Mediastinal mass caused by syphilitic aortitis

R Hofmann-Wellenhof, W Domeij, C Schmid, D Rossmann-Moore, P Kulling, M Annelli-Monti

Abstract
A 47 year old man presented with hoarseness and chest pain found to be due to proliferative syphilitic aortitis. The case is unusual as the syphilitic aortitis caused a mediastinal mass without affecting the lumen of the aorta.

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Syphilitic aortitis, the hallmark of cardiovascular syphilis, has become rare and is hardly considered by today's clinicians in their differential diagnosis. We describe a patient with a recurrent laryngeal nerve palsy resulting from a mediastinal mass caused by cardiovascular syphilis.

Case report
A 47 year old man was admitted to Karl Franzens University Medical Centre because of progressive hoarseness and angina like chest pain over the previous six months. The patient's past medical history included two episodes of syphilis at the age of 20 and 22 which, according to the patient, had both been treated with penicillin. The presence of coronary artery heart disease (NYHA II–III) had been verified by coronary angiography one year previously but the patient had refused treatment.

Upon admission the patient was in good general health, with hoarseness being the only abnormality on physical examination. There was no evidence of any syphilitic stigmata such as aortic incompetence, Argyll Robertson pupils, tabes dorsalis, or gummas. Laboratory tests revealed a normal haemoglobin, white blood cell and differential count, and platelets. C-reactive protein and sedimentation rate were both elevated. Other routine laboratory tests were within normal limits. Serum VRDL and IgM SPHA (solid phase haemadsorption) test results were negative; TPHA (Treponema pallidum haemagglutination) test results were positive at a low dilution, and FTA-ABS (fluorescence-Treponema pallidum antibody-absorption) test results were also positive, both consistent with previous syphilis.

Laryngoscopy revealed paresis of the left vocal cord and chest radiography showed enlargement of the aortic knuckle with tracheal deviation suggesting an aortic dissection. Computed tomography of the mediastinum revealed a mass which extended from the aortic knuckle down to the bifurcation of the trachea (fig 1). Magnetic resonance imaging showed a mass extending from the carina to 3 cm above the aortic arch which suggested a lymphoma or thymoma. An aneurysm of the aorta was ruled out by these imaging procedures.

In order to identify the nature of this mass, exploratory thoracotomy was planned. Coronary angiography performed before thoracotomy showed progression of his coronary artery disease so coronary artery bypass surgery was planned for the same operation. Further preoperative examinations such as bronchoscopy, carotid artery sonography, bone scan, and angiography of the aortic arch showed nothing remarkable.
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Surgery revealed an extensive solid mass reaching from the ascending aorta to the main vessels of the aortic arch, encasing them as well as the thoracic aorta and left pulmonary hilum. Further exploration showed the coronary arteries to be encased by solid nodular tissue. Microscopic examination of a frozen section of the mass suggested malignant lymphoma; syphilitic aortitis, however, could not be excluded. A large portion of the tumour mass was removed; the planned coronary bypass, however, was technically not possible.

Permanent sections of the mediastinal mass revealed connective tissue with extensive sclerosis, fibrosis, and dense infiltration by small and medium sized, occasionally binucleated, plasma cells interspersed with a small number of Russell bodies and macrophages. Lymphoid follicles and multinucleated giant cells were also seen. Within areas of sclerosis and fibrosis small arteries with endothelial swelling and inflammatory infiltration of the vessel wall were found. A massive infiltration by lymphocytes, plasma cells, macrophages, and giant cells was present near the small vessels (fig 2). These findings are considered diagnostic of syphilitic aortitis.

The patient was treated with high dose parenteral penicillin for three weeks. Computed tomography of the mediastinum three months later showed considerable reduction in the size of the mediastinal mass, mainly as a result of the surgery. The results of serological tests for syphilis showed no change after surgery or treatment with penicillin.

Discussion
Fifty years after the introduction of antibiotics into clinical practice cardiovascular syphilis has become a rare disease. Screening of 971 786 patients admitted to public hospitals in Vienna between 1980 and 1984 detected 28 090 cases of untreated latent syphilis. Syphilis therefore remains an important public health problem.

About one third of patients with untreated syphilis develop severe late complications; 10% have cardiovascular symptoms, 7% neurosyphilis, and 16% a gumma. The most common complications of syphilitic aortitis are aortic insufficiency, coronary ostial stenosis, and aortic saccular aneurysm. The latent period between primary infection and late manifestation may be 5–20 years.

Our patient did not have the common complications of cardiovascular syphilis. Hoarseness was a result of the mediastinal mass, and the cardiovascular syphilis caused the mass but without changes in the aortic intima. Serological tests for syphilis only showed evidence of past infection and aortitis was not suspected as the configuration of the aortic arch was normal on computed tomography.

We suspect that this atypical form of cardiovascular syphilis was the result of insufficient antibiotic treatment at the time of the first or second episode of syphilis some 25 years earlier. Although the occurrence of a mediastinal mass caused by a syphilitic aortitis is a rare occurrence, syphilis should be considered in the differential diagnosis of mediastinal tumours.

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