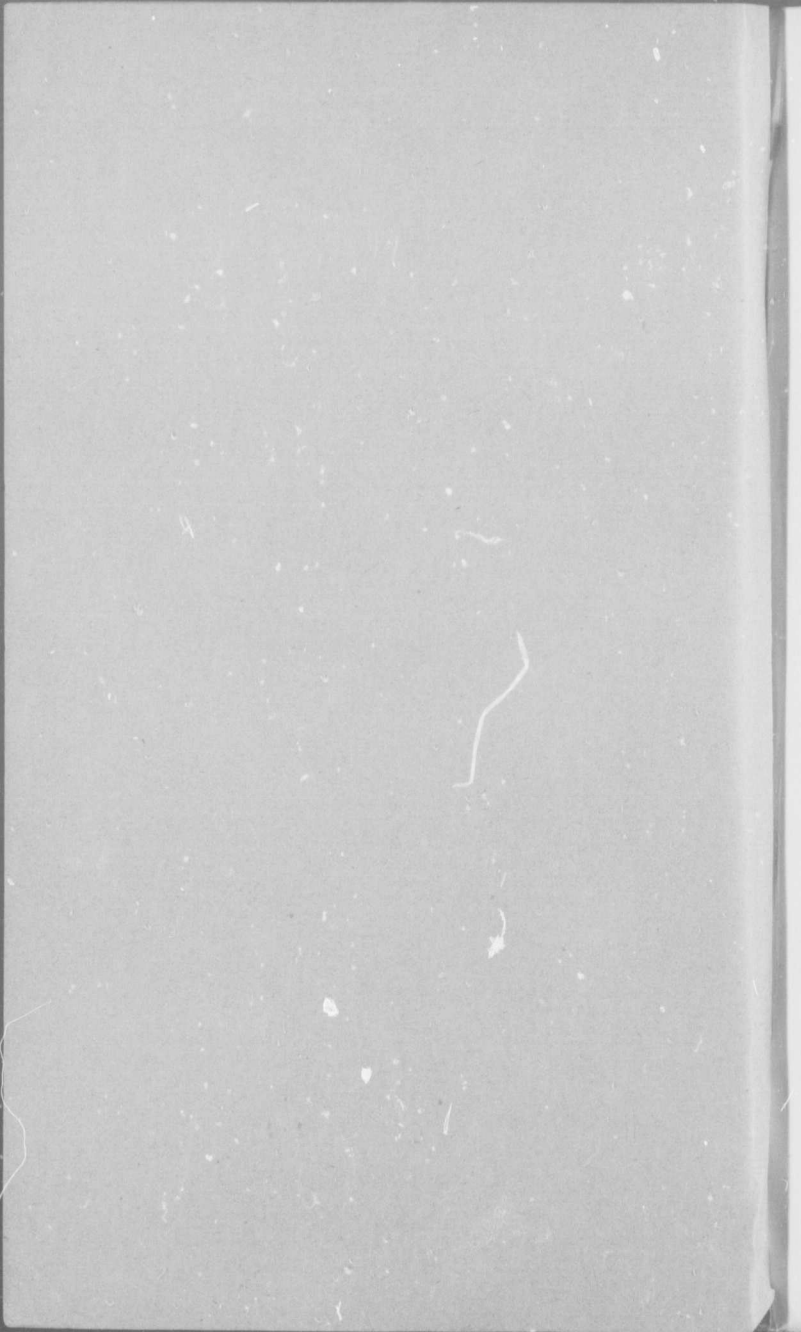


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A CASE OF CONGENITAL SYPHILITIC  
AORTITIS.

By OSKAR KLOTZ.



## A CASE OF CONGENITAL SYPHILITIC AORTITIS.

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(PLATE IV. FIG. 1.)

THOUGH in recent years much stress has been laid on the occurrence of acquired specific lesions in the larger arteries, there has been remarkably little attention paid to the arterial lesions in congenital syphilis. Of the latter condition there have been several reports of the changes found in the omphalic vessels, by Winkel, Birch-Hirschfeld, and Chiari, while there are also descriptions, in the literature, of changes occurring under similar conditions in the veins.

Now that we know that the syphilitic virus passes from the mother to the fœtus, and that all the fœtal tissues become infected as they do in the acquired disease, it is to be expected that the lesions in the two instances will bear some similarity, save that the less resistant tissues of the fœtus will be damaged the more severely.

Our knowledge of acquired syphilitic aortitis has been especially added to by the reports of Heller and his pupils. They pointed out that the syphilitic affections of the aorta have certain characteristics which are constant. The syphilitic lesions, they reported, are most frequently found in the ascending aorta; and these lesions, when viewed from the intimal surface, show a peculiar scarring, with deep furrows and an extensive endothelial proliferation. Fatty degeneration of the vessel wall was seldom seen in these areas, and calcification almost never.

In 1885 Doehle first described the microscopical changes taking place in syphilitic arteritis, and in the main his descriptions have been accepted by later workers. A constant inflammatory infiltration is found present about the vasa vasorum, even in the earliest lesions. This inflammatory zone lies in the adventitia and the outer third of the media, and in itself is quite characteristic in syphilis. It may be that, when Koester was describing the mesarteritis as the usual affection of beginning arterio-sclerosis, he had before him specimens of syphilitic arteritis, and that it was from such that he drew the general conclusions concerning arterio-sclerosis. It is agreed by all authors that in syphilitic arteritis the intimal proliferation is a process secondary to the medial change.

Mraček noted in congenital syphilis that there were numerous ecchymoses in the adventitia of the larger vessels, while the vasa vasorum exhibited a small-celled infiltration about them. Occasionally, too, he noted a slight endarteritis in the carotid, crural, and iliac arteries. Buchta reported a case of congenital syphilis in a young adult of 17, in whom the vessels of the arms

and legs became cord-like, and greatly impeded the circulation, so that partial gangrene of the foot set in.

Bruhns has recently examined nine cases of congenital syphilis, and has been able to demonstrate lesions in the aorta which are very like the lesions met with in mesoarteritis productiva, as described by Chiari in adults. The dilated vasa vasorum in the adventitia showed a constant small-celled infiltration about them, while the condition was also to be traced in the outer zone of the media. The dense inflammatory infiltration of the media led to a pushing apart of the elastic fibres, so that the structure of the vessel became looser in this region. The leucocytic collections in his specimens consisted both of mononuclear and polynuclear cells, while epithelioid cells were also present.

Bruhns concluded that, in congenital syphilis of the aorta, acute inflammatory infiltrations are present in the outer layer of the media and in the adventitia, particularly about the vasa vasorum. He held that the lesions of congenital syphilis in the aorta are identical with those of the acquired type as described by Chiari.

The main point which Chiari brings out in his studies is the differentiation between the two forms of endarteritis chronica deformans. His Type A, which is the ordinary endarteritis deformans as described by Virchow, and which is the disease of the aorta ordinarily considered as arterio-sclerosis, is a primary disease of the intima, in which degenerative changes later take place. The syphilitic aortitis belongs to his Type B, in which the intimal changes are secondary to inflammatory and other changes in the media. This latter type he speaks of as mesoarteritis productiva.

Wiesner has studied the arterial changes in ten undoubted cases of congenital syphilis in children, and found constant characteristic lesions in the arteries. The aorta with its larger branches, and the pulmonary artery, were the most frequent sites of the pathological conditions which he found. In these vessels he distinguished a boundary zone between the media and adventitia, in which, as is also the case in the arterial lesions of acquired syphilis, the primary alterations in the tissue are to be looked for. He found a constant hyperæmia of the vasa vasorum both in the adventitia and in the boundary zone, while in some cases a thrombosis, and even a hæmorrhage, occurred in these regions. The hæmorrhages in his cases occurred most frequently in the adventitia. Another constant feature found in the arteries in acquired syphilis is the presence of a round-celled infiltration following the vasa vasorum from the adventitia into the media. In congenital syphilitic children several weeks old, Wiesner found a perivascular fibrosis replacing the cellular infiltration, and in some cases an obliteration of the nutrient vessels.

In one instance of a child 3 months old he noted the occurrence of a connective-tissue production in the media, so that the elastic fibres had almost entirely disappeared in this region. Wiesner considered that this was a later stage of the inflammatory infiltration seen about the vasa of the aorta in newborn children.

The following case, from the Montreal Maternity Hospital, came to post-mortem:—

CASE.—R., a still-born male child, was born of a healthy mother. An indefinite history of syphilis was obtained from the father. The anatomical findings in the child were—atelectasis of the lungs, petechial hæmorrhages of the thymus and pleura, enlarged spleen, hæmatoma of the scalp, rudiment of spina bifida over the coccyx, and syphilitic aortitis.

The aorta showed at the arch an area of change in its wall with some blood infiltration of its deeper layers, which could be seen through the intima as a bluish-purple patch. This arterial change extended from the arch towards the ascending aorta, and the affected area was somewhat raised above the general

level of the intima. When the aorta was viewed from the intimal surface it was noted that irregularly radiating grooves furrowed the surface over the arch. These puckerings of the intima resembled the aortic lesions of acquired syphilis. Macroscopically no break was found in the intima to account for the infiltration of blood in the vessel wall.

On section, the vessel wall was found to be very much altered, so that some difficulty was experienced in locating the different layers. The intima over the discoloured area was thinned, a little fibrous tissue with the cells lying parallel to the endothelial cells being all that represented the otherwise thickened intima. The surface of the intima was not smooth, but seemed to be marked by sulci. The whole thickness of the intima was permeated by fine elastic threads, which formed a kind of network through it. The internal elastic membrane was present as a broken-up lamina, which, besides being partially destroyed, was also split up longitudinally into several lamellæ. The connective tissue of the intima was fairly cellular, but there was no infiltration of leucocytes. At one portion of the intima, where it lay over the hæmorrhagic area in the media, there was a loss of tissue, though true necrosis was not present in it. The deeper portion of the intima stopped abruptly on either side of the medial blood cyst, but a thin layer of intima separated the blood cyst from the lumen of the vessel.

The media showed the most change. The outer zone, or, as Wiesner terms it, the boundary zone of the media, shows a small-celled infiltration which is particularly localised about the vasa vasorum. In a few places the small-celled infiltration is found to advance into the middle zone. Everywhere it is noted that the vasa are unusually patent, and in one location they are found to pass unusually far into the media, and to lie here as large open-mouthed capillaries.

At the site of the blood infiltration the medial tissue is almost entirely lost. The blood cyst, if we can call it such, extends from beneath the intima to the outer third of the media, and in the region where the blood is found there are no remains of the tissue of the media. Clotted blood with a meshwork of fibrin and entangled leucocytes occupy this site. Towards the lateral margins of the hæmorrhage the media is seen in the process of destruction, in which the muscle and elastic fibres are both involved. A stringy tissue without nuclei is all that remains of the tissue of the media. The elastic fibres exist only in isolated patches, and are here represented by bunches of short granular strands. This process of necrosis also advances close to the adventitia.

The media in some parts showed small dense aggregations of fibrous tissue, which was advancing from the adventitia, and was replacing the normal structure of the coat. Nowhere, however, were giant cells to be found.

The adventitia showed a small-celled infiltration about the vasa. These leucocytes were mostly of the character of lymphocytes with few polymorphonuclear cells among them.

The interesting points in the microscopical examination of the artery are that it had all the usual characters of aortitis syphilitica, as they are met with in the acquired disease. The slow but not fatty degeneration of the media, with the secondary fibrosis in it and in the intima, the proliferation of the vasa vasorum with their surrounding inflammatory infiltration, and the low grade of inflammation present in the adventitia, all point to the syphilitic nature of the disease. This is further supported by the site of the lesion in the aorta, and the lymphoid infiltration of the vessel coats. As to the nature of the hæmorrhage which burrowed through the middle and inner zone of the media, and then broke through the greater part of the intima, it is hard to say. However, the hæmorrhage is not like a dissecting aneurysm,

as it has no connection with the blood in the lumen of the aorta. It is more likely that the aorta, having been degenerated at this point by the specific disease, a rupture occurred of one of the much dilated vasa. The nature of the hæmorrhage working its way from the deeper tissues towards the intima, and advancing in the vessel wall in the wake of the degenerative processes, would bear this out. Thromboses, which were found in several of the vasa, also illustrate the severe changes which had affected these capillaries.

That great degenerative changes were going on in the aortic wall is seen in the destruction of the elastic and muscle fibres of the media, and that the process was a chronic one is demonstrated by the thickened intima. These features are all in accord with a syphilitic affection of the artery.

Beyond the affected area in the aortic arch the vessel showed little change. In the pulmonary artery there was some inflammatory infiltration of the adventitia and media, but the abdominal aorta was without this change.

*To summarise*, we have in this case an indefinite history of congenital syphilis, with, however, lesions in the aorta which are identical with those of acquired syphilis, though these are more extensive than is usually seen.

As Wiesner noted in one of his cases, the inflammatory reaction which is found about the small vasa is eventually converted into connective tissue, so that fibroses of the vessel wall occur. These fibroses produce the puckerings and stellate scarring in the aortic arch, which in the adult have always been considered a sign of acquired syphilis even when such a history has been absolutely denied. May it not be that in some of these cases the scarrings of the aorta are the healed results of a congenital syphilis of the aorta? It must be that some of the cases of congenital syphilis with early lesions in the aorta recover, and that the remains of the arterial disease are to be recognised only in the fibroses and chronic inflammation of the tissues.

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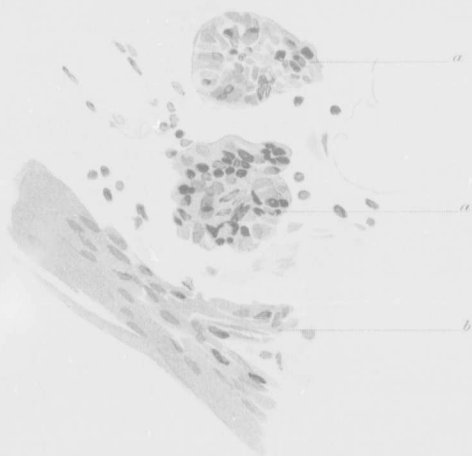


Fig 9

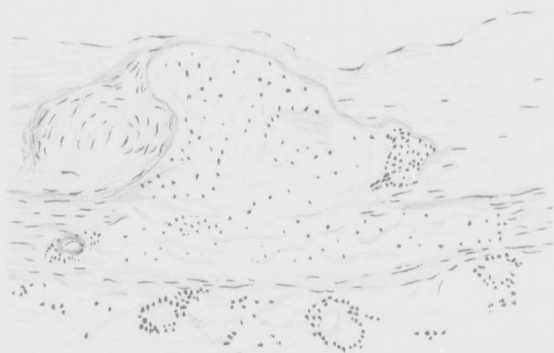


Fig A.