Congenital Vascular Tumors

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KEYWORDS

• Vascular tumors • Pyogenic granuloma • Congenital hemangioma
• Kaposiform hemangioendothelioma • Tufted angioma • Infantile myofibroma
• Epithelioid hemangioendothelioma • Enzinger intramuscular hemangioma
• Angiosarcoma • Cutaneovisceral angiomatosis with thrombocytopenia

KEY POINTS

- Vascular tumors and malformations comprise the field of vascular anomalies.
- Although the majority of tumors affect the skin, lesions involving the mucous membranes have been noted.
- Tufted angioma, or angioblastoma of Nakagawa, shares clinical and histopathologic features with kaposiform hemangioendothelioma, suggesting these vascular tumors exist together along a spectrum.
- Fewer than 1% of soft tissue sarcomas are classified as angiosarcoma and only 1% of angiosarcomas affect children.

INTRODUCTION

Vascular tumors are benign neoplasms, which result from proliferating endothelial cells. These lesions present during infancy or childhood, may affect any location, and exhibit postnatal growth. Local complications include bleeding, tissue destruction, and pain whereas systemic sequelae include thrombocytopenia, congestive heart failure, and death. Vascular tumors should be differentiated from vascular malformations, which present at birth, have a quiescent endothelium, and grow in proportion to the child. Together, vascular tumors and malformations comprise the field of vascular anomalies.

Infantile hemangioma is the most common vascular tumor of childhood. For more information on this topic, see Denise M. Adams's article, "Infantile Hemangiomas in the Head and Neck Region", Marcelo Hochman's article, "The Role of Surgery in the Management

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of Infantile Hemangiomas: What is the Best Timing?" and Milton Waner's article, "The Surgical Management of Infantile Hemangiomas" in this issue. Other congenital tumors, in order of most common, include pyogenic granuloma (PG), congenital hemangioma (CH), kaposiform hemangioendothelioma (KHE), tufted angioma (TA), infantile myofibroma (IM), epithelioid hemangioendotheloma (EHE), Enzinger intramuscular hemangioma, angiosarcoma, cutaneovisceral angiomatosis with thrombocytopenia (CAT).

PYOGENIC GRANULOMA

A small, solitary, red, bleeding or ulcerated lesion that presents in childhood (**Fig. 1**) is pathognomonic for PG, formerly *lobular capillary hemangioma*. These tumors are small compared with other vascular tumors, with 75% measuring less than 1 cm in diameter.[1] PGs are twice as common in boys and the mean age of onset is 7 years; only 12% develop during the first year of life. Although a majority of tumors affect the skin (88%), lesions involving the mucous membranes (12%) have been noted. Lesions affect the head and neck (62%), trunk (19%), and extremities (18%). Within the head and neck, specific sites include cheek (29%), oral cavity (14%), scalp (11%), forehead (10%), eyelid (9%), or lips (9%).[1] Complications include bleeding (64%) and ulceration (36%). Twenty-five percent of patients have a history of preexisting trauma or underlying cutaneous conditions, such as a capillary malformation or arteriovenous malformation. History and physical examination are sufficient for diagnosis; imaging is unnecessary.

With bleeding and crusting, PG may shrink, but regrowth is common. Treatment with curettage, shave excision, or laser therapy is ineffective because the lesion extends into the reticular dermis, which is often inaccessible by these approaches.[1,2] These modalities have recurrence rates as high as 50%.[1,2] Cure rate of approximately 100% is achieved after full-thickness skin excision.[1,2] Alternatively, the lesion can be shaved and the base cauterized with a Bovie through the thickness of the dermis.

CONGENITAL HEMANGIOMA

CHs are solitary, red-violaceous lesions with coarse telangiectasias, central pallor, and peripheral pale halos. The incidence of CH is currently unknown. CH more commonly

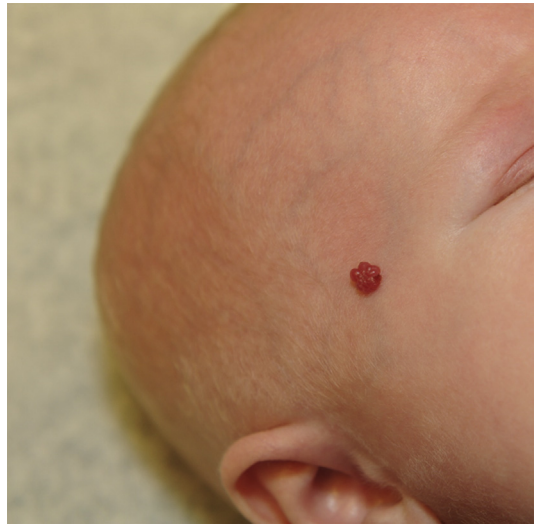

Fig. 1. Two-month-old boy with PG of the face.