disordered growth of both endodermal (respiratory epithelium) and mesodermal (vascular) elements satisfies criteria for the pathological diagnosis of a hamartoma. Primitive mesenchyme is fetal tissue. By this definition and according to the hypothesis of Mark, mesenchymal cystic hamartoma of the lung is hamartomatosus and congenital. Only seven cases have been reported to date. The nodules and cysts increase very slowly over a period of many years and, although the disease may be detected in infancy, three or four decades usually elapse before it comes to clinical attention. Neither the epithelial nor the mesenchymal cells have malignant features, but malignant degeneration has been described. The nodules and cysts represent different stages of the disease. The nodules arise from mesenchyme proliferation in the interstitium. When the nodules reach 1 cm in diameter bronchiolectasis occurs and small cysts form, lined with normal or metaplastic bronchiolar epithelium. Bleeding from systemic arteries into the cysts causes haemoptysis, and rupture of subpleural cysts causes pneumothorax or haemothorax. New nodules continue to appear when the cysts have already developed. Surgery is needed for diagnosis and for treatment of a pneumothorax or haemothorax. The possible benefit of resecting nodules and cysts to preclude malignant transformation must be weighed against the multicentric nature and benign course in most patients.

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Aortic valvulitis complicating Wegener’s granulomatosis

A D Fox, S E Robbins

Abstract

In a case of Wegener’s granulomatosis aortic valve replacement was performed for worsening congestive cardiac failure secondary to aortic incompetence. Two paravalvular lesions and an isolated intra-leaflet deficiency of the non-coronary cusp were identified at operation. Historical changes were consistent with a connective tissue disease.

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Case report

A 20 year old man presented with arthralgia, haemoptysis, inflated fauces, conjunctivitis and weight loss. On clinical examination he was distressed, pale, febrile (39–3°C) with a tachycardia (blood pressure 110/80 mm Hg). A vasculitic rash was present on his shins and elbows. Auscultation revealed bibasilar fine crackles and a blowing left parasternal diastolic murmur. He was anaemic (haemoglobin 8.7 g/dl) with a leucocytosis (13.2 × 10⁹/l), urea 12.5 mmol/l and creatinine 199 μmol/l. Radiographic examination demonstrated cardiomegaly and flabby basilar pulmonary opacifications. Echocardiography showed mild aortic and mitral regurgitation, mild left ventricular dilatation, and normal valve cusps. No evidence of pulmonary hypertension or vegetations was demonstrated. Renal ultrasound revealed normal sized kidneys of increased echogenicity consistent with glomerulonephritis. Antineutrophil cytoplasmatic antibody (ANCA) titres (>1/320) confirmed the diagnosis. Treatment with prednisolone, cyclophosphamide and plasma exchange commenced immediately.

Five months later worsening congestive cardiac failure, recurrent epistaxes, pleurisy and anaemia (haemoglobin 5.9 g/dl) necessitated readmission. Echocardiography revealed left ventricular dilatation and partially prolapsing thin aortic valve cusps. Despite treatment his condition deteriorated.

Examination demonstrated a collapsing pulse, blood pressure of 240/120 mm Hg, a displaced apex beat, a precordial thrill, and a pandiastolic murmur. Bibasilar crepitations persisted. A diagnosis of severe aortic incompetence with significant left ventricular failure was made.

Left axis deviation and left ventricular hypertrophy (without strain) was noted on the electrocardiogram. Repeat echocardiography confirmed a moderately dilated left ventricle with good contractility, very severe aortic regurgitation with prolapsing aortic cusps, a normal mitral valve, and an ejection fraction of...
Aortic valvulitis complicating Wegener's granulomatosis

60%. Deteriorating renal function was confirmed (urea 16.8 mmol/l, creatinine 338 μmol/l).

Angiography confirmed severe aortic regurgitation and excluded coronary vasculitis. Two aneurysmal swellings were noted originating from the right coronary and non-coronary sinuses of Valsalva, suggesting involvement of the aortic and para-aortic tissues in the disease process (figure).

Emergency aortic valve replacement was performed. Macroscopically the aortic valve cusps were apposed but a discrete deficiency (6 mm diameter) in the non-coronary cusp was identified. The aneurysmal swellings were well circumscribed diverticula with smooth intimal linings and without evidence of debris or vegetations. The incompetence was due to the defect within the non-coronary cusp. Culture of the valve revealed no growth.

Microscopical examination showed degenerative myxomatous and mucoid changes typical of a collagen vascular disease. Granulomas were not visualised and no neutrophilic or lymphocytic infiltrate was identified.

Treatment for persisting renal failure (creatinine 326 μmol/l, urea 30.5 mmol/l) continued during an uneventful postoperative recovery. Two months later severe left ventricular failure led to readmission and echocardiography confirmed a paravalvular leak. Clinical deterioration necessitated re-exploration and resuture of the valve. No spare tissue was available for further histological analysis. Postoperative recovery was uneventful.

Discussion

Wegener’s granulomatosis tends to begin in the respiratory tract and terminates in a focal necrotising glomerulonephritis, other tissues occasionally being involved. Although Anderson et al did not identify any cardiac complications in their large series of 256 cases, involvement has been reported in 12–30% of patients with the disease. Pericarditis, paravalvular and coronary arteritis may occur. Aortic valve involvement has rarely been reported in the literature, but angiography confirmed this to be the underlying cause in this patient.

Yanda et al described a 77 year old woman who presented initially with a cavitary lung lesion but subsequently progressed with extensive pulmonary, ocular, dermatological, and aortic valvular symptoms requiring replacement. Histological examination showed myxomatous change, degeneration of collagen with an insignificant inflammatory response, a feature consistent with our case. Macroscopically the valve leaflets were thickened but there was no mention of paravalvular involvement.

Although characteristic granulomas were not identified in the aortic tissue, the microscopic changes were consistent with a degenerative tissue collagen vascular disease. The aneurysmal dilatations and the defect in the non-coronary cusp were similar with their macroscopically smooth intimal architecture.

These defects were presumed to be acquired manifestations of the underlying disease process. There was no evidence of an infective cause isolated from either tissue or blood microbiology. This was substantiated by a lack of neutrophil or lymphocytic infiltrate in the specimen.

Although our case required further intervention there was no evidence to suggest that the paravalvular leak was due to extension of the underlying disease process rather than a technical complication of valvular repair.

Review of the literature has not revealed any cases where associated paravalvular lesions were found, and indeed, no reports of acute incompetence secondary to the development of an isolated intraleaflet deficiency.

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