Iceberg hemangioma: A segmental cutaneous lesion marking extensive extracutanous involvement

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CASE 1

A 3-month-old white girl presented with a soft, bluish scalp mass extending from the posterior hairline to the occipital scalp, just right of midline (Fig 1, *A* and *B*). She also had stridor, plagiocephaly and decreased neck range of motion. After failure to respond to treatment for suspected croup, plagiocephaly and decreased neck range of motion prompted a workup for an invasive infantile hemangioma. Direct laryngotracheoscopy and computed tomography angiography (CTA) identified subglottic narrowing (Fig 2, *A*). Magnetic resonance imaging (MRI) of the head, neck, and upper thorax found additional multifocal extracutaneous lesions (Table I and Fig 1, *C*). Skin biopsy from a neck site found histologic findings typical of infantile hemangioma. The baby had a mixed, segmental hemangioma diagnosed. PHACE syndrome (Posterior fossa, Hemangioma, Arterial abnormalities, Cardiac abnormalities, Eye abnormalities, Sternal Cleft) was considered, but a thorough workup, including echocardiogram, ophthalmology evaluation, and CTA ruled out any intracranial, arterial, cardiac, or ophthalmic abnormalities. Abdominal ultrasound scan helped rule out any other visceral involvement. The patient was started on oral propranolol at 0.3 mg/kg every 8 hours, increased over 3 days to 1.0 mg/kg every 8 hours. Stridor resolved within 1 week, and repeat laryngotracheoscopy confirmed a significantly increased airway diameter (Fig 2, *B*). At 1 month follow-up, the cutaneous component of her hemangioma was less bulky, and neck range of motion and plagiocephaly were dramatically improved.

Abbreviations used:

CTA:	computed tomography angiography
IH:	infantile hemangiomas
LUMBAR:	lower body hemangioma and other cutaneous defects, urogenital anomalies, ulceration, myelopathy, bony deformities, anorectal malformations, arterial anomalies, and renal anomalies ()
MRA:	magnetic resonance angiogram
MRI:	magnetic resonance imaging
PELVIS:	perineal hemangioma, external genitalia malformations, lipomyelomeningocele, vesicorenal abnormalities, imperforate anus, skin tag
PHACE:	posterior fossa, hemangioma, arterial abnormalities, cardiac abnormalities, eye abnormalities, sternal cleft

CASE 2

An 18-week-old white girl presented for evaluation of a small bump on the scalp, which was first noted shortly after birth but rapidly enlarged interfering with head position when supine. The soft, 11- × 15-cm bluish mass had a central area of pallor with

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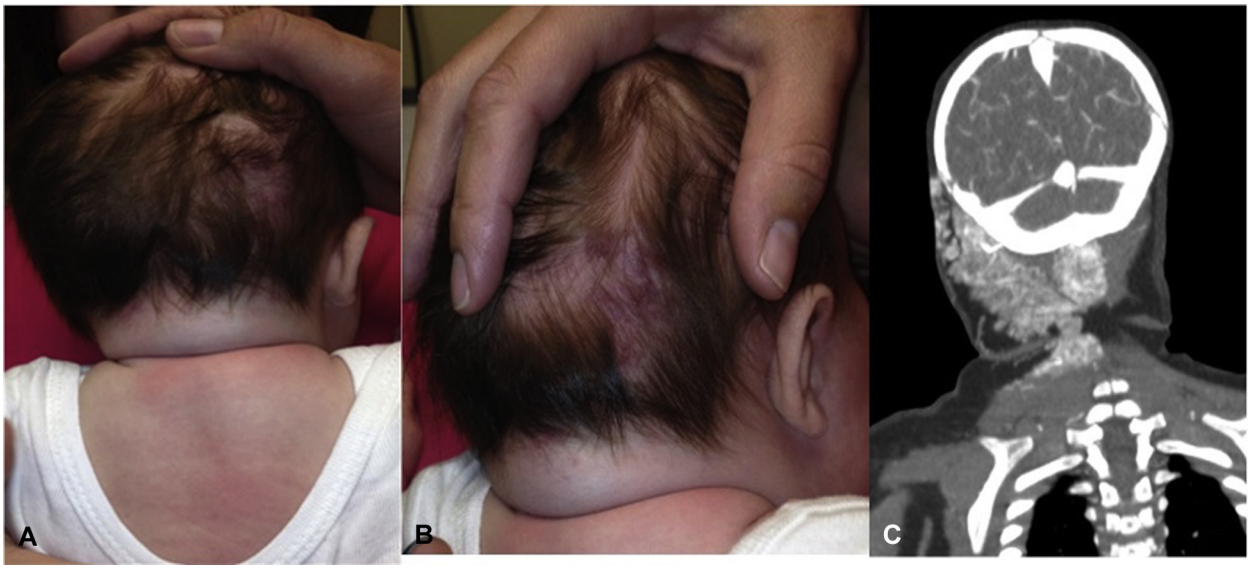

Fig 1. **A**, Soft bluish hemangioma of posterior scalp and nuchal area on physical examination. **B**, Oblique view of soft bluish hemangioma on the scalp and posterior neck. **C**, Entire posterior view with MRI showing diffuse multifocal subglottic and mediastinal involvement.

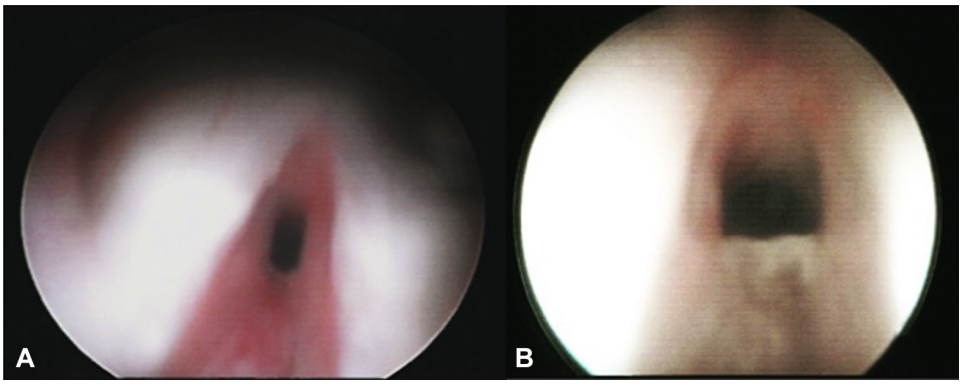

Fig 2. **A**, Bronchoscopy shows life-threatening airway compromise caused by paratracheal hemangioma. **B**, Significant increase in size of airway after 7 days of propranolol.

fine telangiectasia and extended from the nape to the occiput with associated hair thinning. There was no palpable thrill. A similar 3-cm soft bluish mass was noted on the midback. MRI found extensive hemangiomas involving the paratracheal space and the paraspinal area with involvement of the neural foramina and epidural space causing spinal cord deviation (Table I). Oral prednisone yielded 50% reduction of the cutaneous component. Repeat MRI at 15 months found similar partial involution of the extracutaneous lesions. Prednisone was gradually tapered and discontinued at age 17 months. At age 9 years, her cutaneous examination was still notable for a 9- × 7-cm soft, mobile mass at the nape and inferior scalp without overlying skin changes. MRI found evidence of fat with some increased vessels. Surgical debulking was planned, as propranolol was not standard therapy when this child presented.

CASE 3

An 8-week-old white girl presented to the emergency room 3 times within 1 week for evaluation of progressive stridor and respiratory distress that did not respond to oral corticosteroids, racemic epinephrine, and inhaled albuterol. Her skin examination featured a soft, blue 1- × 0.6-cm papule on the right upper eyelid and a 1.0- × 2.5-cm pink patch with central bright red 2-mm papules on her occipital scalp, left of midline. Airway symptoms prompted bronchoscopy, which found a subglottic mass occupying 25% of the airway with extension down to the posterior wall of the trachea and carina. MRI and magnetic resonance angiogram (MRA)