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Subglottic hemangioma: Understanding the association with facial segmental hemangioma in a beard distribution

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ABSTRACT

Objective: A subglottic hemangioma (SGH) is a benign tumor of infancy that can cause severe obstruction of the airway. Infantile hemangiomas, in general, are the most common head and neck tumor in children, affecting 4–5% of the pediatric population. This retrospective cohort study characterizes subglottic infantile hemangiomas at a single vascular anomaly center over a 5-year period (2013–2017) during the era of propranolol treatment.
Methods: Queried the Vascular Anomaly Database at Children's Hospital of Pittsburgh for all infantile hemangioma(s) and then identified case of subglottic hemangiomas. Characterized key features of presentation, natural history and management for subglottic hemangiomas. A secondary differentiation focused on differences between subglottic hemangiomas associated with Beard Distribution (BD) vs not (NBD).
Results: Analysis of 761 cases of infantile hemangiomas demonstrated only 13 patients with subglottic hemangiomas (1.7%). Of those 13 patients, only 4 patients (30%) had BD while 2 patients (15%) had other cutaneous hemangiomas and 7 patients (55%) had no cutaneous hemangiomas. Secondarily, a total of 31 case of beard distribution cutaneous hemangiomas with 11 patients having oropharyngeal involvement (35%) but only 4 patients with subglottic hemangiomas (13%). Interestingly, 2 of the 4 BD patients had treatment failure on propranolol and required second line treatment with steroids or surgical excision while only 1 of 9 NBD patients failed propranolol treatment. As well the same 2 BD patients which failed propranolol also had PHACES syndrome.
Conclusion: Subglottic hemangiomas are a rare presentation of infantile hemangiomas but with significant morbidity. While the classic teaching that a segmental beard distribution hemangioma raises concern for a subglottic hemangioma, this cohort indicates subglottic hemangiomas occur in a NBD presentation (1.3%), and demonstrated only an approximate 10% incidence rate with a beard distribution. But more importantly, this study raises the question that beard distribution in setting of PHACES syndrome may herald a more recalcitrant and complicated natural history for a subglottic hemangioma. This is of significant concern as risk for CVA in setting of PHACES is highest with use of steroid treatment. None of our patients had high risk extra or intra cranial vascular arterial anomalies and no CVA were noted.

1. Introduction

The infantile hemangioma (IH) is a proliferative vascular tumor consisting of endothelial cells which can occur anywhere in the body. IH is the most common head and neck tumor in children with an incidence of 4–5% in the general population [1] but involvement in the subglottic region is both rare and life threatening. The classic teaching is that the IH is not present at birth but then develops in the first months of life. The IH first enters a proliferation phase and rapidly grows for 3–5 months. This is followed by a prolonged involution phase with spontaneous regression over the course of years [2]. Because of the small caliber of the infant airway, during the proliferative phase the rapidly growing IH of the subglottic region can lead to a life-threatening airway obstruction. A study by Perkins et al. evaluating US trends in

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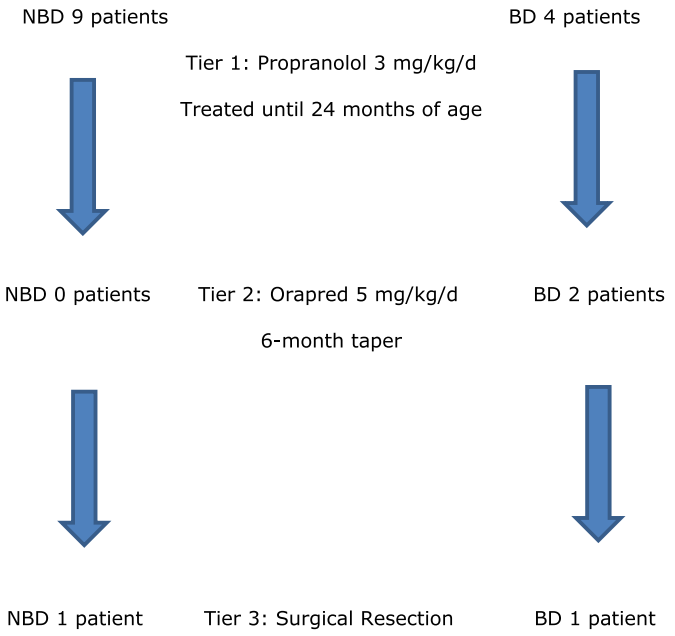

Fig. 1. Pathway comparison of patients with BD v NBD subglottic hemangiomas in response to treatment.

management of subglottic IH demonstrated increased morbidity (30% re-hospitalization) and mortality (2%) in this subpopulation [3]. Therefore, early diagnosis and efficacious treatment modalities are crucial to the successful management of children with subglottic IH.

The standard teaching is that a segmental IH in a beard distribution heralds airway involvement. One study demonstrated that airway involvement in the oral cavity, oral pharynx, hypopharynx, glottis, or sub glottis was seen in 29% of the children with beard distribution hemangioma [4]. In contrast, another classic presentation of subglottic IH is after a persistent croup-like illness without any cutaneous involvement [5]. The literature has not given a complete picture of presentation and natural history of the subglottic IH. The narrative has become more complicated with the recent advent of propranolol and a shift in treatment modalities away from oral steroids and surgical resection as demonstrated by a recent multi-center trial [6].

The purpose of this study is to characterize the presentation, natural history and management of subglottic IH at a single vascular anomaly center over a 5-year period in the age of propranolol management. The aim is to determine indicators of early diagnosis and markers of disease prognosis.

Table 2
Patients with Subglottic hemangioma at time of diagnosis including associated cutaneous hemangioma, age at presentation, age at diagnosis and percent obstruction at diagnosis.

Associated Cutaneous Hemangioma	Age at Presentation (weeks)	Age at Diagnosis (weeks)	Percent Obstruction at Diagnosis
None	12	14	80%
None	8	9	70%
None	8	16	80%
None	8	12	90%
None	20	22	75%
None	10	11	50%
None	20	24	90%
Facial Zone 3	2	2	30%
Facial Zone 1	8	8	80%
Facial Zone 1	8	8	70%
Facial Zone 3 & 1	2	2	10%
Facial Zone 3	4	4	10%
Facial Zone 3	2	2	41%

Table 3
Subgrouping of patients with Subglottic hemangioma without associated cutaneous hemangioma demonstrating delay of time between presentation and diagnosis and number of evaluations in interim period.

Age at Presentation (weeks)	Age at Diagnosis (weeks)	Number of Eval prior to Dx	Diagnosis at prior Evaluation
12	14	1	Croup
8	9	1	Upper Respiratory Infection
8	16	1	Gastroesophageal Reflux
8	12	1	Bronchiolitis
20	22	4	Upper Respiratory Infection
10	11	1	Gastroesophageal Reflux
20	24	1	Upper Respiratory Infection

2. Method

This study was IRB approved by the Institutional Review Board at Children's Hospital of Pittsburgh of UPMC. Retrospective review was preformed to identify all patients presenting to the Vascular Anomaly Center at Children's Hospital of Pittsburgh between 2013 and 2017 with an infantile hemangioma of the subglottic region. Data was collected on patient demographics, cutaneous involvement, characteristics of subglottic obstruction and response to treatment. A secondary analysis reviewed all patients with segmental hemangioma in facial zone 3 and compared subglottic IH associated with beard distribution vs not to

Table 1
Patients with Subglottic hemangioma at time of diagnosis including associated cutaneous hemangioma, treatment course, rebound after treatment and associated PHACES.

Associated Cutaneous Hemangioma	Treatment	Rebound	PHACES Anomalies
None	Propranolol until 24 months	Yes	None
None	Propranolol until 24 months	No	None
None	Propranolol until 24 months	No	None
None	Propranolol until 24 months	No	None
None	Propranolol until 24 months	No	None
None	Propranolol until 24 months	No	None
None	Propranolol 1 week then surgical resection	No	None
Facial Zone 3	Propranolol until 24 months	No	None
Facial Zone 1	Propranolol until 24 months	No	None
Facial Zone 1	Propranolol until 24 months	No	None
Facial Zone 3 & 1	Propranolol 4 months + Surgical resection	Yes	Sternal cleft
Facial Zone 3	Propranolol 24 months	No	None
Facial Zone 3	Propranolol 24 months + Steroids 6 months	Yes	Hypoplasia left common carotid