS: Croup, propranolol, subglottic hemangioma

INTRODUCTION

Subglottic hemangioma (SGH) is one of the rare hemangiomata which is a potentially life-threatening condition. It accounts for 1.5% of all congenital laryngeal abnormalities. The clinical presentations are heterogeneous and at time can lead to critical airway obstruction with mortality rate close to 50%.[1] Although both medical and surgical interventions are described in literatures, the common interventions are steroid (systemic and local), α-interferon, propranolol, carbon dioxide laser, bleomycin, excisional biopsy, and sometimes even tracheostomy.[2]

Here, we report a case of SGH who presented to us as croup and responded dramatically with oral propranolol.

CASE REPORT

A 3-month-old girl was presented with cold, cough and fever for 3 days, noisy, and difficulty in breathing for 1 day. At admission, she was febrile (102°F) and had fast breathing (respiratory rate: 68/mints), tachycardia (heart rate: 158/mints), blood pressure: 82/54 mmHg (50th centile), and SpO₂: 88%–90% on room air. On examination, inspiratory stridor was heard with moderate chest retraction. A working diagnosis of acute severe laryngotracheobronchitis was made, and she was started on humidified oxygen, nebulization with adrenaline and a single dose of intramuscular dexamethasone (0.6 mg/kg) was administered and admitted to the pediatric intensive care unit (PICU). She improved symptomatically over the next 2 days and was transferred to the ward. Overnight, she again developed severe respiratory distress with stridor for that she was shifted back to the PICU and started on supportive measures including nebulization. Routine investigations (hemograms, C-reactive protein, and blood culture) were within normal range including chest X-ray.

In view of recurrent episodes and only partial improvement with medication, airway anomalies were suspected, and upper airway endoscopy was performed which revealed a mass-like lesion arising from the right lateral wall in the larynx. An endoscopic biopsy was performed which revealed a subglottic hemangioma. Oral propranolol was started, and following that, there was a dramatic response within 48-h of therapy, and complete remission after 1 year of therapy.

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the subglottic region and occluding about 70% of the airway lumen [Figure 1a]. Contrast-enhanced computed tomography (CT) scan was also performed which revealed small, well-defined enhancing lesion in the submucosal location of the subglottic region on the right lateral and the posterior wall causing significant airway compromise likely SGH [Figure 1b]. Screening Doppler ultrasound of the abdomen and cranium and echocardiography were normal.

A final diagnosis of SGH was made based on upper airway endoscopy and imaging. Oral propranolol was started at an initial dose of 1 mg/kg/day, which was gradually increased to 2 mg/kg/day in three divided dosages with close monitoring of heart rate (electrocardiography), blood pressure, and blood sugar. The clinical condition was dramatically improved within 48 h of starting propranolol. Subsequently, she was discharged and kept on follow up initially weekly and then every month. Upper airway endoscopy was repeated every 3 months, last one at 12 months of follow-up which showed almost complete resolution of subglottic lesions with no luminal obstruction [Figure 1c]. After 1 year of therapy with complete clinical and endoscopic improvement, propranolol was stopped and she was continuing to follow-up. In the next 6 months, there was no recurrence of symptoms, and she gained adequate weight and achieved normal developmental milestones for age.

DISCUSSION

SGH is considered as common pediatric benign tumor of the airways. The natural course of SGH like other hemangioma has well-described patterns, appears in the 1st month of life, starts proliferation for next 6–9 months, followed by plateau at 1 year, and then enters into involuntary phase which may take 3–7 years. SGH may occur alone or concomitant with other cutaneous hemangioma, especially in a segmental distribution of “beard” area.[8]

The majority of infants with SGH usually present during the proliferation phase with sudden onset of biphasic stridor, feeding difficulty, excessive dry, and respiratory distress leading to critical airway obstruction, so early diagnosis and intervention is critical. A superimposed viral infection can lead to further airway compromise. The condition often gets confused with croup as both have a similar time of onset and also the fact that SGH often improves transiently with adrenaline nebulization.[9] In this index case, also the initial diagnosis was croup and she responded transiently with adrenaline nebulization and dexamethasone.

In isolated SGH, a very strong clinical suspicion is required. The diagnosis is usually established with upper airway endoscopy, but some time imaging (CT and/or magnetic resonance imaging) may be required to document the extent and depth of the lesions and to exclude other abnormalities in suspicious cases. The role of ultrasonography as a noninvasive modality is emerging in the management of SGH.[10]

For SGH, a number of treatment modalities are available which are often used either alone or in combination. Steroid remained the standard of treatment for a long time; however, due to its major adverse effect with prolonged use and only partial improvement, it is now not considered as first-line treatment.[6]

Propranolol has been recently added in the armamentarium of infantile hemangioma management.[7] Although it is now recommended as the first-line therapy for infantile hemangioma, there is a paucity of literature on its use in SGH. The usual recommended dose is 1–3 mg/kg/day in three divided doses, starts with lower dose, and gradually titrates the dose with heart rate, blood pressure, and blood sugar monitoring.[8] In this index case, the maximum given dose was 2 mg/kg/day. It is usually well tolerated among infants and its effect starts within 48 h. The usual adverse effects of propranolol are minor but sometime may develop major adverse effects, namely hyperglycemia, hypotension, and bradycardia which need close monitoring.[8] In the index case, it was well tolerated and she did not develop any adverse effect. Malik et al. in their randomized controlled trial had concluded that propranolol is rapid and consistently efficacious than prednisolone and the combination of both had comparable efficacy and was not superior than propranolol alone.[10]

CONCLUSION

SGH although a rare airway anomaly, but it is a potentially life-threatening condition. In isolated SGH, a high degree of suspicion is essential for the diagnosis. Oral propranolol is an effective noninvasive medical management of SGH which is well tolerated by infants, has rapid onset of...
action and showed complete remission after 12 months of therapy.

**Informed consent**
Parents' written consent for this publication was taken.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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