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Two autopsy cases of a peculiar disease, so called "Pulseless Disease"

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Abstract

Two autopsy cases of "Pulseless Disease", a peculiar disease of the aorta and its branches, are reported in this paper. In "Pulseless Disease" autopsy cases of old persons are relatively rare. The two cases are both of old persons, and show not only stenosis or obstruction as indicated by the name, "Pulseless Disease", but also marked dilatation of arterial lumen. There are no histological differences between stenosed and dilated portions. One of the two cases demonstrates so-called "Atypical coarctation of the aorta", and the other is combined with severe atherosclerosis with marked calcification.

Introduction

The disease known by the name, "Pulseless Disease", was first reported by a Japanese ophthalmologist, Takayasu. In 1908 he made a report on "a case of peculiar changes of the central retinal vessels", in which he pointed out a wreath-like arteriovenous anastomosis surrounding the optic disc, but no reference was made by him of pulselessness. At the same time Onishi and Kagoshima supplemented each a similar case with absent radial pulses. In 1948 Shimizu and Sano described the symptomatology of this disease, and first used the name, "Pulseless Disease". They mentioned clinically three cardiac symptoms of this disease: (1) absent pulse of upper limbs, (2) arteriovenous anastomosis surrounding the optic disc, and (3) an acceleration of carotid sinus reflex. The "Pulseless Disease" was reported under various names; Takayasu's disease, Takayasu's arteritis, aortic arch syndrome and aortitis syndrome etc, and pathologically named as truncoarteritis productiva obliterans by a Japanese pathologist, Nasu. This disease was found predominantly in Asia, however infrequent in other continents, and seems to have a predilection for young female. Some
cases of so called "Atypical coarctation of the aorta", noticed by Inada et al, was considered to belong in the same category of this disease, and recently frequent association of hypertension with this disease has been noticed. The etiology of this disease is still unknown.

Clinically this disease was diagnosed mainly as follows; pulseless disease, valvular disease, coarctation of the aorta, aortic arch syndrome and renovascular disturbance. Causes of death were chiefly heart failure, death under operation, sudden death and uremia.

Pathologically the main lesions of "Pulseless Disease" are mainly localized to the arteries of elastic type including pulmonary artery. The chief changes in the initial stage are supposed to be limited to adventitia and outer portion of media, based on an inflammatory reaction, which in turn brings about several following changes in arterial wall; marked fibrous thickening of adventitia, destruction of media and intimal thickening.

Case reports

Case 1: The patient is a 54 year-old Japanese female, who has noticed a pulsation of neck at the age of 34 years but did not seek medical attention. At the age of 45 years she saw a doctor with the complaints, sense of pressure at chest and angina-like attacks. At that time hypertension and cardiomegaly were pointed out, so she was admitted to the Nagasaki University Hospital and was diagnosed as aortic insufficiency and aortic arch syndrome. The angiocardiography revealed obstruction of the left common carotid artery, aneurysm of the right subclavian artery, stenosis of the descending aorta and kinking of the right common carotid artery, and thrill and pulsation were noticed in all portions mentioned above. After treatment in the hospital for two months she had been followed up as outpatient for eight years. One day prior to her death anorexia and orthopnea appeared and suddenly she fell on her back and was dead. The entire clinical course is approximately 18 years.

Case 2: The patient is a 66 year-old Japanese male. About two years ago he noticed a dyspnea at the rest and a strangulated sense of chest, and visited a doctor to be diagnosed as hypertension and cardiac asthma. Three months after he was admitted to the Nagasaki University Hospital and was diagnosed as severe atherosclerosis, aortic arch syndrome, coronary insufficiency with ventricular hypertrophy on both sides and gastric ulcer etc. The left radial pulse was difficult to palpate, and the variation of blood pressure between left and right arm was noticed. The angiocardiography revealed aneurysmatic dilatation of the ascending aorta, narrowing of the left common carotid artery and stenosis of the left subclavian artery. After medication for about one month the symptoms were improved, but the sense of pressure at chest, dyspnea and stridor appeared and he died of increased heart failure.

The both cases are negative for Wassermann reaction and positive for tuberculin reaction. The blood pressure of Case 1 is 186/30—0 mmHg, and that of
Case 2 is 170/90 mmHg in the right arm, 110/80 mmHg in the left arm and 230/90 mmHg in the lower extremities. The erythrocyte-sedimentation rate moderately accelerates in both cases, but the other examinations are within normal limits. Case 1 has a history of pulmonary tuberculosis at the age of 43 years, and Case 2 has that of pleurisy at the age of 28 years.

**Autopsy findings**

Case 1: The main lesions correspond to the artery of elastic type including pulmonary artery (Fig. 1). The wall of affected arteries are diffusely thickened and the inner surface reveals relatively smooth appearance with occasional irregular white plaques (Fig. 2). But neither atheroma nor thrombi are noticed. The wall of the left common carotid artery is strongly thickened and markedly contracted in the entire length, so that the lumen is stenosed or obstructed (Fig. 4, Fig. 5). The lumen of the descending aorta is narrowed to the diaphragmatic portion as described as "Atypical coarctation of the aorta" while that of the abdominal aorta is not remarkably changed. On the contrary the aortic arch, brachiocephalic artery, right common carotid artery and right subclavian artery are markedly dilated and partially aneurysmatic. Findings of other organs are as follows; cardiomegaly (600 g) with moderately thickened and contracted aortic valves, occasional pulmonary thromboarteritis with infarction in both lungs, partial atelectasis of the left lower lobe, two calcified nodules in the left lower lobe, cloudy swelling of the liver, moderate nephrosclerosis, cholelithiasis and capillary hemorrhage of the stomach and intestine.

Case 2: The distribution of "Pulseless Disease" is similar to that of Case 1 (Fig. 1). The wall of the affected arteries are diffusely thickened. In this case, there is severe atherosclerosis with marked calcification of the aorta and its large branches, especially of the thoracic aorta, brachiocephalic artery, right subclavian artery and right common carotid artery, the inner surface of them being rough and irregular (Fig. 3). The lumen of the left common carotid artery and that of the left subclavian artery are almost obstructed (Fig. 4, Fig. 6). The thoracic aorta, especially of the descending aorta, brachiocephalic artery, left subclavian artery and proximal portion of the right common carotid artery are markedly dilated. Findings of other organs are as follows; cardiomegaly (680 g) with fibrinofibrotic pericarditis, subacute splenitis, severe fatty liver, pyelocystitis and moderate pulmonary edema.

**Histological findings of arteries**

In the adventitia, fibrosis is more remarkable. Diffuse and marked proliferation of collagenous fibers with partial hyalinization are arranged in various pattern; a nodular, fascicular and strationform ones. The adventitia is strongly
thickened and occupies the almost half or more width of the arterial wall. Several granulomas consisting of lymphocytes also plasma cells, a small number of histiocytes and some giant cells of Langhans type, are found in the adventitia and also in the media at the proximal portions of the pulmonary artery and ascending aorta in Case 1 (Fig. 8, Fig. 9). Beside these changes, there are perivascular cell infiltrations of various degree chiefly composed of lymphocytes in the adventitia and partially in the media of all the affected arteries, which sometimes show lymph-follicle like appearance (Table 1). In Case 2, the perivascular cell infiltrations are almost equal to Case 1, but granuloma formation is not seen. The granuloma seems to be prone fibrosis.

In the media, the changes are found chiefly at the outer one third to half portion, and are summarized as follows; newly proliferated capillaries and arterioles, deposition of mucoid substance, partial fibrosis and interruption and coagulation of elastic fibers. At some parts the media is more thin oppressed by the nodular fibrosis of the adventitia.

In the intima, fibrocellular proliferation is remarkable with newly produced elastic fibers and deposition of mucoid substance.

The wall of vasa vasorum at the affected portions are markedly thickened with fibrocellular proliferation of the intima and deposition of mucoid substance, so that the lumen is stenosed or obstructed.

There are no histological differences between the stenosed and dilated portions (Fig. 5, Fig. 6, Fig. 7).

Table 1. Perivascular Cell Infiltration in Media and Adventitia

<table>
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<tr>
<th>Artery</th>
<th>Media</th>
<th>Adventitia</th>
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<tr>
<td>Aortic arch</td>
<td>+</td>
<td>#</td>
</tr>
<tr>
<td>Brachiocephalic</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>R. common carotid</td>
<td>-</td>
<td>#</td>
</tr>
<tr>
<td>L. common carotid</td>
<td>-</td>
<td>±</td>
</tr>
<tr>
<td>R. subclavian</td>
<td>-</td>
<td>±</td>
</tr>
<tr>
<td>L. subclavian</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Thoracic aorta</td>
<td>-</td>
<td>±</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>#</td>
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Summary

Autopsy cases of "Pulseless Disease" in old persons are relatively rare. The two cases reported in this paper are both of old persons. In "Pulseless Disease", the lumen of the affected artery tends to be stenosed or obstructed, but also can be dilated and even partially aneurysmatic as in these two cases. The stenosis of the lumen is chiefly caused by the contraction of the adventitial fibrosis overcoming the pressure on the arterial wall, promoted with the intimal thickening. Dilatation of that may be caused by increased arterial pressure, which exceeds the contracting effect of wall, and also influenced by the duration of the disease. There are no histological differences between stenosed and dilated portions. One of the two cases demonstrates so-called "Atypical coarctation of the aorta", and the other is combined with severe atherosclerosis with marked calcification.
Two autopsy cases of a peculiar disease, so called "Pulseless Disease"

The Pulseless Disease has been reported during the last 60 years in Asia, chiefly in Japan, but might be found more frequently in tropical and subtropical countries of Asia, as more attention will be paid to this condition.

References


日本の奇妙なる動脈疾患

一脈なし病一

山下兼彦・国田五重

長崎大学熱帯医学研究所病理学部

摘要

いわゆる「脈なし病」の名で呼ばれ、大動脈とその主要分枝に主病変を有する疾患の剖検例を経験したので報告した。「脈なし病」の高令者の剖検例の報告は比較的少ないが、本症例は高令の症例である。

又、本疾患ではその名が示すとおり動脈内腔の狭窄あるいは閉塞を来たすことが特徴であるが、こうした所見に加え、本剖検例では一部に動脈瘤様となった部を含め著明な動脈内腔の拡張が同時に認められた。一方、組織学的には、狭窄部、拡張部の間に差達は認められなかった。二例の中一例は異型大動脈縮窄症の所見があり、他は著明な石灰化を伴った高度の動脈硬化症を伴っている。
Fig. 1. Schemas presenting the arterial lesions.

Case 1.

Case 2.

Affected Portion of Arterial Wall

Fig. 2. Aorta and its large branches (Case 1).

Fig. 3. Aorta and its large branches (Case 2).
Two autopsy cases of a peculiar disease, so called "Pulseless Disease"

Fig. 4. Transverse section of left common carotid arteries (right: Case 1, left: Case 2).

Fig. 5. Typical arterial lesion showing the narrowing of lumen (the left common carotid artery of Case 1).

Fig. 6. Typical arterial lesion showing the obstruction of lumen (the left common carotid artery of Case 2).
Fig. 7. Typical arterial lesion at the portion of dilated lumen (the right common carotid artery of Case 1).

Fig. 8. Granuloma in the media of the pulmonary artery of Case 1.

Fig. 9. Granuloma in the media.