Transient Embolic Aorto-Arteritis

Presentation of a Patient

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SUMMARY The clinical and postmortem data from a 14-year-old girl who had transient embolic aortoarteritis are presented. This disease was first described in 1974 by Peiris and Wickremasinghe in Hindu patients and is characterized by repetitive embolic occlusion of intra- and extracranial vessels of the brain, originating from focal thrombotic lesions in the aorta and its primary branches. The main histopathological lesion is located in the vasa vasorum of the media in these arteries with alterations that vary from mild arteritis of the vasa vasorum to degeneration and destruction of the media which predisposes to aneurysm formation. This new entity apparently affects only adolescents and young adults and has no relation to atherosclerosis or other non-specific arteritides previously described. To our knowledge, this is the first such patient reported in America.

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THIS REPORT presents data on a patient at the Hospital Universitario of the Universidad Autonoma de Nuevo Leon with "transient embolic aortoarteritis," a new clinico-pathological entity, recently described by J. B. Peiris and H. R. Wickremasinghe, of Sri Lanka. A 14 year old girl was admitted to the hospital August 8, 1979, because of shortness of breath and abdominal pain.

Past medical and family histories were unreliable and scanty. One of the patient's older sisters, who died at the age of 22, had cardiac surgery at the age of 10 and had had pulmonary tuberculosis. The patient's present illness started 2 months prior to admission with progressive shortness of breath and orthopnea. Four days before admission the patient developed paroxysmal nocturnal dyspnea. She also had epigastric intermittent colicky pain, radiating to both upper quadrants. She had lost several kilograms in weight and complained of asthenia and anorexia.

Physical examination revealed an underdeveloped girl, with malnutrition and generalized pallor who was uncooperative. Her blood pressure was 90/50 (decreased on the right side and was barely detectable on the right). There was a pulsatile mass in the right supraclavicular region approximately 4 cm in diameter. Bilateral jugular engorgement was observed when her head was at an angle of 45°. She had bilateral basal rales. There was a carotid pulse decreased on the left side and a holosystolic murmur, grade III-IV, more pronounced in the left interscapular region. She had grade I-II hepatomegaly, but her liver was not tender and was without nodules. Pulses were absent in the upper extremities. She had generalized muscular hypotrophy.

Neurological examination revealed that she was confused and unable to cooperate. No speech abnormalities were found. Cranial nerve examination was unremarkable except for anemic retinopathy. The tendon reflexes were diminished throughout. Laboratory evaluation on admission showed Hb: 4.6 gm. Hct: 17, WBC: 13,700 with 70% neutrophils, 30% lymphocytes. Her blood chemistry was: glucose: 100 mg%, BUN: 13.6 mg% creatinine 0.8 mg, arterial blood gases with pH of 7.54, were Po2: 64, PCO2: 33, HC 3: 28 with normal blood electrolytes. Her platelet count was normal.

Chest x-ray showed increased bilateral pulmonary vasculature with bilateral pulmonary edema. She had cardiomegaly and there was opacity in the upper mediastinum (right side), with a prominent pulmonary artery. Fluoroscopy of the chest showed that the mediastinal mass was pulsatile. The ECG showed unspecific changes of ST and T waves.

Clinical Course

The diagnosis of left cardiac failure was established and believed to be probably secondary to her anemia. Congenital heart disease was suspected. Treatment with digoxin and furosemide was instituted. Also considered as a possible cause was a vascular mass in the upper mediastinum on the right side; malignancy was not excluded. No explanation of the anemia was found. The patient developed a second degree heart block (Wenckebach type) and she was transferred to the Intensive Care Unit. The cause of the heart block was considered secondary to digitalis intoxication. She developed paroxysmal auricular tachycardia which was controlled with 2.5 mg of I.V. verapamil. The patient returned to her room 40 hours later and the heart block was now first degree with sinus cardiac rhythm. She was transfused with elevation of Hb to 10.7 gm and the signs of cardiac failure disappeared. On Aug. 13, 1979, the patient developed behavior changes, delirium and a right hemiparesis which progressed rapidly to quadriaparesis and coma.

She was transferred again to the Intensive Care Unit and, on Aug. 15, right carotid angiography showed complete occlusion of the right internal carotid artery at its origin with collateral circulation.

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through the occipital artery and with retrograde filling of the vertebral–basilar circulation. Left carotid angiography showed thrombosis of the internal carotid artery at the siphon, with important collateral circulation through the occipital artery and retrograde filling of the vertebral–basilar system with occlusion of the right posterior cerebral artery.

A right femoral artery catheterization was performed but the catheter could not be passed through the aortic arch. The films demonstrated multiple aneurysmal dilatations of the thoracic and abdominal aorta with occlusion of both subclavian arteries and stenosis of both renal arteries with normal intrinsic circulation in both kidneys. In view of these findings pulmonary angiography was performed showing a good filling of the right side of the heart, with marked pulmonary circulation, filling of the left ventricle and the pulsatile mass at the origin of the left subclavian artery and occlusion of both subclavian arteries. There was good filling of the innominate artery and the left common carotid artery as well as both internal and external carotid arteries on both sides. A biopsy of the superficial temporal artery was performed as well as a lumbar puncture. Both tests were reported normal.


**Postmortem Examination**

The brain weighed 1200 g. There was cerebral edema and the entire right hemisphere was soft (see fig. 4). There was herniation of the tonsils of the cerebellum and hippocampal gyrus. The right and left middle cerebral arteries were occluded (fig. 1) by a recent thrombus and there was no evidence of atheromata.

**Arterial Pathology**

There were 3 aneurysms of the right subclavian artery (5×4×3 cm) at 1.5 cm from its origin. The root of the left subclavian artery was also dilated (1.8 cm). The largest aneurysm was in the aorta starting at the left subclavian artery and extending down to the renal arteries. In all, there were mural thrombi which extended into the lumen of both subclavian and common iliac arteries. The occlusion in the subclavian arteries extended for 2.5 cm but only 0.5 cm in the right and left common iliac arteries. The left external iliac artery was also occluded by a recent thrombus. No atheromata was seen (figs. 2 and 3).

The heart was normal. Lungs, liver, spleen, and kidneys were congested.
Microscopic Examination

Multiple representative sections from the elastic arteries and peripheral arteries were examined microscopically. In all of the central elastic arteries there were active or healed lesions involving mainly the media. The aorta and both subclavian arteries showed active lesions of many small but confluent foci of proliferating dilated vasa vasorum surrounded by an inflammatory exudate of polymorphonuclear leucocytes, lymphocytes, and macrophages. There was fragmentation and destruction of elastic lamella. This was more evident after staining for elastic fibers (figs. 4, 5, 6). There were relatively recent or initially organizing mural thrombi adherent to the intima that were focally edematous with slight fibroblastic hyperplasia. The extensive destruction of the elastic lamina contributed to formation of aneurysms in these arteries.

Sections from other central elastic arteries showed healed lesions consisting of areas of fragmentation and depletion of elastic lamella with fibrous scarring and occasional calcification (figs. 7 to 10). In some places small vasa vasorum were seen and inflammatory exudate was usually absent. Thromboemboli were seen in the lumina of these arteries except in the left common carotid artery and its branches. There were no atheromatous plaques or fatty streaks in any of these sections. There was a very small (0.3 cm) recent thrombus adherent to the endocardium in the left ventricle. At this site the endocardium showed changes similar to those seen in the intima of the aorta and subclavian arteries. Some of the peripheral cerebral arteries had recent thromboemboli in the lumen associated with small cerebral infarcts and/or signs of....

FIGURE 3. Aortic arch and branches showing the aneurysms and thromboembolus occluding the lumina.

FIGURE 4. Upper surface of brain. Recent infarcts and marked softening of right hemisphere.

FIGURE 5. Aorta. Proliferating vasa vasorum in the media and polymorphonuclear leucocytes exudate.

FIGURE 6. Right subclavian artery. Elastic fibers stained to demonstrate fragmentation of elastic fibers scarring and a few mononuclear inflammatory cells.
cerebral ischemia. No emboli were present in organs other than the brain.

Discussion

This patient presented a serious diagnostic problem. Even with the information obtained from the postmortem examination it was difficult to integrate the clinical and pathological findings into one nosological entity. After reviewing the paper published by Peiris et al. the diagnosis of this clinico-pathological entity was clarified.

Peiris et al. described 10 patients with this disease. The natural history and the probable pathogenesis of transient embolic aorto-arteritis (TEAA) are summarized as follows:

The primary arterial lesions are found in the aorta, the innominate artery, the common carotid arteries and the origin of the internal carotids and subclavian arteries. These arteries, classified from the histological point of view, are elastic arteries.

The earliest histopathological change in the primary lesion is a focal fragmentation of the elastic lamina of the media. This lesion is followed by an arteritic process in the media, characterized by one or more exudative, discrete, inflammatory foci which may extend into the intima and may be associated with mural thrombi on the endothelial surface.

Occlusion of the intracranial arteries is caused by emboli originating in the mural thrombosis of the aorta and the great vessels of the neck. Clinically, emboli are manifested by transient ischemic attacks and cerebral infarction either in evolution or established. The topography of the thrombotic lesions in the elastic arteries and their relationship with the infarcted areas confirm the thromboembolic pathogenesis of this type of stroke.

After a lapse of 3 to 6 weeks, there is healing of the lesion in the media which leaves small foci with fibrosis with elastic lamina absent.

Due to repeated emboli from one or more of the primary arterial lesions, the patients had repetitive transient cerebral ischemic attacks and changing clinical patterns in their neurological symptoms and signs which can occur several years apart.

Degenerative atherosclerosis does not appear to play a role in the pathogenesis of these arterial lesions.

Some patients may have an increased sedimentation rate during the process of aorto-arteritis. When this heals, the sedimentation rate returns to normal.

In the analysis of the published patients it was demonstrated that there is no relationship of transient embolic aorto-arteritis with rheumatic fever, rheumatoid disease, polyarteritis, systemic lupus erythematosus, temporal arteritis, polymyalgia rheumatic, Takayasu's disease, moyamoya disease or syphilis, and no relationship either with cervical trauma or cellulitis in the neck. In the published reports it was found that the histopathological lesions affected the
elastic arteries, which showed arteritic lesions in the media. The histopathological picture indicates that the elastic tissue of this layer is the main target in this disease.²

The focal lesions of the media sometimes resemble Aschoff nodules of rheumatic fever, but the clinical, serological and histopathological findings do not support this association.

In 3 of 10 published reports of TEAA, active tuberculosis was found but no bacteriological or serological evidence has been found to support the possibility of TB as the cause of TEAA.

Transient Embolic Aorto-Arteritis does not have the natural history of Takayasu’s disease which is a progressive chronic aortitis. TEAA is characterized by intermittent cerebrovascular insufficiency. The lesions in Takayasu’s disease and in TEAA both affect the elastic tissue of the media of large arteries and in some there is an association with active tuberculosis. Our patient had evidence of tuberculosis in the lung and in the liver (apparently inactive).

An important point about this clinical-pathological entity is that it affects predominantly children, adolescents and young adults. The literature regarding the pathogenesis of occlusive cerebrovascular disease in this age group is often obscure and there is often no evidence of atherosclerosis or infectious, active arteritis. TEAA may be considered as a variant of moyamoya disease, (which has been described by Taveras as “multiple progressive occlusion of the intracranial arteries”).³ This is supported by the fact that both entities have a similar pathogenetic mechanism (thromboembolism), but the angiographic patterns seen in moyamoya are not found in TEAA.

Another important feature in the latter is that the arterial lesions showed different stages of activity and healing in the same patient which suggest an additional factor that can produce multiple arterial occlusions which progress over several years.

References
Transient embolic aorto-arteritis. Presentation of a patient.
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