

Propranolol as first-line treatment of a severe subglottic haemangioma

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Abstract

Subglottic haemangioma (SGH) is a rare, benign tumour in children, which is potentially life-threatening because of airway obstruction. We report the case of a full-term 2-month-old infant girl admitted to our institution with stridor, dyspnoea and oxygen desaturation caused by a SGH and treated with propranolol. Neck-chest computed tomography (CT) revealed a contrast-enhancing, 10-mm, subglottic elliptical lesion, referable to SGH. Pre-treatment fibrobronchoscopy showed a sub-occlusive SGH closing more than 75% of the laryngotracheal airway. In agreement with our neonatologists and ear, nose and throat (ENT) specialists, we decided to begin oral propranolol therapy, which rapidly and dramatically improved respiratory symptoms. Fibrobronchoscopy six days after treatment confirmed a reduction of subglottic narrowing. Six months later the patient is doing well and without respiratory symptoms. To the best of our knowledge, this is the first reported case of the successful treatment with propranolol of an SGH obstructing more than 75% of the airway. The case is evidence of the effectiveness of oral propranolol as first-line treatment in the management of severely-obstructive paediatric SGH and the importance of CT and fibrobronchoscopy in the diagnosis; it also demonstrates the importance of multidisciplinary cooperation between thoracic surgeons, anaesthesiologists, neonatologists and ENT specialists in the treatment of these patients.

Keywords: Subglottic haemangioma • Laryngotracheal stenosis • Propranolol • Fibrobronchoscopy.

INTRODUCTION

Although haemangioma is the most common benign head and neck tumour in infants, infantile subglottic haemangioma (SGH) is rare (less than 5% of all congenital laryngotracheal abnormalities) but potentially life-threatening [1]. After an initial silent period, proliferation of SGH leads to biphasic stridor and other respiratory symptoms [2, 3]. If untreated, airway obstruction can occur shortly afterwards. However, if the airway is stable enough and the stenosis minimal, without severe respiratory symptoms, approximately 50% of SGHs left untreated involute by age 5 [2, 3]. Nevertheless, the natural history of SGHs is unpredictable [2, 3]. The problem lies in the ability of the lesion to compromise the airway before it involutes.

Management of infantile SGH remains a major challenge for paediatricians and surgeons, as SGH can be treated in various ways, but a standard method has not been established. Alternatives are (i) wait and see (with or without tracheotomy), hoping that laryngotracheal growth will reduce the relative degree of narrowing, (ii) systemic and intralesional steroids, (iii) interferon, (iv) CO₂ laser and (v) open surgery (laryngotracheoplasty, submucous resection, tracheostomy) [2, 3]. The appropriate management strategy remains controversial, although attempts have been made to rationalise the most effective treatments according to the size and location of the SGH [3].

Recently, a new medical approach with propranolol has been proposed with variable results [4]. We report the first case of a severely symptomatic SGH, successfully treated with a rescue conservative medical treatment using propranolol.

CASE REPORT

A full-term 2-month-old girl, weighing 5 kg, without cutaneous haemangioma, was presented to our institution with stridor, dyspnoea, oxygen desaturation (88%) requiring oxygen support of 2 l/min, and a heart rate of 180 beats per minute. She had already undergone chest X-ray (unremarkable) about one month before, at the onset of respiratory symptoms, and received pharmacological empiric therapy with albuterol, dexamethasone (0.6 mg/kg/day) and erythromycin, without relief.

Neck-chest CT scan, with transverse, coronal and sagittal planes images, revealed a contrast-enhancing 10 × 10 mm subglottic elliptical lesion, encroaching upon the tracheal air column, referable to SGH (Fig. 1A). Fibrobronchoscopy with a 2.7 mm instrument under sedation (intravenous midazolam, 2.5 mg and ketamine, 10 mg) in spontaneous ventilation, showed a soft, blue-tinted sub-occlusive lesion (Fig. 1B, [Supplementary Video 1](#)), closing more than 75% of the laryngotracheal airway (Myer-Cotton System, Grade III [5]), (Fig. 2), with a residual



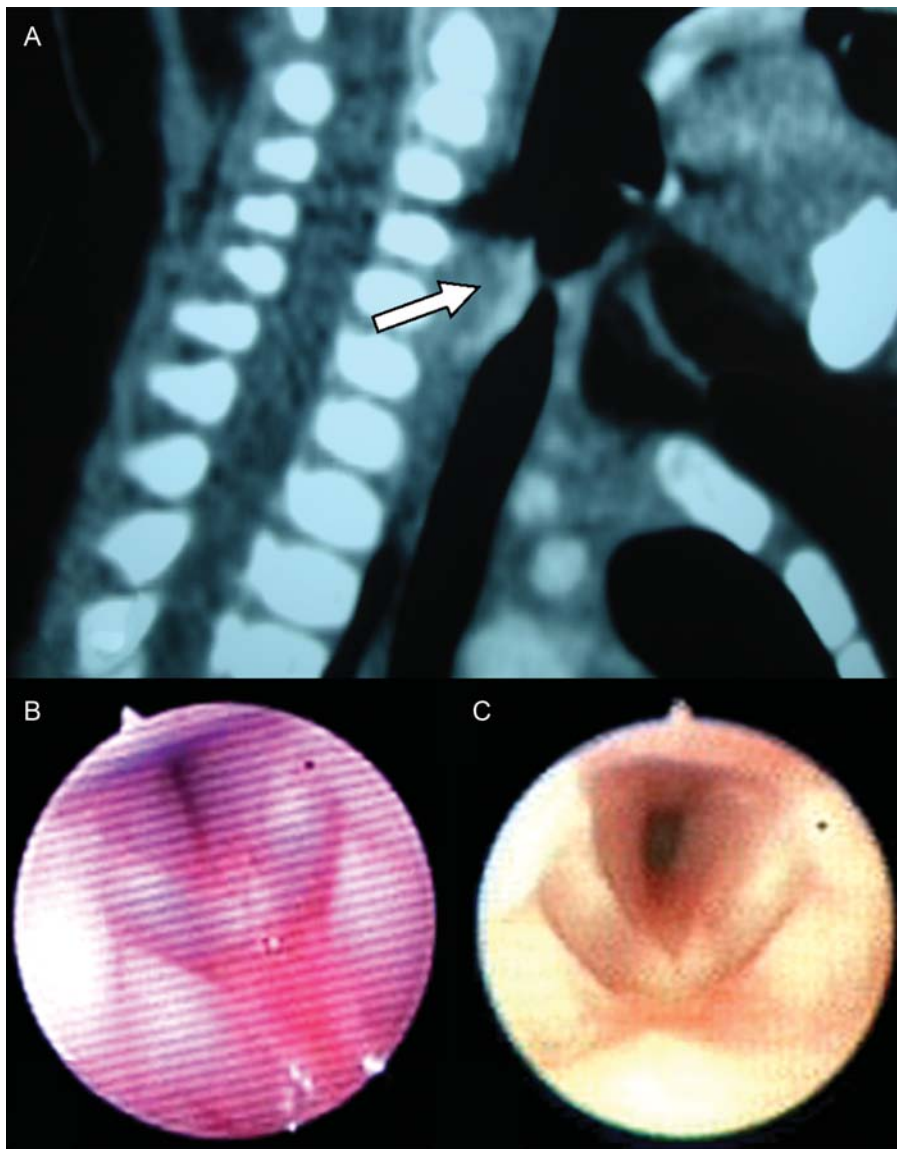


Figure 1: (A) Mid-sagittal contrast neck-chest CT image showing an elliptic mass in the subglottic area; the lesion is clearly visualized as a contrast-enhancing mass (arrow) encroaching upon the tracheal air column; (B) endoscopic appearance of the circumferential blue-tinted subglottic haemangioma before and six days after (C) propranolol treatment.

subglottic space of about 3.97 mm^2 , impossible to penetrate with the bronchoscope and a cuffless tube without being occlusive. The typical endoscopic appearance confirmed the radiological diagnosis of SGH [3].

Considering the patient's stable clinical condition at the time of endoscopy and the strict monitoring applied to the child, in agreement with our neonatologists, ENT specialists and with the parents' consent, we chose not to perform a tracheotomy and instead initiated a pharmacological therapy with oral propranolol, continuing dexamethasone (0.4 mg/kg/day for 14 days). Before starting, a dose test of 5 mg of propranolol was administered, carefully monitoring blood pressure, heart rate and blood glucose, which remained in the normal ranges. During the first 24 hours, the child's respiratory symptoms rapidly and dramatically improved. Medical treatment with propranolol *per os* (2 mg/kg/day) was planned, dividing the total daily dose (10 mg) into two administrations.

Post-treatment fibrobronchoscopy with a 2.7-mm instrument, six days after the first propranolol administration, revealed the

precise inferior limit of the lesion, under the cricoid cartilage, and confirmed reduction of both SGH and airway narrowing (Fig. 1C, [Supplementary Video 2](#)).

Clinical follow-up was set at two weeks after hospital discharge and subsequently every month: six months later the patient is doing well without respiratory symptoms.

DISCUSSION

SGH is a rare but potentially life-threatening disease. Females are more often affected (female-male ratio 2:1) [2]. Approximately 50% of these children also have cutaneous haemangioma, with a beard pattern distribution [3]. Histologically, most of them (91.6%) are capillary; the others are cavernous or mixed [2]. SGH is usually not evident at birth but grows rapidly during the first year of life [2]: the proliferation phase begins around 1–2 months of age, causing intermittent airway obstruction which is more evident during periods of agitation

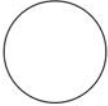




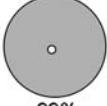
Classification of obstruction	From	To
Grade I	 0%	 50%
Grade II	 51%	 70%
Grade III	 71%	 99%
Grade IV	No detectable lumen	

Figure 2: The Myers-Cotton subglottic stenosis grading scale (adapted from [5]). Developed at Cincinnati Children's Hospital Medical Center (USA), it is used to standardize the measurement of subglottic stenosis and group together patients having similar severity of disease, in order to allow discussion among specialists caring for these patients.

and crying [3]. The most common symptoms are stridor (usually biphasic but more prominent during inspiration), respiratory distress, cough, hoarseness, dysphagia, vomiting and haemoptysis [3].

Diagnosis is the main challenge, especially in an unstable patient with respiratory distress.

As a first diagnostic step, neck-chest X-ray can determine the location and the symmetry or asymmetry of subglottic narrowing [3]. Neck-chest CT with multiplanar reconstruction represents the main method of investigation through imaging, revealing the precise site, contrast enhancement and shape of the SGH. Magnetic resonance imaging is an adjuvant tool but less available and used [2, 3].

Definitive diagnosis can be made by endoscopic examination under sedation during spontaneous breathing (endotracheal intubation would conceal the lesion) but meticulous care is required to maintain an air passage during the procedure. A detailed and accurate airway endoscopy is critical to determine the diagnosis and the best treatment option. The endoscopic characteristics of the lesion (colour, consistency) should be noted: endoscopic tissue colour—along with radiographic findings—confirms the diagnosis of SGH, avoiding the need for histological sampling [3]. If possible, the airway is sized with cuffless endotracheal tubes and graded on the Myer-Cotton system [5].

Our case is the first described severe SGH (Myer-Cotton System, Grade III) successfully medically treated (using propranolol) without initially performing surgical procedures [3].

Endoscopic photography and video-recording allowed later review of the airway with ENT colleagues, confirming the diagnosis and planning the treatment with neonatologists; also it was useful for a short-term (six days) endoscopic follow-up, which gave substance to improvements in respiratory symptoms, allowing long-term treatment to be planned. Clinical and endoscopic follow-up of SGH is recommended, at the outset closer and then deferred in time [3].

In conclusion, multiple techniques have been utilized in the treatment of SGH and no single option is acceptable for all patients [2, 3]. Treatment should aim to reduce the size of SGH, resolve symptoms and maintain a stable lesion involution. Based on our experience we suggest that, subject to an accurate clinical diagnosis, propranolol medical treatment should be considered in all SGH cases, in place of procedures which have previously resulted in higher mortality and morbidity.

SUPPLEMENTARY MATERIAL

Supplementary material (Videos 1 and 2) is available at *EJCTS* online.

Video 1: Fiberbronchoscopy with 2.7 mm instrument under sedation in spontaneous ventilation, showing a blue tinted sub-occlusive lesion, closing more than 75% of the laryngotracheal airway, impossible to overcome with the bronchoscope and a cuffless tube without being occlusive; the typical endoscopic appearance confirmed the radiologic diagnosis of SGH.

Video 2: Post-treatment fiberbronchoscopy with 2.7 mm instrument under sedation in spontaneous ventilation, 6-days after the first propranolol administration, confirming both SGH and airway narrowing reduction; it was possible to overcome the subglottic area with the bronchoscope, visualizing the trachea and the main carina.

Conflict of interest: none declared.

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