Volume twenty-two

Symposium on vascular malformations and melanotic lesions

Editor

H. Bruce Williams

Plastic Surgery Educational Foundation of the American Society of Plastic and Reconstructive Surgeons

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Symposium on vascular malformations and melanotic lesions

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Preface

This book is directed to the multiple medical and surgical disciplines involved with the clinical management of vascular malformations and melanotic lesions in children and adults. The in-depth approach to these two problem areas of patient care is designed to clarify the current controversies and treatment methods. The goals of this book are to assist in the treatment plan as specific patient problems arise and to give a balanced opinion as to their management.

The book is divided into the two major subjects of vascular malformations and melanotic lesions, with the various sections and chapters grouped in a logical sequence. Part I, Vascular Malformations, includes embryology, classification, pathogenesis, and prognosis. Specific diagnostic and treatment methods include the use of Doppler ultrasound, steroids, hypotensive anesthesia, and surgery. Newer techniques with superselective angiography and embolization are evaluated, and the promise for improved treatment in the future using these methods is clearly evident. The discussion of port-wine stains assesses current management of these problems with surgery, tattooing, and the increasing emphasis on argon laser therapy. For lymphatic abnormalities, a similar format includes management of the different malformations. Extremity problems are discussed as to asymmetry, macrodactyly, and gigantism, and recent techniques with lymphovenous anastomoses are assessed. Part II is devoted to melanotic lesions and includes the diagnosis and management of both benign and malignant conditions. The controversial topics of giant pigmented nevi and melanomas in children are discussed, and the clinical management of malignant lesions and their relationship to depth of invasion are evaluated. The current status of tumor markers and the clinical value of the leukocyte adherence inhibition (LAI) test in melanoma therapy are discussed, and other treatment methods such as lymph node dissection, perfusion, chemotherapy, and immunotherapy are evaluated as to their present use.

The panel discussions held at the end of each session are included because they were of considerable value to the content of the symposium. The succinct, carefully prepared presentations of this international faculty and the enthusiastic participation of those in attendance make this book a valuable educational tool.

I would like to express my appreciation to all members of the teaching faculty and to those participants at the symposium and also to express a special thanks to the members of the Educational Foundation Symposium Committee; to our Educational Coordinator, Ms. Victoria Doretti; and to Ms. Carol Larson-Lazier, who helped greatly in the many details of organization. Last, my appreciation to Miss Karen Berger of The C.V. Mosby Company and the staff who gave their usual valuable advice and exhibited great patience in the preparation of this book.

H. Bruce Williams

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Part I

Vascular malformations

Chapter 1

Embryology and classification of hemangiomas

Melvin Spira

DEFINITION OF TERMS

In this and the chapters to follow a wide variety of terms referring to tumors of blood vessels will be employed. A brief description of the terminology is in order.

Angioma is a tumor composed of either blood vessels or lymphatics. The former and most common is called a *hemangioma*; the latter, which is relatively rare, is called a *a lymphangioma.*

Hamartoma, or *vascular hamartoma,* comes from the Greek word "to err." It refers to a tumorlike, but primarily nonneoplastic, formation, an inborn error of tissue development characterized by an abnormal mixture of tissues indigenous to the part, with an excess of one or more of these tissues. Obviously, many hemangiomas will fit in this category.

Telangiectasias are a dilation of previously existing vessels. There is no associated new growth.

The word *nevus* actually comes from Latin and means a "birthmark" or a "blemish." It is occasionally used when referring to hemangiomas, as in nevus flammeus or vascular nevus.

Aneurysm refers to a circumscribed dilation of an artery. When the word "arteriovenous (AV)" is added, we refer to a blood-containing swelling connected with both an artery and a vein. When a direct communication between the two vessels exists without any intervening sac, the condition is called an *aneurysmal varix.* Where there is a sac between the two, it is called a *varicose aneurysm.*

A *cirsoid aneurysm* or *racemose aneurysm* is a dilation of a group of blood vessels due to a congenital malformation with AV shunting. The word *racemose,* or "full of clusters," histopathologically refers to the branching with nodular terminations that resemble a bunch of grapes seen in aneurysms. The word *cirsoid* refers to a varix, which again merely means one or more enlarged or tortuous veins, arteries, or lymphatic vessels.[8]

EMBRYOLOGY OF HEMANGIOMAS

Blood vessels first make their appearance in several scattered vascular areas that are developed simultaneously between the endoderm and the mesoderm, first in the yolk sac and a bit later in the body of the embryo. Here a new type of cell, the angioblast, or vasoformative cell, differentiating from the mesoderm, divides and forms small, dense masses that soon join other similar masses to form plexuses. Thus what will become the vascular tree appears as an interlacing or interconnecting system of blood spaces. These blood lakes contain mixed blood with no separate venous and arterial channels existing. This is the stage that Woollard[10] in 1922 and later Szilagyi and co-workers[9] described as a stage of undifferentiated capillary networks. This developing system (Fig. 1-1, *A*), which is from a 20-day pig embryo, would correspond to about the thirtieth day of development in the human. With development, the principal arterial stems become differentiated, and the blood space is either absorbed or partly coalesced. Separate venous and arterial conduits or channels appear on either side of the capillary network (Fig. 1-1, *B*). This is what we would see at approximately the forty-eighth day of development in the human; it is referred to as the retiform stage and consists of large, plexiform structures formed by coalescence of the original equipotential capillaries. The final stage, which involves the appearance of mature vascular stems after the disappear-

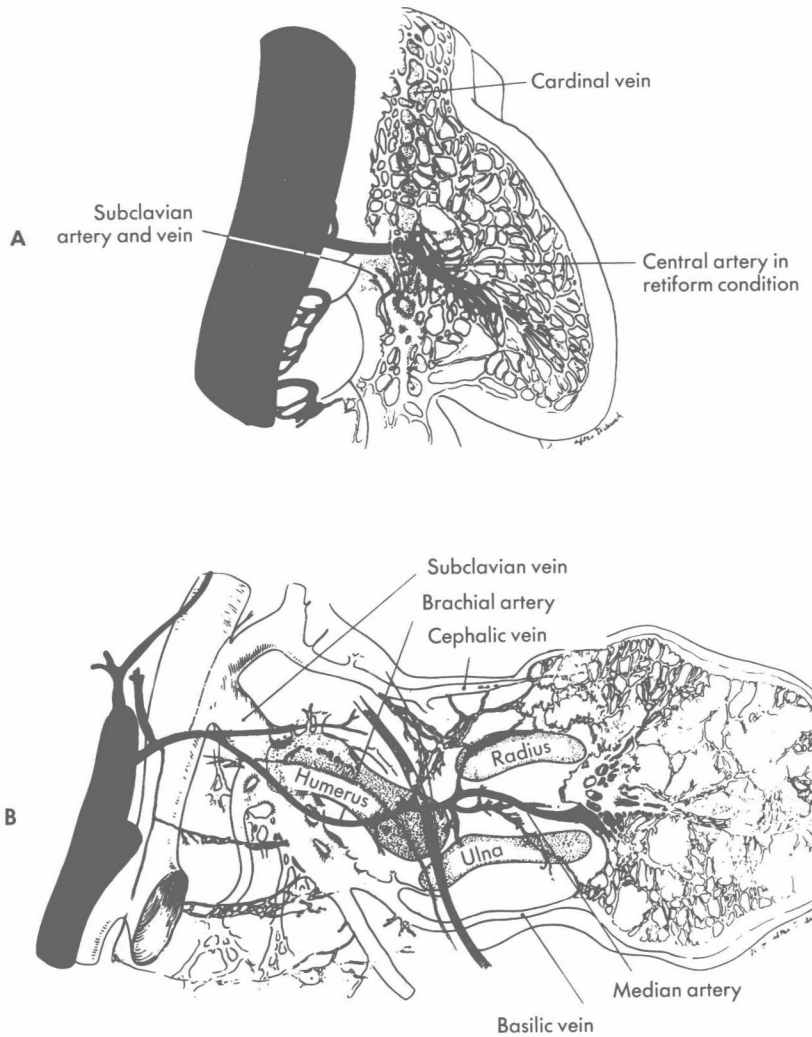

Fig. 1-1. Developing vasculature in the pig embryo. **A,** Retiform stage development (20-day pig embryo). **B,** Stage of gross differentiation (27-day pig embryo). (From Szilagyi, D.E., Elliott, J.P., DeRusso, F.J., and Smith, R.F.: Surgery **57:**61, 1965.)

ance of the primitive elements, occurs within the first 2 months of embryonic development.

Szilagyi and co-workers[9] have classified a group of peripheral congenital hemangiomas and AV fistulas according to the embryologic events just described (Table 1-1). Arrested development in the capillary network stage will typically result in formation of a capillary hemangioma. Where development was arrested in the retiform stage, microfistulas, or minute AV aneurysms, were formed. A clinical example for this category would be a cavernous hemangioma. Going slightly up on the scale where AV aneurysms are larger and contain macrofistulas, a further step in the retiform stage, we have typical AV malformations. When vascular channels develop anomalously, as after trauma, the vessels are mature and would be classified in the last group. In essence, the rest of the direction of normal development may take place at any stage and give rise to anomalous structures. Mulliken and others[7] have described Malan's classification,[6] which also is based on embryological development. A vascular malformation may either be a venous hemangioma, an AV hemangioma, an arteriovenous fistula, or a capillary hemangioma with individual variations (Fig. 1-2).

Table 1-1. Classification of hemangiomas (embryology)*

Designation	*Stage of embryologic development*	*Example*
Hemangioma	Capillary network stage	Cavernous hemangioma
Microfistulous AV aneurysm	Retiform stage	Capillary hemangioma
Macrofistulous AV aneurysm	Retiform stage	AV malformation
Anomalous mature vascular channels	Stage of gross differentiation (arterial stem formation)	Traumatic aneurysm

*Modified from Stedman's medical dictionary, ed. 22, Baltimore, 1972, Williams & Wilkins Co.; Szilagyi, D.E., Elliott, J.P., DeRusso, F.J., and Smith, R.F.: Surgery **57**:61, 1964.

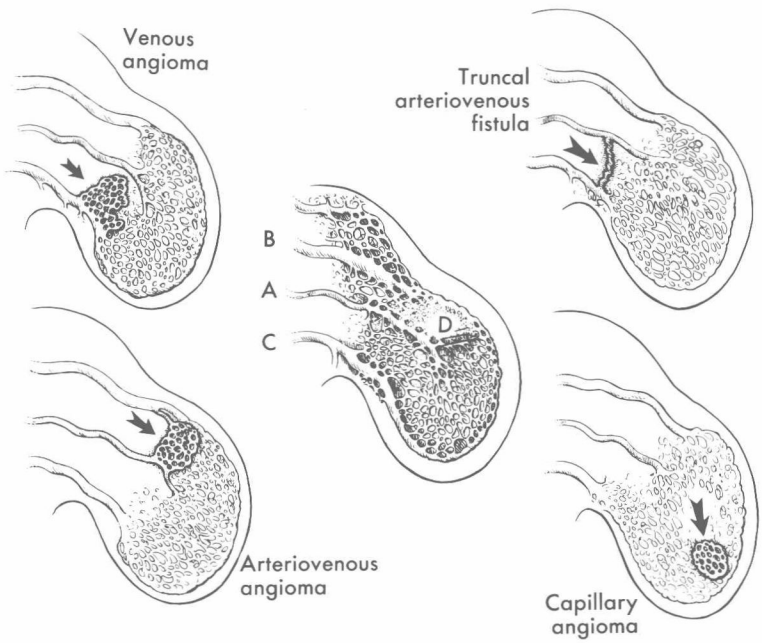

Fig. 1-2. Embryologic development. *A,* Retiform central artery, *B,* Cephalic vein. *C,* Basilic vein. *D,* Primitive capillary plexus undergoing resorption. (Modified from Mulliken, J.B., Murray, J.E., Casteneda, A.R., and Kaben, L.B.: Surg. Gynecol. Obstet. **146:**168, 1978. By permission of Surgery, Gynecology & Obstetrics.)

CLASSIFICATION OF HEMANGIOMAS

A commonly employed classification of hemangiomas is as follows:

A. Benign hemangiomas
   1. Typical
      a. Capillary hemangioma
      b. Cavernous hemangioma
      c. Mixed-combined hemangioma
      d. Port-wine stain—nevus flammeus
      e. Angioma racemosum
      f. Angiokeratoma (Mibelli)
   2. Atypical
      a. Sclerosing hemangioma
      b. Pyogenic granuloma
      c. Spider telangiectasia (nevus araneus)
      d. Glomus tumor
      e. Hemangiopericytoma
      f. Juvenile nasopharyngeal angiofibroma
      g. Venous lakes

B. Syndromes—diseases
   1. Rendu-Osler-Weber syndrome
   2. Sturge-Weber-Dimitri syndrome
   3. von Hippel-Lindau disease
   4. Maffucci syndrome
   5. Blue Rubber Bleb syndrome
   6. Kasabach-Merritt syndrome
   7. Klippel-Trenaunay syndrome

C. Malignant hemangiomas
   1. Angiosarcoma
   2. Kaposi sarcoma
   3. Dermatofibrosarcoma protuberans

Benign typical hemangiomas

The commonest in this group is, of course, the *capillary hemangioma,* also referred to as a *strawberry mark*[1] (Fig. 1-3). It consists of one or more bright red, soft lobulated tumors. The lesion usually appears within the first month of life, increases in size for several months, and then, in contrast to some other forms of hemangiomas, regresses spontaneously, involuting completely by age 7. Bean's[2] remarks are worth repeating, "Lister's classic observations[5] on the natural history of strawberry nevi are a landmark of observation in what was a sea of confusion, vaguely mapped out by shoals of ignorance and hearsay." It was Lister who was first impressed by the fact that the strawberry nevi so common in infants were rare among children or adults. He did not believe that they all had been treated successfully in infancy.[1,2,4] Histologic examination (Fig. 1-4) of a typical capillary hemangioma reveals a proliferation of endothelial cells in which the capillary lumina are either nondeveloped or obscured. The endothelial proliferation may extend from the dermis into the subcutaneous tissue, where, with involution, fibrosis replaces the capillaries, leading to shrinkage and disappearance of the lesion. The typical *cavernous hemangioma* is primarily subcutaneous in location, frequently deeper, and is often ill-defined as to depth (Fig. 1-5). Cavernous hemangioma may be combined with an overlying capillary hemangioma, with the entity being referred to as a combined, mixed, or capillary-cavernous hemangioma (Fig. 1-6). Histologically (Fig. 1-7), these tumors are composed of large, irregular endothelial-lined spaces filled with blood, spaced between fibrous tissue of varying thickness. The cavernous hemangioma, unlike the pure capillary type, frequently may not involute spontaneously and, when present in other systems and organs, may combine with associated signs and symptoms to form a series of syndromes and diseases, which will be discussed later in the chapter.

The *port-wine stain* (Fig. 1-8), otherwise known as nevus flammeus, is characterized as primarily macular, varying in color from light pink to dull red to deep purple and involving one or more distributions of the trigeminal nerve. It is usually unilateral, occasionally bilateral, and may even involve the extremities. Unlike the capillary-cavernous hemangioma, it does not regress but later in life may enlarge, becoming papular or even nodular. Histologically (Fig. 1-9), the tumor is confined primarily to the skin and consists of mature capillaries and papillary ectasias lined with mature endothelium. It does involve the dermis and with age extends deeper into the subcutaneous tissue.

The *AV malformation,* or racemose aneurysm, has a variety of names. The lesion is frequently a large hemangioma that transcends all tissue planes from the skin into the subcutaneous tissue down into the muscle; it may lead to significant enlargement of the involved part of the body (Fig. 1-10). Histologically, the AV malformation is similar to cavernous hemangiomas with multiple arteries and veins involved, the latter even becoming arterialized in terms of vessel thickness and character. Large tortuous vessels are formed, and all the prime elements participate in the growth of the vessels, with the hyperplasia of the muscle formed within the vessel wall being particularly prominent.

Angiokeratomas, one of which is the angiokeratoma of Mibelli (Fig. 1-11), show, in addition to elements of the capillary cavernous hemangioma, multiple dark red papules with a verrucous surface on the skin. They are located commonly on the extremities and appear in childhood. Histo-

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