2) Pathogenesis of Truncarteritis Productiva Obliterans; So-called Pulseless Disease or Aortic Arch Syndrome[†]

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The nosological designation of "truncarteritis productiva obliterans" proposed by the present authors is applied mainly to such vascular diseases as non-syphilitic, young female arteritis of the aortic arch syndrome in the classification of Ross and Mc KUSICK. Therefore, such disease mostly presents a typical pulseless syndrome though some lack it. Twenty six autopsy cases were collected from all over Japan which presented diffuse lesions in the aorta or cardinal arteries branching from the aorta. They demonstrated arterial lesions histologically identical with those of pulseless disease and stenosis or obstruction (not dilatation!) of the arterial lumina. The pathogenesis of the present disease is discussed on the basis of this histological examination.

The main histological changes in this disease may be classified as follows;—

Type A; This lesion consists mainly of tuberculoid granuloma with epithelioid cells and LANGHANS giant cells, sometimes associated with coagulation necrosis. These lesions may be present not only in the wall of the cardinal arteries but in the lung, lymph nodes, liver and kidney. In some of the Type A patients there were found active tuberculosis of the lung, intestine, adrenals or bladder.

Type A': In this type small abscesses apparently due to acute exacerbation by steroid therapy etc. are found in the center of the tuberculoid granulomas occurred as in Type A. These lesions may not be confined in the arterial wall alone, but are present also in other organs as in Type A.

Type B: This type lesion represents the foci of coagulation necrosis with reactive proliferation of neighbouring connective tissue cells. Foreign body giant cells are frequently present. Histologically, these lesions resemble subcutaneous rheumatoid nodules in rheumatoid arthritis.

Type C: This type shows irregular, diffuse productive inflammation with lymphocyte or plasma cell infiltration. Sometimes these are associated by newly formed blood vessels and fibrosis. This may be a chronic form of granulomatous changes.

Type D: This type represents cicatricial lesions which were most commonly seen in the whole series. Inflammatory cell reaction is scanty and fibrosis is prominent.

Type D': This type lesion is a pronounced fibrosis that forms fibrous nodules, and thus the media demonstrates distortion of architecture.

Fig. 1 shows the relation of the above mentioned lesions viewing from their morphogenesis. The A, A' and B types may be not of independent etiology and may be placed not on

Fig. 1. Relation between each type of the histological changes viewing from the morphogenesis.

[†] (Received for Publication, August 3, 1965)

the same level. They are therefore arranged in a declined fashion as shown in the figure. This suggests also the possibility that B type lesions may represent in the late stage of Type A lesions.

ETIOLOGIC CONSIDERATION ON THE BASIS OF
MORPHOGENESIS OF THE LESIONS
(cit. Fig. 2)

Because of the not uncommonly associated active tuberculosis in Type A, the present authors once had a strong suspicion that the tuberculoid granulomas in this type were nothing but tuberculosis. Even at present, there are some cases that appears to justify this assumption. On the other hand, recent study has revealed that tuberculoid granulomas can occur without tuberculous lesion in other organs. Histologic demonstration of tubercle bacilli, treponema, fungi, bacteria has been fruitless.

These findings oppose a rush conclusion that the tuberculoid lesions all result from tuberculosis. Nevertheless, provided that the lesion is not of tuberculous origin, it seems at present most appropriatively to interpret that they would be caused by some infection or allergic mechanism following infection. In the pathoge-

nesis of this disease, therefore, one can not deny that infection plays an important role as a trigger factor, although the infection theory may not give sufficient explanation for the preference of the disease for young females. Taking into consideration that the Type B lesion with coagulation necrosis and proliferative reaction of the connective tissue resembles the rheumatoid nodule and sometimes gives rise to nodular fibrosis as in Type D' lesion, it can not be denied that in the pathogenesis of this disease rheumatoid or rheumatic processes may play a role. Nevertheless, the present study has failed to find ASCHOFF body or endocarditis as evidence of rheumatic carditis. In only 4 of 26 cases clinical history or laboratory examination suggests the existence of rheumatoid arthritis.

Therefore, the interpretation of the pathogenesis of this disease on the basis of the classical concept of rheumatic disorder is not fully supported by the morphological evidence in this study.

The Type D lesion of the cicatricial state may be variously interpreted. Whether it may be nothing but the terminal picture of A, B and C lesions, or, in addition, even congenital metabolic disorders, e. g. MARFAN's syndrome, may show such lesions as the present type D in their late stage, remains undetermined. Furthermore, we have found one case with lupoid hepatitis that showed acquired lesions in the arterial wall simulating MARFAN's syndrome. As the typical MARFAN's syndrome tends to demonstrate dilatation of the vascular lumina, there is some doubt whether MARFAN's syndrome of congenital or collagen disease origin anyhow can develop the lesions identical with the scarred lesions in this disease.

SUMMARY

Twenty six autopsy cases of "truncarteritis productiva obliterans" were collected in Japan according to certain criteria. The morphogenesis of the lesions was studied on the ground of histology of the arterial wall lesion. It has been suggested that infection or allergy following infection of the arterial wall may be the most probable etiologic factor, or may act at least as a trigger in the early morphogenesis.

Fig. 2. Etiologic relation between each type of lesions.

Fig. 3. Type A lesion showing a tuberculoid granuloma in the media of a common carotid artery consisting of typical Langhans giant cells in the center and epithelioid cells mingled with lymphocytes in the periphery.

Fig. 4. Type A' lesion showing a small abscess in the center of tuberculoid granuloma in the adventitia of a pulmonary artery.

Fig. 5. Type B lesion showing a foci of coagulation necrosis with reactive proliferation of the neighbouring connective tissue cells in an aortic media.

Fig. 6. Type D' lesion showing fibrous nodules which cause the distortion of architecture of an aortic media.

It has been suggested, too, that immunopathological mechanism of a rheumatoid nature also may play a role in the pathogenesis.

This work was aided by a grant from the Ministry of Education and from Waksman Foundation of Japan Inc.

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