

ACRAL ARTERIOVENOUS TUMOR

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Abstract. A unique, superficial, lobulated, benign, vascular tumor of the skin, found in the acral areas of adult males, is described. The combination of three different elements—arterial, venous, and transitional vascular channels—make up the predominant histologic features of this tumor. The transition of fibromuscular channels indicates a venous lesion, but the acral location of the lesions and the structure and staining characteristics suggest that the lesion is a hamartomatous proliferation of the Sucquet-Hoyer canal of the true glomus.

Key words: Acral tumor; Benign vascular tumor; Hamartomatous proliferation; Skin tumor; Vascular tumor

Benign vascular tumors are usually classified as arterial or venous, capillary or cavernous, and hemangioma (endothelioma) or hemangiopericytoma. Some vascular tumors such as leiomyoma, glomus tumor, and multiple glomus tumor are distinctive because of the amount of smooth muscle in the vascular wall. Our recent study of a special benign vascular lesion indicated that three elements are present: an arterial, a venous, and a connecting transitional element that resembles a hamartomatous proliferation of the Sucquet-Hoyer canal. The discovery that the distribution of these lesions was predominantly acral prompted us to suggest "acral arteriovenous tumor" as the name for this lesion and to describe the clinical and histologic pattern of this unique vascular mass.

MATERIAL AND METHODS

Fifteen cases of acral arteriovenous tumor originally diagnosed as hemangioma or venous hemangioma comprise the series. Sections of tissue stained with hematoxylin and eosin were available for review in all cases. In addition, sections were stained with alcian blue and periodic acid-Schiff, Masson trichrome, aldehyde fuchsin, elastic Giemsa, and Bodian. The clinical records were reviewed in all cases.

CLINICAL DATA

The mean age of the 15 patients (12 men and 3 women) at diagnosis was 58 years, ranging from 27 to 83 years. The patients' occupations were diverse and had no apparent relationship to the lesion.

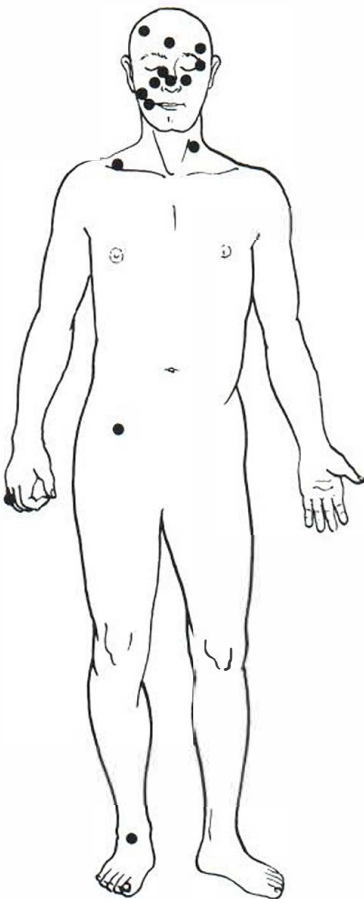


Fig. 1. Anatomic distribution of acral arteriovenous tumor.



Fig. 2. Elevated, dark-red papule of acral arteriovenous tumor.

Ten patients had the lesion on the skin of the face and scalp (Fig. 1). One lesion was located on the neck, one on the clavicular area, one on the dorsum of the finger, one on the dorsum of the foot, and one on the skin of the abdomen. The sizes of the lesions ranged from 3 mm to 4 cm. Most of the lesions were less than 1 cm in diameter. The largest tumor was located in the neck.

Nine lesions were described as elevated dark-red papules (Fig. 2); five lesions were nodular or tumoral, and one was verrucous. No pain or bleeding was noted in any case.

Most of the lesions had been present for between 6 months and 6 years. In 3 patients, the lesions were present for 25, 20, and 10 years and appeared when the patients were 33, 42, and 29 years old, respectively.

The common clinical diagnoses of these tumors were hemangioma, nevus, pyogenic granuloma, and Kaposi's sarcoma. No associated significant diseases or tumors were observed.

HISTOPATHOLOGIC FINDINGS

The lesions presented as circumscribed growths of large, mature-appearing blood vessels in the superficial and middle dermis, without evidence of infiltration into the deep dermis and subcutaneous tissue (and with no suggestion of capsule formation). The vascular mass was separated from the epidermis by a rim of normal connective tissue (Fig. 3).

The tumors were formed by a proliferation of thick-walled, transitional, vascular channels lined by a single-cell layer of endothelial cells and with a wall rich in fibrous cells. The wall-to-lumen ratio and the histologic features superficially resembled those of venous blood vessels. In cross section the channels were piled atop one another in the upper and middle dermis. No internal elastic lamina was present. The fibromuscular wall was deceptive, because at low power it appeared to be smooth muscle, but at high power and with special stains, it was apparent that it was largely fibrous, with occasional round cells or smooth muscle cells. The elastic stain showed fine fibrillar staining throughout the wall. The wall had a minute muscle component, as revealed by trichrome staining. In addition, an arterial component was found in all the tumors, represented by a small tortuous artery with a PAS-positive wall and an internal elastic lamina detected by elastic Giemsa staining (Fig. 4). Typical veins were observed at the periphery of the tumor.

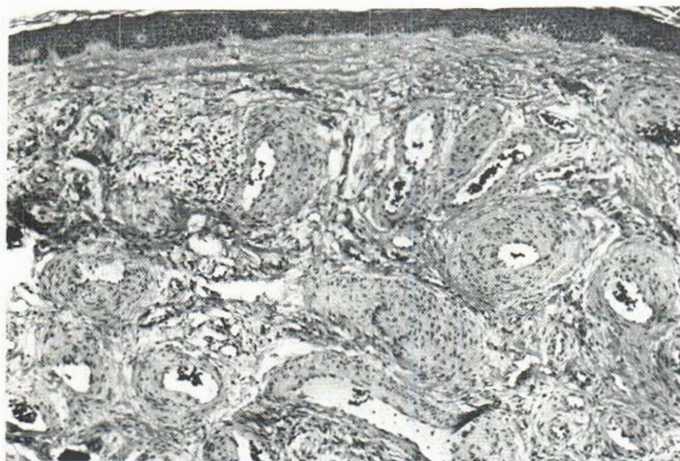


Fig. 3. Circumscribed growth composed of arteriovenous blood vessels in superficial and middle dermis. (Hematoxylin and eosin; $\times 54$).

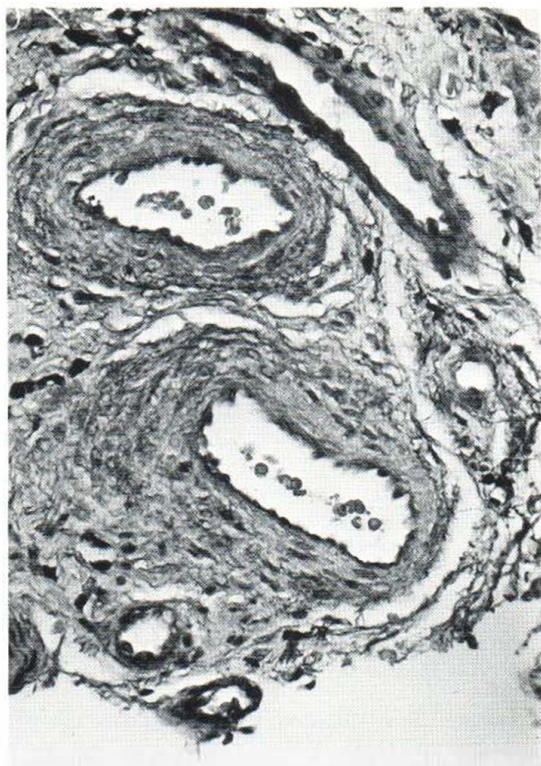


Fig. 4. Arterial vessels with demonstration of partial internal elastic lamina. (Aldehyde fuchsin-Giemsa; $\times 250$.)

Usually, the arterial channels were found in the periphery of the vascular mass, especially in the deep parts of the tumors. Only in four tumors were arterial vessels with elastic lamina seen, mixed with the venous blood vessels.

In nine lesions, round glomoid cells in the vascular wall were observed in the transitional channels. In four tumors, Bodian staining revealed nerve fibers in the stroma.

In addition to the large or small arteriovenous vessels, all lesions contained lobular masses of capillary dilatation, similar to the senile angioma, in the upper dermis.

The overlying epidermis was normal, but in 3 patients, hyperkeratosis was noted. Six patients had prominent dermal changes of actinic elastosis. In 9 patients, no elastic fibers could be detected in the dermal tissue of the tumor area.

Some lymphocytic infiltration was seen in 4 cases, particularly around the capillary vessels beside the tumor. No sclerotic changes were noted around the blood vessels of the tumor. Mast cells

were found in areolar connective tissue surrounding the vessel masses. Minimal amounts of ground substance were present in the stroma of the tumor.

COMMENT

The term "arteriovenous tumor" already has been used to designate the vascular proliferation, multiple or single, whether related to vascular syndromes or not (2, 4, 10, 12, 13, 15, 16), and which demonstrates arteriovenous connections as well as vessel proliferations. Such tumors are usually located in the subcutaneous and deeper tissue, and they are described by different names, depending on the most prominent component (arterial or venous) or the tissues in which they develop (1, 6, 8, 9, 14).

In contrast, we present the clinical and histopathologic features in 15 cases of an unusual vascular tumor localized superficially in the skin and composed of a vascular mass of arterial and venous blood vessels with transitional vascular channels. These lesions are present in adults and are found most frequently but not exclusively in males, located predominantly on acral areas (face, hand, and foot).

We found no references in the literature concerning this form of vascular tumor, which suggests that no systematic study of this lesion exists because of its infrequent occurrence and its benignity and because such lesions are rarely biopsied. We believe that the usual diagnoses are senile angioma, hemangioma, so-called venous angioma (11), and benign pigmented or epithelial tumors; histologic verification is usually not sought.

The clinical presentation is not characteristic. The red papular or nodular form is the most frequent clinical appearance, and with the distribution on acral areas and the clinical appearance of the lesion, thrombosed capillary aneurysm (5) and venous lakes must be considered. Venous lakes have been described by Bean & Walsh (3) in 1956 as vascular macules or papules, but the lakes are a varix type of venous dilatation that are easily distinguishable histologically from the acral arteriovenous tumor. Clinically, the acral arteriovenous tumor is a single lesion with a long evolution and without enlargement, bleeding, or ulceration. We believe that the lesion can be diagnosed clinically but that the histopathologic features are truly and distinctively characteristic and diagnostic.

We believe that the acral arteriovenous tumor is similar to the arteriovenous shunt (7) in which there are arterial, venous, and transitional vessels, but the shunt is seen in deeper locations. The acral arteriovenous tumor is not the same as the glomus tumor, as the former is composed of lobules of complete channels, whether arterial, capillary, venous, or transitional. In the deeper part of the acral tumors, there are true arterial vessels that represent the afferent artery to the tumor. A few smooth muscle cells are present in the vessel walls of the acral tumor, but these cells are different from the typical glomus cells (smooth-muscle cells) that proliferate in the walls of the ill-defined vascular mass of the glomus tumor.

We believe that the transitional channels resemble the Sucquet-Hoyer canal of the glomus and that the acral arteriovenous lesion could represent hamartomatous proliferation of that particular structure. This is suggested by the presence of the thin-walled venous segment, of the transitional channels with a narrow lumen and thick wall without internal elastic membrane and very fine fibrillar elastic-staining fibers about the cells in the fibrovascular wall, and of true arterial segments in the deeper part of the tumor. The presence of the nerves in this lesion and its location in the middle and superficial part of the dermis, as well as its clinical presentation on acral areas (which was discovered by clinical review of these cases), are also confirmatory features. The acral arteriovenous tumor may ultimately be considered the true glomus tumor, or at least a glomus hamartoma.

REFERENCES

- Allen, P. W. & Enzinger, F. M.: Hemangioma of skeletal muscle. An analysis of 89 cases. *Cancer* 29: 8, 1972.
- Bean, W. B.: The arterial spider and similar lesions of the skin and mucous membrane. *Circulation* 8: 117, 1953.
- Bean, W. B. & Walsh, J. R.: Venous lakes. *Arch Dermatol* 74: 459, 1956.
- Enzinger, F. M., Lattes, R. & Torloni, H.: International Histological Classification of Tumours. No. 3. Histological Typing of Soft Tissue Tumours. World Health Organization, Geneva, 1969.
- Epstein, E., Novy Jr, F. G. & Allington, H. V.: Capillary aneurysms of the skin. *Arch Dermatol* 91: 335, 1965.
- Fu, Y.-S. & Perzin, K. H.: Non-epithelial tumors of the nasal cavity, paranasal sinuses, and nasopharynx. A clinicopathologic study. I. General features and vascular tumors. *Cancer* 33: 1275, 1974.
- Gray, H. R.: Arteriovenous shunt. In *Dermal Pathology* (ed. J. H. Graham, W. C. Johnson & E. B. Helwig). Harper & Row, New York, 1972.
- Jenkins, H. P. & Delaney, P. A.: Benign angiomatous tumors of skeletal muscles. *Surg Gynecol Obstet* 55: 464, 1932.
- Jones, K. G.: Cavernous hemangioma of striated muscle. A review of the literature and a report of four cases. *J Bone Joint Surg (Am)* 35: 717, 1953.
- Masson, P.: *Traité de pathologie médicale et de thérapeutique appliquée*, vol. 27, p. 164. A Maloine et Fils, Paris, 1923.
- Montgomery, H.: *Dermatopathology*, 1247 pp. Hoeber Medical Division, Harper & Row, New York, 1967.
- Naranjo, J. & Mogollon, G.: Hemangioma venoso. *Angiologia* 22: 235, 1970.
- Pack, G. T. & Ariel, I. M.: Treatment of Cancer and Allied Diseases: Tumors of the Soft Somatic Tissues and Bone, vol. 8, pp. 408-411, 2nd ed. Hoeber Medical Division, Harper & Row, New York, 1964.
- Scott, J. E. S.: Haemangiomata in skeletal muscle. *Br J Surg* 44: 496, 1957.
- Stout, A. P. & Lattes, R.: Tumors of the soft tissue. In *Atlas of Tumor Pathology*, Second series, Fasc. 1. Armed Forces Institute of Pathology, Washington, D.C., 1967.
- Winer, L. H. & Levin, G. H.: Acquired vascular tumors of the skin in the adult. A report of five unusual cases. *Arch Dermatol* 79: 17, 1959.

Received February 10, 1976

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