Airway Hemangiomas in PHACE Syndrome

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ABSTRACT

OBJECTIVE: To describe the clinical presentation and airway characteristics of infants with airway hemangiomas and concomitant PHACE syndrome and to determine the prevalence of airway hemangiomas in PHACE subjects at our institution.

STUDY DESIGN: Case series

METHODS: Retrospective review including clinical presentation, airway findings, treatment measures, and outcomes

RESULTS: A total of 23 subjects were diagnosed with PHACE at our institution between September 1, 2005 and September 1, 2011. Twelve (52%) of these subjects had documented airway hemangiomas, six of whom were diagnosed and treated at our institution. All six subjects underwent direct laryngoscopy and bronchoscopy by a pediatric otolaryngologist.

Five (83%) subjects had subglottic hemangioma. Three subjects (50%) had additional hemangioma within the airway. Five subjects (83%) were treated with propranolol, five (83%) were treated with systemic steroids, and one subject received vincristine. One subject required laser ablation of subglottic hemangioma and tracheotomy. All subjects were airway symptom free at last follow up.

CONCLUSIONS: Airway hemangiomas can be a life threatening complication of PHACE syndrome. At our institution, 52% of all PHACE subjects were diagnosed with airway hemangiomas. Early detection of airway involvement is paramount. Given the high rates of airway involvement, we recommend performing direct laryngoscopy and bronchoscopy in all PHACE patients with respiratory symptoms. We also recommend having a low threshold for airway evaluation in asymptomatic PHACE patients, especially those who will not be otherwise started on propranolol.

REFERENCES


Table 1. Clinical characteristics of subjects