Editorial

Thromboangiitis Obliterans: Fact or Fancy

In a recent publication the entire experience with Buerger's disease at the Beth Israel Hospital in Boston was analyzed and the various considerations that have perpetuated it as an entity were reviewed. It was concluded from this study that the disease originally described by Buerger is indistinguishable from atherosclerosis, systemic embolization, or peripheral arterial or venous thrombosis, singly or in combination. Because of the difficulties inherent in establishing a negative hypothesis, any attempt to prove that thromboangiitis obliterans does not exist can approach such a goal only asymptotically.

To this observer, however, an impartial examination of the entire evidence provides overwhelming support for the opinion that Buerger's disease has never been and is not now an entity in either the clinical or pathologic sense.

The more carefully one examines the various factors associated with the diagnosis of Buerger's disease, the less convincing they appear. In view of current knowledge of the relative frequency of atherosclerosis among young men, it is no longer reasonable to consider age and sex as distinguishing features of Buerger's disease. To appreciate how this association may initially have come about, however, it is necessary to recall that Buerger's original investigations were completed in an era when neither the frequency of clinically significant atherosclerosis nor the ubiquity of thromboembolism was fully recognized. Herrick's classic treatise on the correlation of the pathology of coronary occlusion with the clinical picture of myocardial infarction, for example, was first published 4 years after Buerger described the disease that bears his name. Buerger's initial studies also preceded by several decades the extensive observations on the high incidence of coronary atherosclerosis in young men. Although the use of tobacco and the presence of clinical phlebitis have been considered important findings in the diagnosis of Buerger's disease, no reports were uncovered in which these relationships were established through the use of control studies. It is now also clear that the secondary and tertiary lesions of thromboangiitis obliterans are nonspecific and may result from arterial obstruction from any cause, and that the extent of perivascular fibrosis is no greater in thromboangiitis obliterans than in other thrombotic disorders.

Since the etiology is unknown, the clinical features of the disease are not diagnostic, and the intermediate and healed morphologic lesions are nonspecific, validation of the disease as an entity must rest, finally, with the acute lesion, which Buerger considered to be "absolutely diagnostic." The acute vascular lesion has occasionally been described in superficial veins, but only rarely in deep veins or in arteries. The paucity of such acute lesions in

From the Medical Research Department of the Yamin Research Laboratory, Beth Israel Hospital, and the Department of Medicine, Harvard Medical School, Boston, Massachusetts.

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the deep vessels has usually been attributed to the fact that the affected limbs are amputated only in the late stages of the disease. However, if the natural history of Buerger's disease is indeed that of an episodic, recurrent, and relapsing process, it is remarkable that the traces of the acute lesion are reported so rarely in these specimens. This is particularly significant, since recent arterial thrombi devoid of acute inflammatory reaction have often been noted in such amputated specimens. Although the literature contains a few photomicrographs that resemble in part the picture described by Buerger, it is often difficult to determine whether the vessel is an artery or a vein, whether the wall is intact, whether the surrounding area is free of infection, or even whether or not a septic endocarditis may have been present. Of particular interest in this regard were Buerger's own photomicrographs relating to the acute arterial lesions as documented in five of his major reports. There are four separate examples in which the identical photomicrographs, allowing for minor differences in photographic contrast, spatial orientation, and magnification, are labeled a vein in early papers and an artery in subsequent ones. This identity takes on greater significance since these lesions, together with an additional lesion shown only once, seem, in fact, to be veins rather than arteries.1 The reader is forced to the conclusion that, although Buerger may indeed have seen an acute arterial lesion, he did not at any time present documentary proof of its existence.

Fortunately, the issue of the identity of thromboangiitis obliterans does not, in the final analysis, turn on the inconclusive evidence of whether or not an acute arterial lesion has been seen. It rests, rather, on the specificity of this acute vascular lesion, a question that, for want of adequate arterial lesions, can best be approached in the vein, where the morphologic findings are well documented. From the pathologic point of view, specificity is based on such features as inflammation without necrosis and the presence of giant cells within the thrombus. As shown in control venous biopsies,1 however, the degree of reactive inflammation may vary greatly and may occasionally be as marked as that observed by Buerger. The specificity of the lesion cannot rest on the presence of giant cells alone, since these vary from section to section, may be entirely absent from some areas and can, moreover, be found occasionally in recently organizing thrombi.

There are now sufficient data concerning the morphologic response of arteries and veins in many pathologic processes to indicate clearly that the arterial response does not necessarily mimic the venous reaction. Even granting the existence of a comparable lesion in arteries, however, one must be able to justify the calling of such a lesion specific in the absence of a definitive clinical picture or pathognomonic, morphologic, or etiologic components. It is only necessary to recall the relation of the sarcoid lesion to sarcoidosis or the Aschoff body to rheumatic fever to indicate how difficult it is to find an "absolutely diagnostic" lesion on pathologic grounds alone. If, in addition, one recognizes that the lesion may be mimicked in whole or in part by other less specific processes and that the evidence for its actual existence is tenuous at best, there seems little justification for permitting the whole case for thromboangiitis obliterans to rest entirely on this lesion.

Skepticism concerning the identity of thromboangiitis obliterans has been expressed by numerous investigators,2-5 and other observers have indicated their failure to find examples of the disease among varied hospital populations. Yet, there remains today a widely held belief that thromboangiitis obliterans is a distinct, if uncommon, disease. Retention of thromboangiitis as an entity has been urged by some investigators because, in their opinion, the diagnosis of Buerger's disease describes a characteristic and recognizable clinical picture. Such findings, however, do not define a disease. Others persist in the belief that thromboangiitis obliterans exists because the symptoms and signs respond to specific therapy. Yet, there is no evidence from controlled clinical trials that any ther-
apy, including abstinence from tobacco, is more beneficial to patients said to have Buerger's disease than to patients of similar age with established forms of peripheral arterial insufficiency of comparable severity. Finally, some observers insist that the existence of an acute pathognomonic vascular lesion justifies acceptance of thromboangiitis obliterans as an entity. These same observers explain the paucity of specific arterial lesions on the fact that the disease is no longer as common as it once was. As indicated above, the specificity of the acute vascular lesion has never been established, and the data provided by Buerger's own photomicrographs lend no credence to the belief that the disease was seen more commonly before than after World War II.

This editorial will perhaps serve its most useful function, if these comments stimulate those physicians who still believe in the entity of Buerger's disease to review critically their own experiences with thromboangiitis obliterans.

STANFORD WESSLER

References


Whether my observations and opinions be disproved or supported, I shall be equally satisfied. Truth is the prize aimed for; and, in the contest, there is at least this consolation, that all the competitors may share equally the good attained.—DOMINIC JOHN CORRIGAN, M.D. The Lancet 1:586, 1829.
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