The Pathology of Vascular ("Arteriovenous") Malformations

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Introduction

VASCULAR malformations of the central nervous system have been known for at least three hundred years (Olivecrona and Ladenheim '57), but are still surrounded by considerable confusion and associated with a complex classification. Detailed morphologic studies of large numbers of cases are rare. Reviews of the many classifications can be found in the works of Evans and Courville '39, Noran '45, Olivecrona and Ladenheim '57, and Raynor and Kingman '65.

While the relationship of vascular malformations to vascular neoplasms is still less than completely clear (Raynor and Kingman '63), it seems relatively certain that arteriovenous malformations are true developmental malformations and not neoplasms (Wolf and Brock '35, Noran '45, Manuelidis '50, Olivecrona and Ladenheim '57, Zülch '57, Russell and Rubinstein '59, Bailey '61, Kaplan et al. '61, McCormick and Nofzinger '66). Zülch '57 states that the distinction between vascular malformations and vascular neoplasms is difficult, and distinguishes between the two groups on the basis of the autonomous growth seen in the latter group. However, as Russell and Rubinstein '59 and others have noted, "in common with the true neoplasms, it is clear that some [malformations] at least are not static but grow and inflict progressive destruction on the adjacent brain." Noran '45 considered that the presence of brain parenchyma between the vessels of the angioma was of "paramount importance" in concluding that it is a malformation rather than a vascular neoplasm. Clearly, however, this does not hold for all malformations, most notably the cavernous angiomas. In spite of these difficulties, one considers the malformations to be distinct from the true neoplasms, and to be caused by faulty embryologic development.

Our purpose is to present a reasonable classification of these lesions and to illustrate the various anatomic types. While the generic term "arteriovenous malformation" is commonly used, by no means all vascular malformations are composed of a mixture of arteries and veins, and thus strict delineation of the terms is necessary. The basic features of this classification are quite similar to those given by Evans and Courville '39, Noran '45, Olivecrona and Ladenheim '57, and Russell and Rubinstein '59.

Classification

Vascular malformations (congenital malformations of intracranial blood vessels, excluding berry aneurysms) consist of:

1. Telangiectasias (Including some cases of Sturge-Weber syndrome)
2. Varix (Including some vein of Galen malformations)
3. Cavernous malformation ("Angioma")
4. Arteriovenous malformation ("Angioma")
5. Venous malformation ("Angioma") (Including some cases of Sturge-Weber syndrome)

All of the 70 vascular malformations personally encountered can be classified into one of the five types (McCormick and Nofzinger '66). Moreover, at least the great majority of the well-documented malformations illustrated in the literature would seem to fall easily into one of these groups. The proper classification of the "hemangioma" of the nervous system is not completely clear. Kernohan and Sayre '52 considered them as true neoplasms. Cases (Burke et al. '64) in which multiple cutaneous and C.N.S. "hemangiomas" were found defy my attempts at precise classification, since no satisfactory photomicrographs of the C.N.S. lesion were shown. Although the term "angioma" is used, it should be clearly understood that it does not mean, as here used,

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a true neoplasm. A brief description of each of these five types of malformations is given.

**Telangiectasias (Capillary Angiomas)**

These are relatively common, typically small, solitary malformations, usually encountered incidentally at necropsy. In my experience, they have been found most commonly in the pons (Fig. 1). Grossly, they present as small areas of red softening, usually with ill-defined borders. Less often, they may appear as a group of dilated vessels resembling a cluster of petechiae (Fig. 2). These malformations seem to be uncommonly associated with massive hemorrhages.

Microscopically, telangiectasias are composed of thin-walled capillaries which are devoid of smooth muscle or elastic fibers. The capillaries may vary greatly in size, and may resemble cavernous spaces in some areas (Fig. 3). More or less normal brain parenchyma is present between the dilated capillaries, although at times the parenchyma becomes gliotic or even heavily mineralized (Fig. 4). In the Sturge-Weber syndrome, such mineralization and gliosis are almost constant (Greenwald and Koota '36, Wohlwill and Yakovlev '57, Roizin et al. '59). Due to common saccular dilatations of the capillaries in telangiectasias ("capillary microaneurysms"), beautifully demonstrated by Russell and Rubinstein '59 and by Courville '63, distinction between some of these malformations and cavernous angiomas may be somewhat difficult. Russell and Rubinstein '59 have stressed this point,