Head and Neck Vascular Anomalies

A Practical Case-Based Approach

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Preface

A better understanding of the nature and source of vascular anomalies has vitalized an interest in this field among numerous disciplines. The language used to describe these lesions is now coherent across specialties and allows for treatment algorithms to be unified. However, each vascular tumor and malformation has a unique management profile based on its type, size, and location as well as disciplines involved. Head and neck vascular anomalies are no exception to this rule and are the subsequent motivation behind this text.

We designed *Head and Neck Vascular Anomalies: A Practical Case-Based Approach* with the goal to provide hands-on, step-by-step, management algorithms for specific vascular anomalies of the head and neck encountered in daily practice. This is a condensed, multidisciplinary, practical guide for both simple and complex lesions. Our colleagues in otolaryngology, dermatology, pediatric surgery, plastic surgery, oncology, and interventional radiology have all contributed amazing cases with clinical detail, scientific evidence, and therapeutic options.

In each chapter, the initial steps to diagnose a vascular lesion are followed by a recommended treatment in a case-based format with photographs, radiographic imaging, and alternative therapies. All cases are based upon current literature with the aim to give state-of-the-art information on the majority of head and neck vascular anomalies. Medical, radiographic, and surgical techniques for frequently encountered and more difficult vascular anomalies are described.

This text is designed to be a reference guide. As you will see, each case follows a consistent and relatively rigid presentation outline. This style is meant to provide clarity, brevity, and simplicity to the reader. As a result, redundancies may be encountered for similar anomalies. For this we apologize, but frankly, we did not design the text to be read from cover to cover. Actually, we hope the reader can simply turn to a chapter and capture a complement of knowledge required to help their specific patient.

Of note, we also did not filter out any author or discipline bias in the chapters. In essence, the authors were allowed to express their opinion and therapeutic approach to their assigned case with the requirement to provide treatment alternatives. This decision was made to maintain the authenticity of opinion that is frequently found in the multidisciplinary field of vascular anomalies.

We, thereby, humbly submit to you *Head and Neck Vascular Anomalies: A Practical Case-Based Approach.* With an increasing number of vascular anomaly centers, patients, and interest in the field, we hope you find this text important to your everyday practice and a valuable aid for your patients.

—Gresham T. Richter and James Y. Suen

1

Infantile Hemangiomas

BASIC TENANTS AND INTERVENTIONS

Gresham T. Richter

Basic Tenants

Infantile hemangiomas (IHs) are the most common vascular tumor. They are composed of proliferating immature endothelial cells that express histologic marks found on placental blood vessels (GLUT-1, Lewis Y Antige, FcyRII, and merosin).[1] IHs are thought to be sporadic events although family lineage has been reported.[2] Coincidentally, IHs are also the most common tumor of infancy and are present in approximately 5% of the population.[3] They have a higher prevalence in females, Caucasians, and premature, and low birth weight infants.[4] They also occur more frequently in infants from mothers with early trimester bleeding, preeclampsia, and placental anomalies.

Infantile hemangiomas are rarely present at birth but early blanching or macular erythema of the skin may be a precursor to their later development. They may present anywhere on the body but involve the head and neck in over 60% of cases. Eighty percent of IH grow within the first 3 months of life and continue to grow up to 1 year of age.[5] IHs undergo predictable proliferative, quiescent, and involution

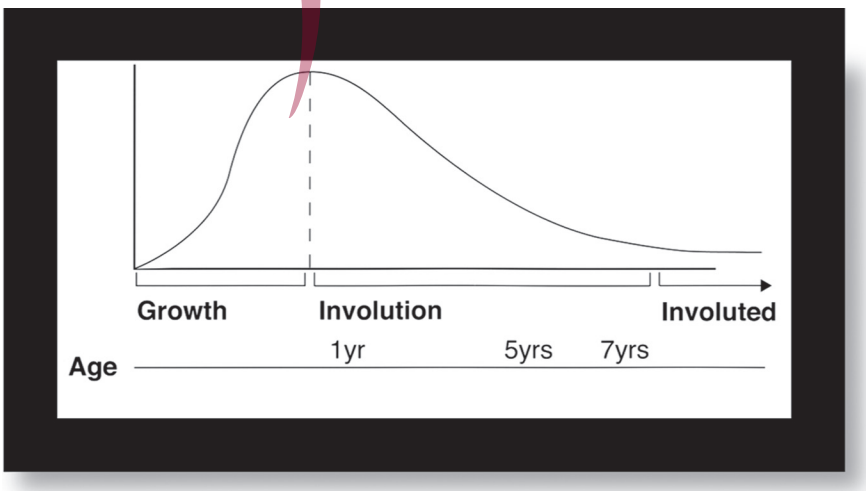

Figure 1–1. Typical growth phases of infantile hemangiomas.

phases as seen in Figure 1–1. The majority of IHs are thought to involute completely by 7 years of age. This natural history can help the clinician differentiate IHs from other congenital lesions and guide management decisions.

The classification of IHs is rather complex. They are first determined to be either focal or segmental. Focal IHs have discrete borders and further characterized as either superficial, deep, or compound. Early nomenclature has been supplanted by this new terminology to describe the majority of IHs (Table 1–1). Superficial and compound hemangiomas present with dark red cutaneous staining in a cobblestone pattern. Compound hemangiomas contain a subcutaneous component whereas deep hemangiomas do not involve the skin and present as a protuberance with an overlying blue skin discoloration (Figure 1–2). It is extremely rare for a focal IH to involve muscle or penetrate beyond subcutaneous fat. An exception is a parotid IH, the most common nonepitheliod tumor of the gland, which is frequently deep.

Problematic focal IHs typically involve the lip, eyelid, orbit, and subglottis where aesthetic and functional problems occur during the rapid proliferative phase. Sixteen percent of infants with 5 or more focal IHs will also have hepatic involvement and should undergo abdominal ultrasound.[6] Segmental IHs have a more complex growth pattern that their focal counterpart. In the head and neck, segmental IHs follow a trigeminal nerve (V) distribution. They are diffuse, compound, and maintain irregular borders. More than one facial subunit is frequently involved. They usually penetrate into deep fascial planes of the head and neck. The beard distribution IH (V3) is most commonly described.[7] These involve the lower lip, chin, neck, and preauricular areas and are frequently accompanied with ulceration. Sixty-three percent of segmental beard distribution will involve the subglottis and require airway endoscopy. All patients with segmentally distributed IHs should undergo systematic evaluation for PHACES (posterior fossa malformations, hemangiomas, arterial lesions, cardiac abnormalities, eye abnormalities, sternal cleft) syndrome.

The cause of IHs remains unclear but is postulated to either be ectopic placental tissue or an endo-

Table 1–1. Old and New Nomenclature for Infantile Hemangiomas

Old Nomenclature	New Nomenclature
Strawberry or Capillary Hemangioma	Superficial Hemangioma
Cavernous Hemangioma	Deep Hemangioma
Capillary Cavernous Hemangioma	Compound (Mixed) Hemangioma

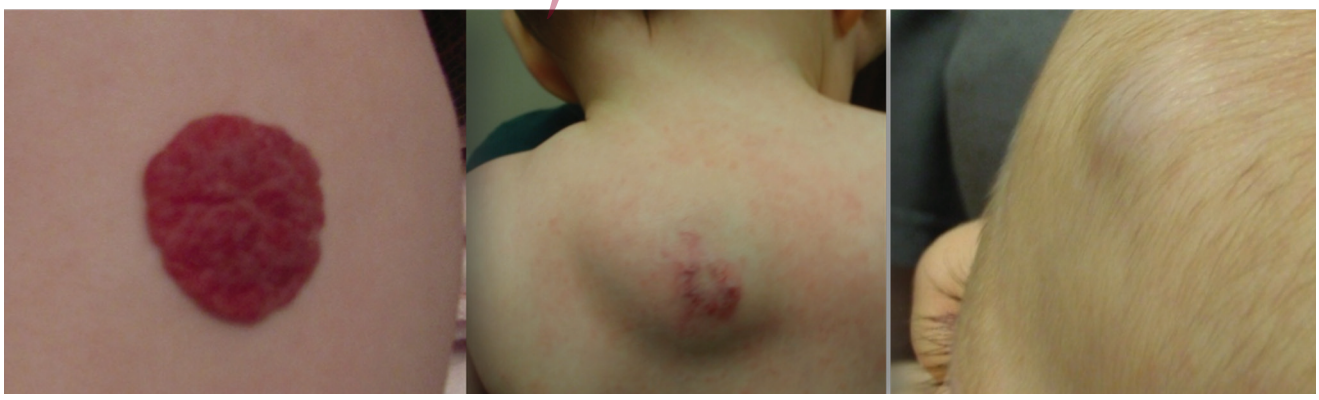

Figure 1–2. Focal hemangiomas described as superficial, compound, or deep (*left to right*).

thelial progenitor stem cell.[8] IHs are not associated with increased morbidity or mortality except in the very large hemangiomas that may rarely cause high output heart failure.

Intervention

Because of their natural involution, IHs were historically managed with observation alone. Although many resolve spontaneously others will cause significant functional and disfiguring consequences. Problematic hemangiomas are defined as those leading to significant events affecting the future life of the child. Most problematic events from IHs occur during the proliferative phase and include ulceration, bleeding, pain, vision disturbance, airway compromise, and feeding difficulties. However, late and deforming sequelae also occur to include scarring, telangiectasias, and fibrofatty residuum (Figure 1–3). Many cease to improve after 4 years of age and up to 69% of IHs will leave residual lesions.[9] At least 10% of IHs persist beyond 9 years of age. The age of self-recognition occurs around 4 years of age and must be considered in the treatment hemangiomas during their early phase of growth. Although it is difficult to predict future consequences for each lesion, early observation for rapid growth, protuberance, segmental disease, and functional compromise will help guide appropriate therapy.

Both surgical and medical interventions are available in the treatment of IHs. These include surgical excision, laser therapy, topical therapy, intralesional corticosteroids, systemic corticosteroids, systemic beta-blockers, and vincristine chemotherapy. Each of these therapeutic modalities is discussed in the following case presentations. Every IH has a unique profile that governs its treatment and is typically based on location and risk of aesthetic and functional compromise. Management during the proliferative phase generally will lead to the best final outcome. However, many IHs require multimodal therapy of which the final treatment occurs during the involution period. Absolute indications for early intervention include an impact on vital structures, active or impending functional impairment, the possibility of permanent scarring, large segmental facial hemangiomas, and ulcerative lesions.

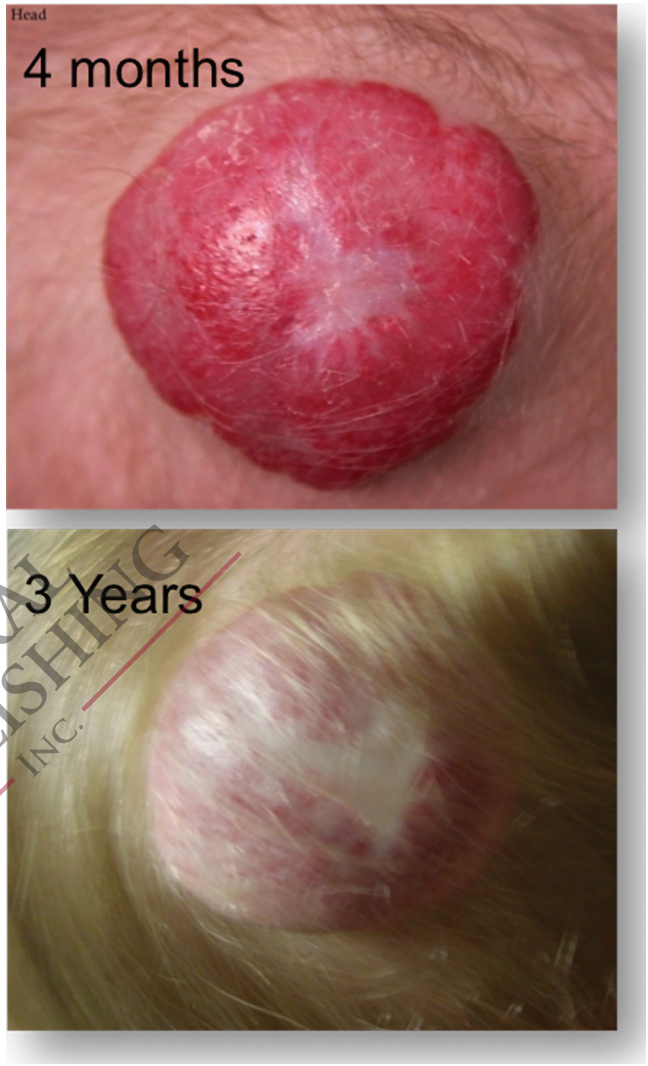

Figure 1–3. Focal scalp hemangioma at 4 months and seen again, untreated, at 3.5 years with resultant residuum that will require intervention.

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CASE STUDY 1–1. ANTERIOR NECK HEMANGIOMA

Abby R. Nolder

Representative Case

A 2-month-old, former 26-week preterm newborn male was referred to the pediatric otolaryngology clinic for evaluation of middle ear pathology following a failed newborn hearing screen. During that visit, the patient's mother expressed concern about a growing mass under the child's chin. It was not present at birth but had been rapidly progressing over the last several weeks. He had a history of intubation for 2 days in the neonatal intensive care unit but had no associated airway symptoms. He was having some feeding difficulties that seemed to be worsening as the mass increased in size.

On physical examination, he was found to have a 4-cm, soft, mobile, cystic appearing submental neck mass with faint blue discoloration of the overlying skin (Figure 1–4). He also had a 3 × 4-cm compound, pedunculated hemangioma on the right posterior scalp without bleeding or ulceration (Figure 1–5). No other lesions were discovered elsewhere on his body. He had mild stertor at rest without significant retractions or increased work of breathing; however, work of breathing increased during bottle feeding resulting in spillage of formula from his mouth.

Overview

Hemangiomas are common vascular tumors, occurring in up to 10% of children.[1] They grow rapidly during the first year of life and depending on the anatomic location can cause significant functional and cosmetic impairments. Hemangiomas of the neck should be managed based on the size and symptoms (eg, ulceration, bleeding) of the lesion. Rapidly growing tumors of the anterior neck can cause compression and result in airway and feeding difficulties in young infants; therefore, prompt

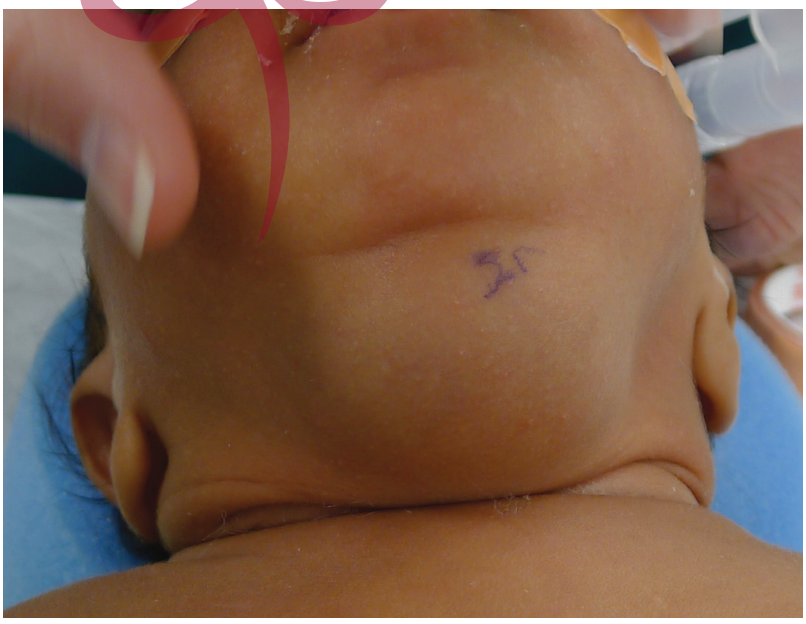

Figure 1–4. Midline anterior neck hemangioma.