The Aortic Arch Syndrome (Pulseless Disease)

A Report of Ten Cases With Three Autopsies

By W. M. Thurlbeck, M.B., Ch.B., and J. H. Currens, M.D.

Robert Adams described a patient in 1827 whose arterial pulses were not palpable (fig. 1), and this is the first recorded case of "pulseless disease" that we have been able to find. The term in recent time has been used to describe a peculiar syndrome in young women, usually Japanese, characterized by the absence of arterial pulsation in the arms and neck, often associated with cataracts and peculiar vascular abnormalities of the retinae. The optic changes were described by Takayasu and the syndrome bears his name, although the absence of radial artery pulsations was mentioned only in the discussion of his report. Any disease of the aortic arch, however, can produce a similar picture of absent arterial pulsation in the arms and neck. Ross and McKusick used the term "aortic arch syndrome" to describe these diseases and their effects. These authors found syphilitic aortitis to be the common cause of these syndromes in Baltimore and considered atherosclerosis alone to be a rare cause.

Experience at the Massachusetts General Hospital suggests that atherosclerosis with or without superimposed thrombosis is a more common cause of this syndrome than is generally realized. It is the purpose of this paper to describe 10 patients with pulseless disease and to discuss the etiology and the pathology with particular reference to 3 patients on whom autopsies were performed.

Methods

Ten patients have been observed in the hospital between 1952 and 1957. All these cases had in common absent or diminished pulsations and blood pressure readings in one or both arms. In addition to the usual clinical and laboratory investigations, the blood pressures were determined in 4 patients by a recording machine, which picks up Korotkov vibrations over the brachial artery when these are indistinct or inaudible on auscultation. Autopsies were performed in the routine manner on the 3 patients who died in the hospital.

Results

The case histories of the 10 patients and the significant autopsy findings in 3 are presented below. Tables 1 and 2 summarize the clinical features of these cases.

Case 1. A 54-year-old man was admitted on March 9, 1954, because of attacks of fainting. In the 6 years prior to admission he suffered dyspnea and substernal choking on exertion, bilateral claudication, a myocardiac infarction, transient episodes of blindness, dizzy spells, 2 episodes of unconsciousness, episodes of numbness and weakness of the right hand, failing memory, and aphasia. Past and family histories were noncontributory.

On physical examination feeble pulsations of the femoral and the dorsalis pedis arteries were the only pulses that were palpable. Blood pressure in the legs was 160 systolic. The fundi showed normal disks, multiple microaneurysms, many peripapillary fusiform vascular dilatations, and irregular venous dilatations with congestion and sluggish flow. The retinal artery pressure was 17/13 mm. Hg O.D. and 11 mm. Hg O.S. with insignificant pulse pressure. Speech was slow and hesitant, words were occasionally misused, and syllables were reversed.

Laboratory examinations of blood and urine were normal. An electrocardiogram showed Q waves in leads II, III, and aVF with upright T waves. X-ray of the chest showed no calcification or dilatation of the aorta. Intravenous pyelogram was negative. The Hinton test for syphilis was negative.

Under hypothermia thrombendarterectomy of the innominate and left common carotid artery was performed (fig. 2). The innominate and left common carotid arteries were found occluded proximally, with no blood flow through the former and little through the latter. After thrombendarterectomy, flow was good through the left common carotid but the innominate artery thrombosed almost immediately. A bypass arterial shunt was
then placed between the ascending aorta and the right common carotid artery. The patient tolerated the procedure well, but during the warming phase postoperatively ventricular fibrillation occurred. Cardiac massage was instituted after 3 to 4 minutes, and the heart began to beat spontaneously after 3 1/4 hours of manual massage and electric defibrillation.

The patient regained full consciousness with a regular and good pulse, but then developed progressive respiratory difficulty from left ventricular failure and died of acute pulmonary edema 48 hours postoperatively.

At autopsy the ascending aorta and the arch of the aorta showed numerous, lipoid plaques involving 50 per cent of the surface of the arch and ascending portion; the intima was ulcerated over 1 plaque, 1.5 cm. in diameter, proximal to the innominate artery and adjacent to the site of the graft. The aortic valve and coronary ostia were normal and there was no dilatation or calcification of the aorta. The abdominal aorta was severely atherosclerotic with many large, ulcerated plaques covered with a thin plate of red-brown thrombus. In the descending thoracic aorta, iliac, and femoral arteries were numerous slightly raised, lipoid streaks.

The entire innominate artery was occluded by a red-brown, shiny, slightly adherent, granular thrombus. Its walls were focally calcified, and beneath the thrombus were streaks of fatty material. The right common carotid artery showed a single lipoid streak. The right internal carotid artery showed some calcification and a plaque that narrowed the lumen about 20 per cent. The homograft connecting the ascending aorta to the right common carotid artery was patent.

The left common carotid artery at the site of incision was patent. The left internal carotid artery in its proximal 2 cm. was obstructed by a fibrofatty thrombus through the center of which was a lumen 1 to 2 mm. in diameter; the wall of the vessel in this region was focally calcified. The proximal 3 cm. of the left subclavian artery was also occluded by a light brown fibrous thrombus with a central lumen 1 to 2 mm. in diameter; the wall in this region was atherosclerotic and calcified. The vertebral arteries were patent, soft, and about twice normal size. The visceral branches of the aorta had occasional atherosclerotic plaques but no significant obstruction.

Microscopic examination of multiple sections through the aorta and its branches showed severe atherosclerosis, but no syphilis or other arteritis. The occlusions of the left internal carotid and left subclavian arteries showed organized, canalized thrombi in severely atherosclerotic vessels, with many pigment-filled macrophages in the subclavian thrombus and a few in the internal carotid thrombus. The innominate artery was occluded by a recent antemortem thrombus.

The heart weighed 520 Gm. and showed signs of cardiac massage. There was severe diffuse sclerosis of the coronary arteries with almost complete, old occlusion of the anterior descending branch of the left coronary artery. The lumens of several other portions of the coronary arteries were reduced by half. Microscopically, the heart showed focal interstitial and perivascular fibrosis.

The brain weighed 1,350 Gm. The cerebral vessels were free of atherosclerosis. Gross and microscopic examination of the brain revealed multiple small areas of infarction, more on the

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<tr>
<th>Case no.</th>
<th>Age (yr)</th>
<th>Duration of symptoms (min)</th>
<th>Cardiac arrest with cardiac massage (min)</th>
<th>Clipping of coronary arteries</th>
<th>Coronary occlusion</th>
<th>Stroke</th>
<th>Mental deterioration</th>
<th>Transient blindness</th>
<th>Diabetes mellitus</th>
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*Three siblings had diabetes.
AORTIC ARCH SYNDROME

TABLE 2.—Clinical Examination and Laboratory Data

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<th>Case no.</th>
<th>Radial L</th>
<th>Carotid L</th>
<th>Femoral L</th>
<th>Arm</th>
<th>Leg</th>
<th>Machine (arm)</th>
<th>Electrocardiogram</th>
<th>Blood sugar (mg.%), Hinton test</th>
<th>Cholesterol (mg. %)</th>
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<td>+</td>
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<td>80 Syst. (R)</td>
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<td>Dicumarol</td>
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<td>+</td>
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<td>140/70 (R)</td>
<td>Myocardial infarction (post.)</td>
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<td>130 Syst. (R)</td>
<td>LVH</td>
<td>—</td>
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<td>LBBB</td>
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<td>130/70 (R&amp;L)</td>
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<td>140/90 (R)</td>
<td>80/60 (L)</td>
<td>LVH, ST and T wave changes</td>
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<td>Neg.</td>
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LVH, left ventricular hypertrophy.
LBBB, left bundle-branch block.
mained for six weeks, with little alteration in his symptoms, except that his strength was observed declining daily and his breathing becoming more difficult: his rest during the night was still more imperfect: during the entire of this distressing period, no pulse was to be felt in any artery in the body: although I daily made the most careful examination, it was in vain.

Fig. 1. From a case report by Mr. Robert Adams published in the Dublin Hospital Reports in 1827. It concerned a 68-year-old physician, whose pulses disappeared after an attack of chest pain, and who died 6 weeks later.

ment and absent peripheral arterial pulsations, except possibly for a weak right carotid pulsation, were observed.

The patient's father had died at age 50 and his mother at the age of 52, each of a "heart attack."

On physical examination Cheyne-Stokes breathing was striking, the blood pressure was unobtainable in either arm, and no distinct pulsations could be felt anywhere in the body except over the abdominal aorta. The optic disks appeared distinct in outline, and there was no arteriovenous nicking. The arterioles, however, were easily obliterated by slight pressure on the eye. His vision was good in each eye. The neck veins were not distended in the upright position and did not pulsate. The heart was enlarged 1 cm. to the left of the midclavicular line and no murmurs were heard. The liver extended 3 finger-breadths below the costal margin in the epigastrium and was nontender. There was a right midthigh amputation. Responses to complex commands were delayed, but there was no definite motor or sensory abnormality, and the reflexes were normal except for absent abdominal reflexes.

Laboratory studies were not significantly abnormal except for the electrocardiogram, in which Q waves and ST and T changes suggested a previous posterior myocardial infarction. Blood Hinton tests for syphilis were negative on 2 occasions.

Severe Cheyne-Stokes breathing and dyspnea persisted without relief. On the third hospital day a venous circulation time with Decholin was 61 seconds with a fair endpoint. Approximately 10 minutes later, he developed severe respiratory distress with cyanosis, rapid breathing, tachycardia of 130, and circulatory collapse, and he died within 5 minutes. This episode was considered to be a fatal drug reaction to sodium dihydrocholate.13

Autopsy revealed severe extensive atherosclerosis of the aorta with much intimal ulceration involving the whole aorta (fig. 4). The aortic cusps and coronary ostia were normal and the ascending aorta was not dilated. The origins of the innominate, left common carotid, and left subclavian arteries were occluded by an adherent red-brown thrombus (fig. 5). The entire innominate artery showed severe atherosclerosis and calcification with occlusion extending to the origins of the right subclavian and common carotid, whose origins were narrowed to slits. Beyond this point the vessels were soft and patent. The left common carotid and left subclavian arteries were almost completely occluded in their proximal portions by calcified fibrofatty material (fig. 6). The left common carotid above the clavicle and the left subclavian and its distal branches were soft and patent. The distal abdominal aorta, common iliae, and left femoral arteries were completely occluded by rubbery, dark yellow clot. Microscopic examination of the aorta showed severe atherosclerosis but no evidence of syphilis or other arteritis. The innominate and left common carotid artery were markedly atherosclerotic and occluded by an old organized canalized thrombus.

The heart weighed 530 Gm. and extensive old infarcts were found in the interventricular septum and posterolateral left ventricle. Recent mural thrombi were present in the lateral wall and apex of the left ventricle and in the right atrial appendage. There was severe coronary atherosclerosis, the lumens being reduced to pinpoint size in many areas. Microscopically old infarction and extensive recent infarction (about 48 hours old) were
found in the lateral left ventricle and the right atrium.

The brain weighed 1,250 Gm. Multiple small friable and slightly discolored areas of infarction in the cortex and subcortical white matter on the superior medial region of both cerebral hemispheres were found that were of varying age and ranged in size from half a gyrus to minute microscopic areas. These lesions were distributed in the "watershed areas" of the anastomosis between the anterior cerebral arteries and the middle cerebral arteries and between the middle and posterior cerebral arteries, suggesting that they were due to diminished hydrostatic pressure. Many of the lesions consisted of reacting microglia rather than more severe infarction or cavitation, also suggesting a markedly diminished, but not completely deprived blood supply.

Case 3. A 54-year-old unmarried plumber, was admitted on July 4, 1956, with scrotal swelling for 1 week. Three years earlier he developed intermittent claudication and femoral arteriograms showed obliteration of both profunda femoris arteries and the left superficial femoral artery. A homologous femoral arterial homograft gave temporary improvement but it thrombosed after 11 months and a left midthigh amputation was necessary later.

On physical examination the blood pressure was 150/90 in the right arm, and no pulse or blood pressure could be obtained in the left arm. Both carotids, the right radial, both femoral, and the right dorsalis pedis arteries were normal. The right popliteal and posterior tibial pulsations could not be felt. The right scrotum was swollen, red, warm, and tender.

The urine showed albumin. No white cells or bacteria were seen in the urinary sediment. The hemoglobin was 11.2 Gm. per cent, and the white blood count was 20,500 with 80 per cent neutrophils. Acidosis and azotemia were present and an electrocardiogram suggested an old posterior myocardial infarct. X-ray of the chest showed cardiomegaly with no abnormality of the aorta. Blood Hinton tests for syphilis were negative on 4 occasions.

On the ninth hospital day a right orchideectomy was performed and acute orchitis and epididymitis with abscess due to Bacillus coli were found. The patient became progressively uremic following surgery and died on the eighteenth hospital day.

Autopsy showed severe atherosclerosis involving the entire aorta. Large ulcerated atherosclerotic plaques, with mushy red-brown grumous material at their bases, involved about one fifth of the aorta and were most marked in the distal thoracic...
and abdominal aorta. Between the ulcerated plaques were many thickened, yellow, lipid streaks. The aortic valve and coronary ostia were normal, and the ascending aorta and arch were not dilated. The origin of the left common carotid artery was narrowed to a slit and the left subclavian artery was occluded near its origin by atherosclerotic plaques (fig. 7). The innominate artery was relatively free of atherosclerosis. The left external iliac artery was occluded by a fibrofatty thrombus and the remaining iliac vessels showed severe atherosclerosis.

Microscopic examination showed severe atherosclerosis, with no evidence of syphilis or other arteritis. The left subclavian and left common carotid arteries were almost occluded by severe atherosclerosis with superimposed, organized and canalized thrombus.

The heart weighed 600 Gm., with a thick left ventricle. An area of old fibrosis involved the posterior left ventricle and septum and there was questionable mild old rheumatic mitral valvulitis. The coronary arteries showed diffuse atherosclerosis with an old occlusion of the right coronary artery midway in its course.

The kidneys weighed 85 Gm. each and showed old pyelonephritis, nephrosclerosis, and healing lower nephron nephrosis.

The brain weighed 1,300 Gm. and the cerebral vessels showed minimal atherosclerosis. Microscopic examination showed multiple small areas of cortical infarction.

Case 4. A 54-year-old woman was admitted on December 5, 1952, because of aphasia.

Seven months earlier no blood pressure was obtainable in either arm. Three months prior to admission she developed pain in the right eye, poor vision, unsteadiness, slight aphasia, and thick speech.

Three siblings had diabetes mellitus. For many years she had an excessive alcohol intake but stopped 3 years previously.

On physical examination the blood pressure was 150/80 in the left leg. Both radial and brachial arteries, the left carotid artery, and both posterior tibial arteries could not be palpated. Pulsations of the right carotid artery were felt by some observers but not by others and both femoral arteries and dorsalis pedis arteries pulsed. Coarse rales were heard at both bases. The left pupil was slightly larger than the right and was irregular and there was a right homonymous hemianopsia and a sustained nystagmus on lateral gaze. Hearing was decreased in the right ear. Coordination was slow on the right. She was disoriented as to place and time. She could follow simple commands but not complex ones. Speech was slurred and she had anomia aphasia of which she was aware.

Abnormal laboratory findings included a white cell count of 17,000 and a serum cholesterol of 405 mg. per cent. On x-ray the heart, aorta, the neck and skull were normal. Lumbar puncture was negative but a pneumoencephalogram showed ventricular dilatation due to cortical atrophy. An electrocardiogram showed an old posterior myocardial infarct. Hinton tests of blood and cerebrospinal fluid were negative.

The patient’s condition changed little during her hospital stay and for the subsequent 3½ years except for a leg amputation.

Case 5. A 55-year-old male clerk suffered over an 8-year period cardiac pain, intermittent claudication, recurrent episodes of visual loss and pain in the right eye, and finally episodes of tingling of the left thumb and weakness of the left arm.

The blood pressure was 160/80 in the left arm and 80 systolic in the right arm by machine. No pulsation was felt in the right arm or right neck or below the femoral arteries. Blood pressure in the right arm by the indirect machine method was 80 mm. systolic. The left carotid and radial arteries pulsed normally.

Examination of the urine and blood was normal, the serum cholesterol was 280 mg. per cent, an electrocardiogram showed an old myocardial infarct, and the blood Hinton test for syphilis was negative.

After a long period of anticoagulant therapy there was good subjective improvement, and weak pulsations were palpable in the right radial and carotid arteries.

Case 6. A 70-year-old man with diabetes mellitus for 12 years and congestive failure for 3 weeks had no blood pressure in either arm. Pulsations were felt in both radial arteries, but there were no pulsations of the dorsalis pedis and posterior tibial arteries. A grade-II systolic murmur was heard at the apex. There was mild pitting
Fig. 5 Top. Case 2. Region of origin of the innominate, left common carotid, and left subclavian arteries viewed from the inside of the aorta. Note the dark areas that represent recent red and brown thrombus formation (A).

Fig. 6 Bottom. Case 2. The innominate (A), left common carotid (B) and subclavian (C) arteries cut in cross section close to their origin from the aorta. Note the severe atherosclerosis and thrombosis that occludes or nearly occludes the arteries.
edema of both ankles and free fluid in the abdomen. The urine was normal. The fasting blood sugar was 231 mg. per cent. Two blood Hinton tests for syphilis were negative. On intra-arterial puncture the blood pressure was found to be 140/70 in the right arm.

The patient died 3 months later at another hospital. Autopsy revealed severe generalized atherosclerosis. The “vessels to the neck” were “tortuous and calcified but patent.” There were severe coronary atherosclerosis and extensive myocardial fibrosis. There was no evidence of syphilis.

Case 7. An 81-year-old woman had absent blood pressure in both arms for 4 years. On physical examination the blood pressure was 130 systolic in the right leg. No pulsations or blood pressure could be found in the arms. There were bilateral subclavian, carotid, femoral, and popliteal pulsations. Physical examination was otherwise unremarkable.

The urine and blood were normal. X-rays of the chest showed cardiomegaly and a tortuous and calcified aorta. X-ray of the ankle showed an old trimalleolar fracture of the right ankle and Paget’s disease of the left os calcis with calcification of the arteries in this region. An electrocardiogram suggested left ventricular hypertrophy and a blood Hinton test for syphilis was not recorded.

The patient died at home 3 years later, allegedly of a “stroke.” No autopsy was performed.

Case 8. A 51-year-old woman had an episode of chest pain lasting a few minutes 10 years before admission. At that time chest x-ray showed cardiomegaly and her blood pressure was said to be more than 200 systolic. One year prior to admission she noticed increasing dyspnea on exertion and a gain in weight, relieved temporarily by mercurial injections.

Four to six months prior to admission she developed anorexia and loss of weight. Four weeks before admission she was given digitalis. Her parents died in their middle 70’s of coronary thrombosis as did a brother at age 54. Two siblings were alive and well.

Pulsations were not obtainable in either arm or in the region of the subclavian, axillary, brachial, or radial arteries. Both carotid arteries had forceful pulsations. Both femoral arteries had poor pulsations and pulsations were absent below them.

The heart was enlarged 3.5 cm. to the left of the midclavicular line and the apex beat was heaving. Dullness and rales were heard at both lung bases, and there was pitting edema of the legs.

Examination of the urine and blood was negative. The nonprotein nitrogen varied from 80 to 114 mg., the cholesterol was 327 mg., and the protein-bound iodine was 1.3 gamma per 100 ml. The radioactive iodine uptake was 22 per cent. The basal metabolic rate was —16.

Chest x-ray and fluoroscopy revealed a greatly enlarged heart with poor pulsations. The aorta was negative. An electrocardiogram showed left bundle-branch block. The blood Hinton test for syphilis was negative on 2 occasions. The blood pressure by the machine method was 150/120 in the right arm. A clinical diagnosis of myxedema was not made although the laboratory data suggested hypothyroidism.

The patient was treated for cardiac failure, but she died after a month; an autopsy was not performed.

Case 9. A 50-year-old woman noted for 1 year muscular weakness and fatigue, particularly in the arms, usually after walking 1 block. Approximately 6 months later she noted transient episodes of complete blindness of the left eye recurring several times daily, unrelated episodes of paresthesia of the hands, nose, and mouth, and aching substernal chest pain, coming on once or twice a week.

The hands and feet were warm and red on examination while the face appeared somewhat cool and clammy. No carotid, brachial, or radial pulses were felt. The femoral pulses were palpable. The blood pressure in both legs was 130/70. No blood pressure could be obtained in either arm by the usual methods. The arm blood pressure by an indirect machine method was 60/45 (fig. 8). A continuous bruit and thrill were noted over the right carotid above the clavicle.

The urine and blood were normal. The serum cholesterol was 394 mg. per cent. Glucose tolerance tests showed a diabetic curve. X-ray of the chest showed a slightly elongated aorta. An
AORTIC ARCH SYNDROME

electrocardiogram demonstrated nonspecific T-wave changes. The blood Hinton test for syphilis and the treponema pallidum immobilization test were positive. The cerebrospinal fluid Hinton test for syphilis was negative.

The patient was treated with penicillin, a strict low-fat diet, and Dieumarol. Two years later, angina pectoris intermittent claudication, and visual disturbances were less prominent than before. Examination at this time showed faint pulsation in the right radial artery and a blood pressure in the right arm of 90/70.

Case 10. A 46-year-old woman 3 years prior to admission gradually developed mood and personality changes, with depression, confusion, poor memory, and weakness and stiffness of the right arm. About 18 months prior to admission after an attempt at suicide she was found to be confused and dysarthric with mood lability. At this time she had a right homonymous hemianopsia and a right spastic monoplegia. Cerebrospinal fluid pressure, skull x-rays, pneumoencephalogram, and electroencephalogram were normal. A few weeks later an episode of cardiac syncope occurred following which atrial fibrillation and poor pulses in the left arm were noted.

Five days before admission she had an episode of confusion accompanied by headache. Immediately before admission she had cardiac syncope again.

Her father died of "angina" and the mother died of a stroke, both at an unknown age. One sister was alive and well.

On physical examination the pulse was 92 and regular, the blood pressure was 140/90 in the right arm, 80/60 in the left. The left common carotid artery did not pulsate. There was a thrill over the right common carotid. The left radial and femoral arteries pulsed poorly and the right femoral artery could not be felt. Crackling rales were heard at both lung bases. There was a moderate systolic murmur at the base of the heart. A systolic bruit was present over the entire abdomen.

The patient was confused and disoriented. Her memory was poor. She named objects and did simple calculations poorly. There was a hypermature cataract in the left eye. The right eye showed some arteriovenous nicking and tortuosity of the vessels. The right arm was weak and spastic. The right plantar response was extensor.

Laboratory examinations revealed albuminuria, elevated white cell count on admission of 23,000 per mm.,* and a serum cholesterol of 354 mg. per cent. A blood Hinton test for syphilis was negative.

X-rays of the chest on admission showed pulmonary edema, which cleared 10 days later. The heart was borderline in size and the aorta was negative. Marked calcification of the abdominal aorta was seen. The left kidney did not fill on intravenous pyelography. An electrocardiogram showed left ventricular hypertrophy and widespread S-T depression and T-wave inversion.

The patient complained of intermittent abdominal pain and later suffered marked dyspnea and left pleuritic chest pain. Pulmonary congestion and left pleural effusion were found. Her cardiac failure improved somewhat but she died at home 4 weeks later and an autopsy was not performed.

DISCUSSION

While atherosclerosis and thrombosis of the internal carotid is fairly common,14-16 atherosclerosis of the arch of the aorta with occlusion of the great vessels is rare as judged by reports in the medical literature. Occasional cases of atherosclerotic occlusion of the common carotid artery extending to the aorta have been recorded17, 18 as have cases with diminution or absence of pulsation in either arm, thought to be due to atherosclerosis.11, 18 Clinical cases with involvement of 2 or more of the great vessels by atherosclerosis are even more rare; only 8 cases being described.11, 20-23 We agree with Ross and McKusick that Bit-torf's24 case is not clearly due to atherosclerosis. The only autopsied case of occlusion of the great vessels due solely to atherosclerotic occlusion that will stand critical analysis is that of Abrams and Gore25 but even in this case underlying syphilitic aortitis cannot be completely excluded on the basis of the data presented. Broadbent's26 patient "had at one
time been a sailor, had lived intemperately and had syphilis,” and at autopsy had a dilated ascending aorta; no microscopic examination was made. Two other cases sometimes accepted as being atherosclerotic in origin should be rejected as such, since in one case there was an extensive panarteritis and medial degeneration and in the other extensive elastic degeneration and fragmentation. Dissecting aneurysm of the aorta may have been the disease in Adam’s case, but there was no description of the aortic arch or vessels.

We have no doubt that atherosclerosis and thrombosis was the cause of the disease in our 3 autopsied cases. These patients had severe diffuse atherosclerosis, intermittent claudication, and coronary heart disease. Despite intensive search there was no evidence of aortitis. Although thrombus was present in the left ventricle in case 2, and this might suggest an embolic origin of the occlusions, the histologic appearance of the occlusions of the great vessels indicated that they had preceded the thrombi in the heart.

We can only guess at the etiology of the remaining cases. Case 9 was the only instance with positive serologic tests for syphilis and may represent syphilitic aortitis. The remaining cases illustrate the difficulty of diagnosis without the benefit of an autopsy. There is evidence of generalized atherosclerosis, or factors predisposing to atherosclerosis, in most of them and we consider that they may be due to atherosclerosis, but syphilitic aortitis or other more obscure forms of aortitis cannot be excluded without complete autopsy. Case 8 had laboratory evidence of myxedema, which may have been responsible for her large, poorly pulsatile heart and could have predisposed to atherosclerosis.

The family histories of these 10 patients as obtained from the hospital records are of interest and indicate a high incidence of coronary heart disease. Of the total of 20 parents 6 died of coronary heart disease and 2 more were thought to have died of coronary disease. None of the parents was listed as having diabetes and only 1 was said to have an elevated blood pressure. There were 33 siblings and 4 of the siblings were known to have coronary disease and 4 more thought to have coronary disease. Four siblings were known to have diabetes and 2 were said to have an elevated blood pressure.

It is thought that the disease is the same in the 3 autopsied cases as occurs in people with atherosclerosis although the element of thrombosis was prominent. The only unusual feature in these patients seemed to be the location of the atherosclerosis and thrombosis to involve the arch of the aorta particularly at the ostia of the large vessels. The gross association of thrombosis and atherosclerosis in these autopsies can be used to support Du- guid’s concept that thrombosis “instead of being an occasional complication of atherosclerosis, is an important factor in its pathogenesis.” Estimation of fibrinolytic activity of the blood was not done.

The sex incidence in the 10 cases reported here is equal, indicating that the disease does not have a predilection for either sex. This is in distinct contrast to the variety that seems to affect the younger people in Japan and that is predominantly a disease affecting females (111 females and 11 males), suggesting an inherited sex-linked factor.

The nomenclature of this group of diseases of the aortic arch remains a problem. The symptoms are similar because they affect the same anatomic region. The syndromes so produced may vary slightly because of the underlying disease process. For example, the aortic arch syndrome as it occurs in Japan would also include the facts that the patients are usually young, female, have a strongly positive tuberculin test, and an elevated erythrocyte sedimentation rate. The best term for the whole group would appear to be “the aortic arch syndrome” and we would describe our cases as “the aortic arch syndrome, due to atherosclerosis and thrombosis” (cases 1, 2, and 3), “probably due to syphilis and atherosclerosis” (case 9), and “of unknown etiology, possibly atherosclerosis” (remaining cases). Many other names have been used to describe this whole group. Of these, “chronic subclavian-carotid syndrome” is one of the best, but perhaps the innominate artery should not be excluded as it is by this
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term. "Reversed coarctation,"30-32 although a striking term, has already been criticized. "Pulseless disease" is manifestly a poor name, since there are only a few cases in which pulses cannot be felt clinically, and pulses in the legs are often easily palpable. Furthermore, we believe that even in the cases where pulses were not clinically palpable, pulses would be detected by the machine mentioned above. The disease is "pulse-poor" rather than "pulseless." The eponymous use of Takayasu10 or Martorell22 to describe this group of diseases has little to recommend it since Adams1 described the first case.

A second problem is the term used to describe the aortic arch syndrome caused by arteritis in young females. The most commonly used term is "Japanese pulseless disease," but since this disease is neither Japanese nor pulseless, it seems a poor name.

Our attention was drawn to the striking symptom of intermittent blindness (amaurosis fugax) in cases 1, 5, and 9. In case 1 this was related to exertion and in case 5 it was on the side of the completely occluded carotid artery. Case 9 was observed in an attack and stasis of the retinal arteries and veins was seen then. This symptom would thus appear to be due to transient ischemia of the eye and due to the diminished blood flow in the ophthalmic artery. The changes in the eye grounds of case 1 were thought to be similar to those described in "Japanese pulseless disease," and they may represent changes due to diminished retinal blood pressure. The eye findings of this patient and the changes found at autopsy and their significance are to be described in detail later.33

TREATMENT

Various forms of treatment have been used in the aortic arch syndromes including surgery,20, 22 antispasmodics,34 corticosteroids,35 anticoagulants,23 and a combination of the last two.9, 36 Fairly good results have been claimed for anticoagulants,23 with which we would agree. The progress of the disease was halted in 2 of our cases that have been kept on long-term anticoagulant therapy; these patients perhaps even improved slightly, with pulsation appearing in previously clinically nonpulsatile vessels. Case 9 also was treated for syphilis, however, and was given a low fat diet; while she was definitely improved, it is not possible to decide which method of treatment was responsible. The dietary restriction of fat with weight reduction may also be of help in patients with hypercholesteremia.

SUMMARY

Ten patients are presented in whom absent arterial pulsations were noted in 1 or both arms. Six patients also had absent carotid arterial pulsations on 1 or both sides.

The blood pressure was determined satisfactorily by a machine devised to record the Korotkov vibrations. This indicates feeble arterial pulsation in spite of inability to palpate them clinically.

Three autopsied cases showed severe ath erosclerosis with superimposed thrombosis.

Long-term anticoagulation used in 2 patients for as long as 3 years seemed to result in some clinical improvement.

SUMMARIO IN INTERLINGUA

Es presentate 10 patientes in qui le absentia de pulsation arterial esseva notate in 1 o ambe bracios. Sex del patientes haveba etiam absentia de pulsation del arteria carotidic a 1 o ambe lateres.

Le tension de sanguine esseva determinate satisfactorimente per medio de un machina construite pro registrare le vibrationes de Korotkov. Iste methodo reflecte debile pulsationes arterial in despecto del facto que illos non es clinicamente palpabile.

Tres necropsias mostrava sever atherosclerosis con superimposition de thrombosis.

Medication anticoagulatori, usate in 2 del patientes durante periodos de usque a 3 annos, pareva resultar in un certe melioration clinic.

REFERENCES


3. Sato, T.: Ein seltener Fall von Artcrien-


33. Smith, Taylor, and Dowling, J.: To be published.


The Aortic Arch Syndrome (Pulseless Disease): A Report of Ten Cases With Three Autopsies
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