SEGMENTAL AORTITIS AND AORTIC INCOMPETENCE

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ABSTRACT

An unusual case of a middle-aged male with segmental aortitis and aortic incompetence is described. To our knowledge, only 13 cases have been previously described in the literature, but only two have had anatomopathological confirmation.

Aortic incompetence is a rare manifestation of segmental aortitis. Although intrinsic abnormalities of the aortic valve have been proposed, the finding of a normal aortic valve at postmortem examination in 2 of the 3 cases in which the aortic valve was examined, supports the premise that the incompetence is probably secondary to disproportionate dilatation of the aortic root.

INTRODUCTION

Savory in 1856 first reported a case of arteritis of unknown etiology affecting the aorta and its branches. In 1908 Takayasu, a Japanese ophthalmologist, described a young woman with cataracts and abnormalities of the arteries and veins around the optic discs. Onishi found that these ocular changes were associated with absent radial pulses. In the years following these reports, the clinical and pathological features of segmental aortitis (Takayasu’s Disease) have been defined.

Aortic incompetence is a rare manifestation of segmental aortitis. Approximately 13 cases have been described in the literature. In 10 of them the diagnosis was established on clinical grounds and only 3 underwent aortography. Pathological description of the aortic valve exists in only 2 cases.

This paper reports a case of segmental aortitis and aortic incompetence in a middle-aged male with angiographic and anatomopathological confirmation.

CASE REPORT

F. C., a 42-year-old male was admitted to our institution for cardiac evaluation. Twelve years prior to admission the patient developed arterial hypertension. Four years later a heart murmur was first heard. The patient did well until 3 years prior to admission when he developed dyspnea on exertion, easy fatigability and exertional chest pain. The symptoms...
slowly progressed to dyspnea on minimal effort and intermittent episodes of orthopnea. Associated symptoms included paresthesias of the external aspect of the left leg and sexual impotence of 2 years duration.

Past history disclosed viral meningitis 20 years prior to admission and a bout of gross hematuria 1 year before admission. There was no history of diabetes mellitus, syphilis or rheumatoid arthritis.

On physical examination, the patient appeared as a well-developed and well-nourished male. The blood pressure in the upper extremities was 110/60 and 160/55 mm Hg. in the lower extremities. The jugular venous pressure was not elevated. The lungs were clear to percussion and auscultation. The heart was enlarged, the point of maximal impulse was palpable

Fig. 1. Chest roentgenogram demonstrating cardiac enlargement with preponderance of left atrium and left ventricle.
at the 6th intercostal space at the anterior axillary line. A systolic thrill was present in the 2nd intercostal space to the left of the sternum. A systolic ejection murmur grade IV/VI radiating to the neck vessels was heard in the aortic area, followed by grade III/VI early diastolic decrescendo murmur that radiated along the left sternal border towards the cardiac apex. There was a ventricular gallop at the apex. There was no hepatosplenomegaly. The pulses were bounding in nature, slightly weaker in the upper extremities when compared to those in the lower limbs. There was no edema, cyanosis or clubbing.

Chest roentgenogram showed cardiac enlargement with preponderance of the left atrium and left ventricle. Calcification of the ascending aorta was observed in the LAO projection (Figs. 1, 2).

![Chest roentgenogram showing cardiac enlargement with preponderance of the left atrium and left ventricle](image)

**Fig. 2.** Note calcification of the ascending aorta.
Admission electrocardiogram showed normal sinus rhythm, normal QRS axis, left atrial and left ventricular enlargement.

Laboratory studies including glucose (fasting and 2 hours postprandial), VDRL, FTA-abs, rheumatoid factor, LE preparation, antinuclear antibody, and serum protein electrophoresis were normal.

Hospital course: Heart catheterization was performed (hemodynamic findings are listed in Table I). Aortic root angiogram demonstrated a dilated ascending aorta with regurgitation of dye into the left ventricle, reflecting moderately severe aortic incompetence (Fig. 3).

Aortic valve replacement was indicated because of the limitation of activities and degree of cardiomegaly. At the time of surgery, a markedly thickened ascending aorta was found with segmental areas of calcification; the aortic valve was incompetent with separation of the cusps secondary to dilation of the aortic root. The ascending aorta was replaced with a 22 mm Dacron graft and the aortic valve with a #3 Cutter-Smeloff prosthesis. When attempting to wean the patient off cardiopulmonary bypass the procedure was complicated by the development of the “Stone Heart Syndrome” which was irreversible. The patient succumbed in the operating room.

AUTOPSY FINDINGS

Gross examination showed left pleural effusion with a small amount of blood in the pericardial sac. The heart weight was 840 gm. Upon opening of the heart cavity marked left ventricular hypertrophy and moderate

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<td>PA</td>
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<td>PCW</td>
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<td>LV</td>
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right ventricular hypertrophy were noted. There was fibrous thickening of the very proximal ascending aorta; the sclerotic process occluded the right coronary ostium and markedly stenosed the ostium of the left coronary. The ascending aorta was replaced 1.7 cm above the coronary ostia with a Dacron graft, 4 cm in length. Beyond the graft there were moderate areas of narrowing of the proximal brachiocephalic and right subclavian arteries. The ascending aorta had a segment 6 cm in length of narrowing with thickening and calcification of the arterial wall (Figs. 4, 5). The rest of the aorta was not involved. Gross examination of other organs showed bilateral pulmonary atelectasis and acute passive congestion.

The aortic valve showed no abnormalities, either on gross or at microscopic examination (Fig. 5).

There was marked thickening of the aortic wall due to collagenization of all layers. The intima had moderate atherosclerotic changes at the coronary ostia. The medial changes included irregular areas of fragmentation and fibrosis. The outer aortic wall accounted greatly for the thickening due to marked fibrosis and obliteration of the vasa vasorum.

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Fig. 3. Aortic root angiogram demonstrating a dilated ascending aorta with moderately severe aortic incompetence.
Fig. 4. Different portions of aorta. In the middle, the largest portion is in the abdominal aorta showing only minimal atherosclerotic changes. Two segments of aortic thickening due to fibrosis are seen on transverse sections. (A. aorta=ascending aorta; D. aorta=descending thoracic aorta). The aortic arch (bottom) shows minimal atherosclerotic change, three leaflets of aortic valve (upper) show no gross changes.

Fig. 5. Close-up view of three aortic valve leaflets and a transverse section through the resected portion of ascending aorta that is markedly thickened due to fibrosis.
A chronic inflammatory infiltrate with a large number of plasma cells was noted throughout these areas, with an occasional lymphoid follicle containing an active germinal center. Marked intimal thickening of the branches of both pulmonary arteries was also found.

The histological features were typical for segmental aortitis, which greatly resemble luetic vascular disease (Figs. 6, 7). However, multiple sections were stained with special silver impregnation techniques and spirochetes were not demonstrated. The findings along with non-reactive serology reactions points toward the diagnosis of a non-luetic segmental panaortitis (Takayasu’s Disease).

**DISCUSSION**

The etiology of segmental aortitis remains unknown. An autoimmune mechanism has been postulated, because the involvement of the aortic arch and its major branches has also been observed in disorders presumably due to an autoimmune mechanism such as systemic lupus erythematosis,\(^5\,14\) polymyalgia rheumatica,\(^20\) and rheumatoid arthritis.\(^15\)

Circulating anti-arterial antibodies have been demonstrated in some patients.\(^21\) There is no definite evidence at present that the circulating antibodies are the direct cause of segmental aortitis. Whether an autoimmunological mechanism may take place in segmental aortitis, or whether the circulating anti-arterial antibodies may be produced only as a result of destruction of the arterial tissue is a question that deserves further investigation.

The basic pathological process is marked intimal fibrosis, fibrous scarring of the media and marked degeneration of the elastic fibers accompanied by chronic inflammatory cell infiltration. Although the process most often involves the arch of the aorta and its major branches, several segments and even the entire aorta, may be involved.\(^3\,8\,22\,23\)

Cardiac symptoms are commonly described. Nakao et al\(^21\) in a review of 84 cases encountered palpitations in 36 patients, dyspnea in 24, and angina pectoris in 11. Cardiomegaly due to left ventricular hypertrophy is commonly seen, usually secondary to arterial hypertension (in the lower limbs) that is seen in over half of the cases. Pericarditis,\(^5\) myocardial infarction and sudden death\(^21\) have occasionally been described.

Although intrinsic abnormalities of the aortic valve have been proposed as responsible for the aortic incompetence,\(^3\,24\) our findings and those described in a clinico-pathological conference of the Massachusetts General Hospital\(^17\) indicate that the incompetence is secondary to disproportionate dilation of the aortic root with secondary separation of the aortic leaflets. The gross and microscopic normal findings of the aortic valve support this premise.

Our case constitutes the first male ever reported with this association; the rest of the cases were females ranging in age from 18 to 40 at the time of the diagnosis.

Congestive heart failure is the most common cause of death when aortic
Fig. 6. Microscopic picture of aortic intima and media. Note a marked thickening due to fibrosis, marked irregularities and focal absence of elastic fibers. Elastic Verhoeff's-Van Gieson: X 106.
Fig. 7. Fibrous thickening of adventitia with multifocal chronic inflammatory change. Hematoxylin-eosin: X 106.
incompetence complicates segmental aortitis. Sudden death has also been described, secondary to involvement of the coronary ostia.21

In brief, aortic incompetence probably occurs in segmental aortitis secondary to disproportionate dilation of the aortic root, rather than to intrinsic abnormalities of the aortic valve. The majority of patients are females with symptoms secondary to involvement of the major aortic branches, (claudication of the arms and jaw, angina pectoris, abdominal angina, or arterial hypertension secondary to obstruction of the renal arteries).5,8,12-18 Segmental aortitis should be included in the differential diagnosis of non-rheumatic aortic incompetence, when associated with calcification of the ascending aorta.

REFERENCES

1. Savory WS: Case of a young woman in whom the main arteries of both upper extremities and on the left side of the neck were throughout completely obliterated. Med Chir Trans Lond 39:205, 1856

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