Association of Cystic Medial Necrosis of the Aorta and Undiagnosed Thyroiditis

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Introduction.

We have recently seen two patients with cystic medial necrosis of the aorta. The first patient died of a dissecting aneurysm of the thoracic aorta. At autopsy, classical Hashimoto's thyroiditis was discovered. The second patient died of a rupture of the ascending aorta. At autopsy, chronic thyroiditis was seen with multiple large germinal centers and diffuse fibrosis. Neither patient was clinically suspected of thyroid dysfunction although the second patient had had a partial thyroidectomy in the remote past.

The association of dissecting aneurysm and post-thyroidectomy myxedema in three patients was reported many years ago.[1] Autoimmune thyroiditis with symptoms of hypothyroidism and dissecting aneurysm have also been observed together.[2] It has also been reported that aortic aneurysm appears to be significantly more frequent in myxedematous patients than in other hospitalized patients.[3] The underlying theme in these clinicopathological correlations appears to be that hypothyroidism may cause weakening of the aortic wall. This report is meant to focus attention on this possibility and point out that subclinical cases of hypothyroidism may also involve the aorta.

Case Report 1.

A 76-year-old black woman who had a history of mild hypertension was brought to the emergency room because of sudden low back pain and "spells" earlier the same day. Her vital signs were a pulse of 68/min, blood pressure 120/84 mmHg, respiration 44/min. She had prominent epigastric aortic pulsations. A chest x-ray showed cardiomegaly and a widened mediastinum probably due to the thoracic aorta. She had multiple PVB's on her electrocardiogram. She suddenly became unresponsive after her physical exam and died.

The autopsy was performed 17 hours after death. There was a dissecting aneurysm of the thoracic and abdominal aorta and a hemopericardium. The aorta showed mild to moderate atherosclerosis, but the thoracic and abdominal aorta were grossly pliable. Marked cystic medial necrosis was seen with hematoxylin and eosin staining and was confirmed by the significant metachromasia seen with the aldehyde fuchsin and toluidine blue stains. The heart weighed 450 gm and the left ventricle was 2 cm thick. Moderate atherosclerosis of the major coronary arteries was present and the myocardium had moderate diffuse interstitial fibrosis. The thyroid had a diffuse lymphocytic infiltrate together with germinal centers and Hürthle cell change. The morphological criteria for Hashimoto's thyroiditis were therefore fulfilled.

Case Report 2.

A 55-year-old black woman experienced the sudden onset of severe sharp midepigastric pain while ascending a flight of stairs and immediately lost consciousness. She was brought to the emergency room in a state of shock. After admission the patient had generalized seizures followed by episodes of ventricular tachycardia and ventricular fibrillation. She responded to resuscitation at first but later developed cardiac arrest and died. She had no history of hypertension and had been taking no medications. Ten

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years prior to this episode she had undergone a partial thyroidectomy for thyroid nodules.

An autopsy was performed four hours after death. A large rupture in the wall of the ascending aorta extended into the adventitia of the aorta and pulmonary arteries. A hemopericardium containing 100 cc of blood and clot was present. The abdominal and thoracic aorta were markedly atherosclerotic. Marked cystic medial necrosis was seen in the ascending aorta and significant metachromasia was demonstrated with the toluidine blue stain. The heart weighed 450 gm and showed left and right ventricular hypertrophy. The left anterior descending coronary artery was 50% occluded by atherosclerosis within 1 cm of the coronary ostia. Scattered areas of early myocardial necrosis were seen microscopically. The thyroid was small and firm, and one lobe had been previously removed. A marked chronic thyroiditis composed of diffuse lymphocytic infiltration, prominent germinal centers, and scarring was present, but there was no Hürthle cell change. The partial thyroidectomy done ten years previously showed the same histologic pattern. A single sigmoid kidney was present on the right as a result of crossed renal ectopia with fusion.

Discussion

The two patients in this report both had cystic medial necrosis of the aorta and died as a result of complications of the weakened aortic wall. Both patients were hypertensive based either on history or on the autopsy findings of cardiomegaly and left ventricular hypertrophy. Systemic hypertension is often present in patients dying of dissecting aneurysms[4] and ruptures of the aorta. Whether the hypertension has a causative role in cystic medial necrosis or only functions to propagate a tear is not fully understood.

Both of the patients in this report had undiagnosed thyroid diseases at autopsy. In the first patient Hashimoto's thyroiditis involved the entire gland. The second had had a partial thyroidectomy and the remaining thyroid tissue showed chronic thyroiditis. There was diffuse fibrosis and multiple germinal centers, but no Hürthle cell change was seen in this patient and we were therefore reluctant to classify this as a classical Hashimoto's thyroiditis. The anatomic evidence suggests that both of these patients could have had episodes of hypothyroidism which were not detected, so-called preclinical myxedema.

The effect of total thyroidectomy without thyroid hormone replacement in hypertensive patients has been documented by Kountz and Hempelmann.[1] Two patients died of dissecting aneurysm within six months. A third patient was treated with desiccated thyroid for two years. She stopped taking her replacement hormone and ten months later died of dissecting aneurysm. All three patients had cystic medial necrosis. The findings suggest that hypothyroidism is a severe risk factor in the development of dissecting aortic aneurysm in hypertensive patients.

More recently an association of Hashimoto's thyroiditis and dissecting aneurysm was reported.[2] This case involved a patient who was also hypertensive and clinically hypothyroid. Thyroid auto-antibodies were present. Medial degeneration was not documented, but severe atherosclerosis was present. An association of hypothyroidism and abdominal aneurysm has also been shown by Niarchos and Finn.[3] They suggest that preclinical myxedema patients might be picked up with a more sensitive screening test, such as with antithyroid antibodies or serum thyroid stimulating hormone (TSH) levels. In addition, we suggest that patients with aortic aneurysm might be screened more carefully for evidence of preclinical myxedema. In such patients, further weakening of the muscle wall might possibly be prevented.

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