# Pediatric Vascular Tumors of the Head and Neck

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**KEYWORDS**

• Hemangiomas • Vascular • Lesions • Pediatric

**KEY POINTS**

- There is confusion regarding the classification of vascular neoplasms and malformations of the head and neck.
- An incorrect diagnosis of hemangioma is often posed.
- The diagnosis of a vascular tumor is mostly based on history and physical examination.
- Giant cell lesions, pyogenic granulomas, and aneurysmal bone cysts are not vascular neoplasms per se, but they have a prominent vascular component.
- Other rare pediatric vascular lesions include hemangioendothelioma, tufted angioma, and juvenile nasopharyngeal angiofibroma.

## INTRODUCTION

Vascular tumors of the head and neck are commonly seen in the pediatric population. Oral and maxillofacial surgeons are often involved in the treatment of these children, particularly if there is bony involvement, or an association with the dentition. The diagnosis and treatment of vascular tumors, particularly those of the head and neck, have been hampered by a confusing nomenclature. An incorrect diagnosis of hemangioma is often posed, even in the presence of lesions that do not regress.

Mulliken and Glowacki[1] suggested a simplified classification based on endothelial characteristics. Vascular lesions are divided into 2 categories: vascular tumors and malformations. Hemangiomas are tumors of endothelial cells, whereas vascular malformations are the result of a structural anomaly of the blood vessels with normal endothelial cells. A detailed classification, based on these broad categories, is also available through the International Society for the Study of Vascular Anomalies (ISSVA Classification of Vascular Anomalies 2014, issva.org/classification).

Although other vascular tumors are rare in children, oral and maxillofacial surgeons may encounter them frequently. The giant cell lesion (GCL), pyogenic granuloma, and aneurysmal bone cyst (ABC) are not considered true vascular neoplasms, but have a strong vascular component, and therefore, they will be discussed here.

## HEMANGIOMA

### *Epidemiology*

Hemangiomas are the most common vascular lesions of infancy and are found in the head and neck in 40% to 60% of cases.[2,3] The incidence is between 3% and 10% by the age of 1 year and is more common in premature infants, girls, and Caucasians.[4–6] Skin lesions are the most frequent

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area of involvement, but these can be seen in internal organs, such as the liver, brain, and gastrointestinal tract (Figs. 1 and 2).[7] They are usually solitary, but multiple lesions are seen in 20% of cases.[2] If more than 4 cutaneous lesions are found, it is suggested to assess for internal lesions. Hemangiomas of the liver can result in life-threatening bleeding.[2] Large lesions of the face combined with other vascular anomalies should prompt suspicion of PHACE syndrome (*P*osterior fossa malformations, *H*emangiomas, *A*rterial lesions, *C*ardiac abnormalities [coarctation of the aorta], and *E*ye anomalies).[2,3,8]

### *Origin*

Hemangiomas are composed of hyperplastic endothelial cells exhibiting increased mitotic activity.[1] The cause remains incompletely understood, although an autosomal transmission is presumed.[7] Two proangiogenic factors are involved during the proliferative phase: fibroblast growth factor (FGF) and vascular endothelial growth factors (VEGF),[9] which is important because it explains, in part, the efficacy of β-adrenergic receptor-blocking agent to treat these lesions (see Treatment section).[10]

### *Evolution*

Cutaneous hemangiomas may not be visible at birth, but can present as a subtle erythematous macula or papule in up to 40% of cases.[1,11] They become more apparent within the first 6 weeks of life and then undergo a proliferative phase during the subsequent year.[11] Some lesions may have a late proliferative phase, starting later and lasting longer. During this phase, increased endothelial activity and hyperplasia are noted. After proliferation, involution of the tumor occurs over the next several years (up to age 7), a phenomenon not observed with vascular malformations.[11] At this stage, cellular activity is characterized by apoptosis of endothelial cells and decreased angiogenesis.[1] The abnormal vascular channels are replaced with fat and fibrous tissue. Different phases may be present within the same lesion. A lesion completely mature at birth that does not undergo a proliferative phase is called a congenital hemangioma.

### *Clinical Appearance*

The clinical appearance may vary depending on the depth of the tumor. The classic bright red strawberry-like appearance may not be evident if the lesion is situated deeper into the subcutaneous tissues. It can then be misdiagnosed as a vascular malformation, with the overlying skin exhibiting a normal appearance. In the past, superficial lesions were referred to as capillary hemangiomas, and deep lesions were referred to as cavernous hemangiomas.[1] These terms should be reserved for histologic description, because they have no bearing on clinical behavior or treatment. Tumors combining superficial and deeper parts (compound) are also encountered.

In a retrospective chart review of 445 children referred for biopsy of cervical masses, Torsiglieri

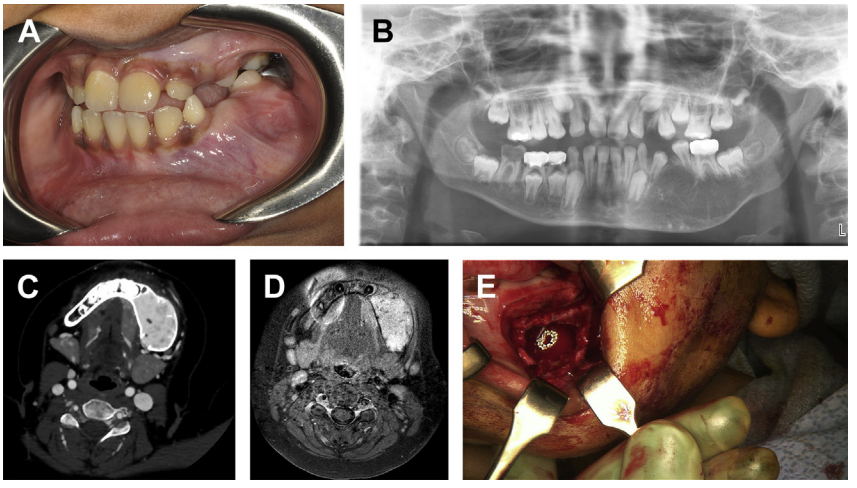


**Fig. 1.** Intraoral photograph (*A*) of an 11-year-old male patient referred for a vestibular mass of the left mandible. The panoramic radiograph (*B*) shows a poorly defined expansile multilocular lesion with displacement of teeth. Computed tomography angiography (*C*) and MRI (*D*) confirmed the suspected diagnosis of intraosseous hemangioma. The lesion was successfully embolized and enucleated (*E*) without complications or recurrence.