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Open airway surgery for subglottic hemangioma in the era of propranolol: Is it still indicated?☆

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ABSTRACT

Objectives: With the emergence of propranolol as the primary treatment for hemangiomas the indications for surgical intervention have been greatly reduced. There remains a role for surgical management in those patients who fail medical therapy, particularly for hemangiomas involving the airway. Detailed is our experience with subglottic hemangiomas, including three patients who failed propranolol treatment and were successfully treated with surgical excision and single stage laryngotracheoplasty (LTP) with thyroid ala graft.
Methods: Retrospective case series (level of evidence: 4).
Results: Six patients were treated with propranolol for subglottic hemangiomas over a 6 year period (2008–2014). Three patients responded to propranolol therapy and required no adjunctive surgical procedures. Three patients failed propranolol treatment, and required open resection of their subglottic hemangiomas and thyroid ala graft placement. Indications for resection were complete lack of response to propranolol in one patient, and initial response to propranolol with subsequent regrowth in the other two patients. All three patients were treated with submucosal extirpation of their hemangioma and single stage LTP; hemangioma was confirmed in all cases by positive GLUT-1 staining. All three surgical patients were successfully extubated post-operatively and none had hemangioma regrowth.
Conclusions: Fifty percent of patients in our series did not have long-term response to propranolol for subglottic hemangioma, highlighting the importance of close follow-up. When identified early, subglottic hemangiomas refractory to propranolol treatment can be successfully addressed with single stage LTP and tracheotomy can be avoided.

1. Introduction

Hemangiomas are the most common benign tumor of infancy, with an incidence of approximately 10%. Fortunately, the majority of these lesions are superficial in nature, involving only skin and subcutaneous tissues. Given their natural history of rapid proliferation during the first year of life followed by spontaneous involution, many of these lesions can be managed conservatively. Subglottic hemangiomas however, are of particular concern, since they can rapidly enlarge and cause significant airway distress.

Historically, several different modalities of treatment have been successful in the management of problematic hemangiomas, including systemic steroids and vincristine. In the case of symptomatic subglottic hemangiomas, tracheotomy has always been considered a safe and effective option. In more recent years, several other treatment modalities, including intralesional steroids, endoscopic laser excision, and open excision have also been described.

Propanolol, a non-selective beta-blocker, was incidentally discovered to rapidly and drastically reduce the size of proliferative hemangiomas in 2008 [1], and this discovery has revolutionized the management of hemangiomas. A follow-up retrospective study in France showed that propranolol effectively reduced 37 of 39 head and neck hemangiomas, and advocated propranolol as a first line treatment for head and neck hemangiomas. Since that time, there have been several successful reports of propranolol use for airway hemangiomas, either as part of a combined therapy with corticosteroids [2–4], or as isolated therapy [5–8]. Leboulangier published a series of 14 patients with subglottic hemangioma recalcitrant to other forms of treatment, all of whom responded to propranolol therapy initially [9]. However, he did report 2 patients

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who had regrowth following premature cessation of therapy, one of whom became resistant to beta blocker therapy and required adjunctive procedures. Several other recent reports have also highlighted the potential for regrowth despite initial response to propranolol [10–12]. These reports highlight the importance of close follow-up in these patients and suggest that propranolol may not be as effective for airway hemangiomas as originally described.

Like many institutions, we have adopted propranolol as first line therapy for hemangiomas, including subglottic hemangiomas. Here, we report our experience with propranolol for subglottic hemangioma in six consecutive patients, three of whom had excellent response to propranolol, and three of whom ultimately required surgical extirpation of their hemangioma.

2. Materials and methods

Six patients who were treated at the Children's Hospital of UPMC for subglottic hemangioma between 2009 and 2014 were included in this retrospective study. Institutional review board approval was obtained through the University of Pittsburgh. Inclusion criteria included patients who had a diagnosis of subglottic hemangioma based on bronchoscopy findings and in whom propranolol therapy was initiated were included in the study. Exclusion criteria included patients who did not have bronchoscopy-confirmed subglottic hemangioma, and patients who underwent therapies other than propranolol as a primary treatment. Patients were treated with 2–3 mg/kg/day of propranolol divided into TID dosing. Patients also received a short course (24–72 h) of IV steroids at the time of initial diagnosis, however none received prolonged steroid treatment. The patients charts were reviewed for demographics, presence of additional hemangiomas, age at diagnosis, response to propranolol and need for additional therapies.

All three patients who failed medical management underwent open surgical excision through an anterior cricoid split approach, as described by Vijayasekaran [13]. Following exposure of the hemangioma through an anterior cricoid split, careful submucosal dissection is carried out for hemangioma resection. This allows for preservation of a mucosal flap to line the underlying cartilage. Following surgical extirpation of the hemangioma with meticulous submucosa, a thyroid ala graft is used to augment the laryngeal framework. Our surgical technique is shown in Fig. 1.

3. Results

Six patients who underwent treatment with propranolol were identified. Interestingly, all 6 patients were female, and the diagnosis was made between 2 and 4 months of age in all cases. All patients had stridor as the presenting symptom which prompted bronchoscopy, and 4/6 patients had concomitant cutaneous hemangiomas, all with involvement of the head and neck. At the time of initial diagnosis, the percent of airway obstruction ranged from 30 to 90 percent. The percent obstruction was determined by sizing the airway with endotracheal tubes and calculating percent obstruction based on age appropriate endotracheal tube size. All patients had follow-up bronchoscopy to assess response to propanolol 2–4 weeks following diagnosis with the exception of one patient who was lost to follow-up for 2 years. None of the patients underwent prolonged treatment with systemic steroids, balloon dilation or steroid injection of the hemangioma.

Three patients had an excellent response to propranolol therapy and did not require any open surgical intervention. Three patients ultimately required open surgical resection. One patient had no significant response to propranolol therapy despite being maximized and 3 mg/kg/day, with continued hemangioma growth despite propranolol treatment. She had worsening stridor and respiratory distress as well as feeding difficulty and underwent open extirpation of her hemangioma with laryngotracheoplasty (LTP) with thyroid ala graft at 3 months of age. Another patient had initial good response to propranolol, with reduction in size of the hemangioma from 50% obstructive to 20% obstructive during the first 2 weeks of therapy. In the ensuing months however she developed worsening respiratory distress and was found to have regrowth to 60% obstruction despite continued treatment with 3 mg/kg/day of propanolol; she underwent surgical resection at age 11 months. The last patient also had excellent response to propranolol, but had regrowth with attempted weaning of propranolol at 1.5 years of age, and then again at 2.5 years of age; with weaning of propranolol, her hemangioma regrew to be approximately 50% obstructive and caused significant stridor and respiratory distress. She ultimately underwent open surgical resection at 34 months following two failed attempts at weaning. The diagnosis was confirmed with positive glut-1 staining on pathology in all three cases. Results are summarized in Table 1.

All three surgical patients underwent extirpation of subglottic hemangioma via anterior cricoid split and anterior thyroid ala graft. Other authors have described successful surgical extirpation of hemangioma, often in a single stage fashion with thyroid ala graft augmentation [10,14]. However, there are other options; Vijayasekaran [13] published a series of 21 patients who underwent open excision of subglottic hemangiomas, and only 10 of them were augmented with either costal or thyroid cartilage. Javia [15] described a series of 13 patients, 8 of whom underwent thyroid ala graft, 4 underwent primary closure without grafting, and 1 underwent costal cartilage grafting. Our preference is to

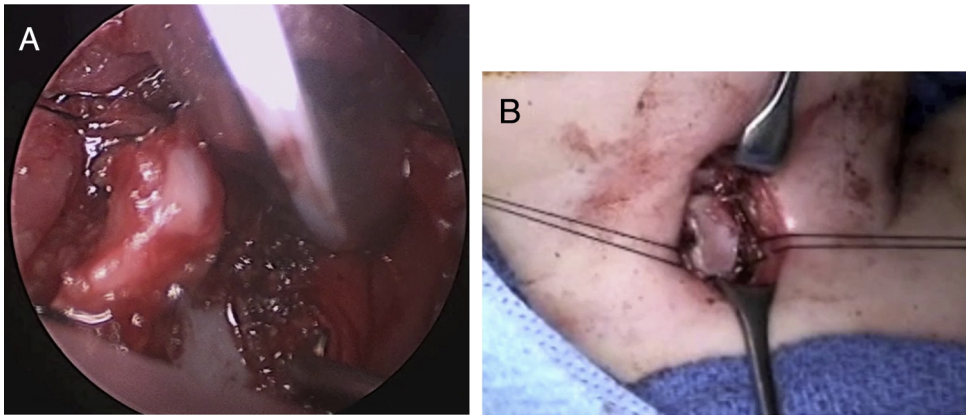

Fig. 1. Surgical technique. (a) Submucosal resection of hemangioma using a curette to maintain a mucosal flap. (b) Airway is augmented with anterior thyroid ala graft.