Aortic regurgitation due to syphilis: unusual case

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SUMMARY A 28 year old man developed signs and symptoms of left ventricular failure and was found to have gross aortic regurgitation. He underwent an aortic valve replacement, and histology showed changes characteristic of syphilis. Both the age of this patient and the mode of presentation were atypical of cardiovascular syphilis.

Case report

A 28 year old heterosexual storekeeper of Portuguese Indian extraction presented with a two week history of cough and progressive shortness of breath, the onset of which coincided with a febrile illness. His appetite had decreased and he had lost over three kilograms in weight. His only ever sexual contacts had been with a prostitute in Goa two years previously and two prostitutes in Kenya 10 years previously. There had been no history of sexual abuse as a child, but 10 years previously he had been tattooed on his forearm.

Physical examination showed evidence of left ventricular failure, cardiomegaly, and the characteristic murmur of aortic regurgitation. Chest radiography, electrocardiography and echocardiography confirmed these findings. His blood pressure was 130/40 mm Hg.

The patient responded to conventional treatment for heart failure, and cardiac catheterisation showed severe aortic regurgitation. Serology showed positive results to the Treponema pallidum haemagglutination assay (TPHA) and the Venereal Disease Research Laboratory (VDRL) test at a titre of 1/256, the erythrocyte sedimentation rate was 47 mm in the first hour, and the white cell count was 12.2 x 10⁶/μl. Treatment was started with prednisolone for 48 hours followed by procaine penicillin 600 mg and probenecid 500 mg every six hours.

An aortic valve replacement was performed, and at operation there was pronounced mediastinal oedema and tense serous pericardial effusion. A size 25 Medtronic-Hall tilting disc prosthesis was inserted.

Histology of the valve showed thickening of the intima with dense hyaline bands of collagen. The media and adventitia showed endarteritis and periarteritis of the vasa vasorum. Dense aggregates of:

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Accepted for publication 22 November 1986

Figure Aortic valve histology showing intimal thickening and obliterative endarteritis of the vasa vasorum and periarteritis, mainly consisting of plasma cells with some lymphocytes (haematoxylin and eosin).
lymphocytes and plasma cells were present, which caused disruption of the elastic fibre.

Discussion

Syphilis of the cardiovascular system is normally preceded by a latent period of 15 to 30 years, and most patients are aged 40 to 55. The three main cardiac problems caused by syphilis are: thoracic aneurysm formation, aortic valve disease, or coronary ostial stenosis, whereas the myocardium is rarely affected directly. There is little evidence of any myocardial sequelae in surviving adolescents and adults with congenital syphilis, although 10% of infants dying of syphilis have myocarditis.1

Aortic regurgitation occurs in 30% of patients with tertiary syphilis, and in a series of 300 cases of aortic valve disease described by Campbell and Shackle, of which 25% were due to syphilis, nearly 80% were aged 40 to 60, and none were younger than 30.2 In Heggtveit's3 clinicopathological necropsy study of 100 cases, the average age was 63, and all patients were over 30.3

The initial symptom of aortic regurgitation in most cases is effort dyspnoea, the duration of symptoms before valve replacement being one to 55 (mean 25.5) months.4 Heart failure as a complication is said to be increasingly uncommon and to occur at a later stage of the disease.5

Valve surgery is not without risk, and one group had a mortality of 20% at operation and late mortality of 40% (unpublished observation) at a time when the mortality for aortic valve replacement in patients without syphilis at the National Heart Hospital was much lower.

The case described was most unusual both in the nature and presentation of symptoms and in the remarkably short latent period.

References

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doi: 10.1136/sti.63.4.277

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