

Prognosis of Subglottic Haemangiomas Associated with Facial Haemangiomas in a Paediatric Population: A Preliminary Study

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Sir,

Subglottic haemangiomas (SGH) are rare benign vascular soft-tissue tumours representing 1.5% of congenital abnormalities of the larynx (1). Not usually present during the first few weeks of life, they then grow over the following 6 to 18 months and spontaneously regress after the 18th month. However, they are potentially life-threatening, with varying degrees of respiratory compromise (2). SGH can be isolated or associated with cutaneous lesions. The frequency of association with skin lesions varies between 30% and 50% (2). Recently, some authors have stated that the presence of a facial haemangioma distributed in a beard pattern was highly correlated with the presence of haemangioma in the upper airway or subglottic regions (3). Are SGH associated with facial haemangiomas in a beard distribution more aggressive than isolated SGH? We designed this retrospective study in order to answer this question.

METHODS

The patient records from August 1995 to January 1999 of all children presenting with SGH in the Otorhinolaryngology Department of our Children's hospital were reviewed retrospectively. Fifteen cases were assembled and diagnosed by the typical clinical history and bronchoscopy. The children were divided into two groups: group I – isolated SGH or SGH associated with skin haemangiomas not localized on the face, and group II – SGH associated with facial haemangiomas in a beard distribution.

Beard distribution indicates that locations of haemangioma are preauricular regions (uni or bilateral), lower lip, chin and anterior portion of the neck, and the score is given on the basis of the number of regions involved (1 to 5). Patients with four or more affected regions had a beard-like distribution, as shown by Orlov et al. (3).

The following information was recorded for each group: age, sex, presenting features, localization and type of skin haemangiomas associated with SGH, other malformations, bronchoscopic data, treatment modalities and outcomes. Statistical analysis using Student's *t*-test was performed.

RESULTS

Fifteen children were identified: nine with isolated SGH (group I) and six with SGH and facial haemangiomas in beard distribution, score ≥ 4 (group II) (Table I). The sex ratio (M/F) was 6/3 in group I and 1/5 in group II. Symptoms began at 3 months of age (ranging from birth to 8 months) in group I and at 1 month of

age (ranging from birth to 3 months) in group II. Abnormalities apart from subglottic or skin haemangioma were present in one child in group II only, and included a minor form of PHACE(S) syndrome (association of posterior fossa brain malformations, haemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, eye abnormalities, sternal non-union) (5). Bronchoscopic data revealed no circumferential lesions. Median occlusion of the subglottic area was 60% in group I and 52% in group II.

The treatments in our series are summarized in Table I. In group I, a short-duration treatment (less than 2 weeks) of oral steroids (mean dose of prednisone 2.3 ± 1 mg kg⁻¹ day⁻¹) was administered in 5 children (56%) and resulted in good evolution. Prolonged treatment (required for a median duration of 148 days) was used in 3 children and prolonged treatment associated with intubation in another 2 children. Mean duration of intubation was 10 days. In group I, three children developed side effects of steroids. In the whole group, mean duration of treatment was 72.8 days.

In group II, a short-duration treatment of steroids (mean dose of prednisone 1.9 ± 0.8 mg kg⁻¹ day⁻¹) was prescribed in 2 children and long-term treatment (median of 137 days) in 4 children, with injection of local steroid and intubation for 14 days in one case. Mean duration of intubation was 9.3 days. In the whole group, mean duration of treatment was 113 days. Two children in group II developed adrenocortical insufficiency.

DISCUSSION

Haemangiomas are benign neoplasms of the vascular endothelium. Recently, the association of facial haemangiomas distributed in a beard pattern and symptomatic SGH has been emphasized (3). Orlov et al. described 14% of superficial haemangiomas of the preauricular region, chin, anterior neck and lower lip and 63% of the children presenting a score of 4 or 5 had symptomatic airway involvement. Tracheotomy was required for 40% of these children. Other authors have observed this association (4). However, the risk of symptomatic haemangiomas occurring in the upper airway or subglottic regions when facial haemangiomas are present has been determined only by Orlov et al. (3).

In our study, 6/15 (40%) of the children had SGH associated with facial haemangioma in a beard

Table I. Symptoms, bronchoscopic data, treatment and outcomes of subglottic haemangiomas (SGH).

Patient	Age at first symptoms (months)	Sex	Symptoms	Skin haemangiomas	Score of Orlow	Bronchoscopic data	Duration of steroid therapy (days)	Duration of intubation (days)	Outcome
Group I									
1	At birth	F	Stridor	No		Anterolateral SGH	198	13	In remission
2	2	M	Laryngitis	No		Left hemicircumferential SGH	86		NED
3	2	M	Laryngitis	No		Posterior SGH	60		In remission
4	At birth	F	Stridor	No		Posterior SGH, laryngomalacia	248	7	In remission
5	8	M	Laryngitis	No		Anterolateral SGH	14		In remission
6	4	F	Dyspnea + cough	No		Posterolateral SGH	14		NED
7	4.5	M	Laryngitis (premature)	Yes (left forearm, dorsal region)		Posterolateral SGH	12		NED
8	4	M	Stridor	No		Anterolateral SGH	10		In remission
9	7	M	Stridor	No		Posterolateral SGH	14		In remission
Group II									
10	2	M	Stridor	Yes (beard, oropharyngeal)	5	Anterolateral SGH	165	9	In remission
11	3	F	Stridor	Yes (beard, oropharyngeal)	4	Left hemicircumferential SGH	14		In remission
12	1	F	Laryngitis	Yes (beard)	5	Anterior SGH	82	5	ACI
13*	3	F	Stridor	Yes (beard, oropharyngeal)	5	Anterior SGH (right) + Posterolateral SGH (left)	292	14	Intermittent stridor
14	At birth	F	Stridor	Yes (beard)	4	Posterolateral SGH	110		ACI
15	At birth	F	Stridor	Yes (beard)	5	Left hemicircumferential SGH	14		Intermittent stridor

Score of Orlow: locations of haemangioma concern preauricular region (uni or bilateral), lower lip, chin and anterior portion of the neck. The score is on the basis of the number of regions involved. Patients with four or five regions involved had a beard-like distribution.

*In remission: SGH is still visible during endoscopy but without clinical signs.

**PHACE(S) syndrome [posterior fossa brain malformations, haemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, eye abnormalities, sternal non-union].

NED=no evidence of disease (SGH is not any more visible during endoscopy); ACI=adrenocortical insufficiency.

distribution. In both groups, symptoms may be present at birth or begin before 3 months of age. Most of the children with haemangiomas in a beard distribution were girls (83%). The mean daily dose of prednisone per kg was not significantly different between groups. Evaluation of these groups in relation to severity of symptoms, treatment modalities and prognostic outcomes did not reveal any significant difference. The low number of children in each group did not allow a level of statistical significance to be reached, but a multicentric study could do so.

Early detection of children with facial haemangioma distributed in a beard pattern complaining of stridor is highly important because these children appear to be at risk of developing symptomatic airway haemangiomas.

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