Renal amyloidosis and non-specific aorto-arteritis—a hitherto undescribed association

S. C. DASH
M.D., D.M., F.I.C.A.
K. K. MALHOTRA
M.D., M.A.M.S.
R. K. SHARMA
M.D., M.A.M.S.
U. N. BHUYAN
M.D.

Department of Medicine and Pathology, All India Institute of Medical Sciences, Ansari Nagar, New Delhi 110029, India

Summary

Two patients presented with the nephrotic syndrome complicating non-specific aorto-arteritis (Takayasu's arteritis). Histologically both had renal amyloidosis. On investigation there was no evidence of chronic infection or any immuno-inflammatory disease known to be associated with amyloidosis. These cases raise the possibility of a significant association between aorto-arteritis of the Takayasu type and systemic amyloidosis.

KEY WORDS: Takayasu's disease, nephrotic syndrome, amyloidosis.

Introduction

The association of amyloid with aorto-arteritis was reported by Misra, Prakash and Agarwal (1959), although both their patients had intermittent tuberculosis. However, in the present cases of aorto-arteritis we failed to find any other known cause of amyloidosis.

Case reports

Case 1

An 11-year-old boy was admitted in September 1980 with giddiness, bilateral leg oedema, puffy face and ascites of 3-week duration. The right radial and brachial pulses were feeble. A systolic bruit was audible over the right renal artery. Blood pressures were 158/95 and 86/60 mmHg in left and right arms, respectively.

On investigation, 24-hr proteinuria varied between 4.5 and 5.6 g. Total serum protein was 50 g/litre with albumin of 18 g/litre. His serum cholesterol was 9.84 mmol/litre and creatinine clearance (C₅₀), 70 ml/min. Chest X-ray was normal, Mantoux test was weakly positive. Anti-streptolysin 'O' titre was <200 Todd units and 'C'-reactive protein was normal. Serological tests for syphilis (STS) were non-reactive. Erythrocyte sedimentation rate (ESR) was 62 mm on 1st hr. Serum electrophoresis did not reveal monoclonal gammopathy. Urine was negative for Bence Jones protein. Sputum and urine examination showed no Mycobacterium tuberculosis on smear. Barium studies of the gastrointestinal tract were normal. Renal biopsy showed extensive amyloid deposits replacing the glomerular architecture. The amyloid stained with potassium permanganate. Aortogram revealed irregularities of the abdominal aorta and stenosis of both renal arteries, the left being almost completely occluded (Fig. 1). Aortic archogram showed narrowing at the root of the right subclavian artery. He was treated empirically with anti-tuberculous drugs and prednisolone. Three months later, 24-hr proteinuria had decreased to 0.8 g. However, his blood pressure showed an elevation which improved after withdrawal of the prednisolone.

Case 2

A 22-year-old woman was admitted in February 1981, with a 6-month history of headache, and a 2-month history of leg oedema, oliguria and increasing weakness. She had facial puffiness and her radial and brachial arteries were palpable on both sides. Blood pressure in the right lower limb was 170/100 mmHg.

She had proteinuria of 5.5 g/24 hr and reduced serum protein (total: 52 g/litre and albumin: 25 g/litre). Serological test for syphilis was non-reactive. Anti-streptolysin 'O' titre was <200 Todd units. The ESR was 45 mm in the 1st hr and C-reactive protein...
Discussion

Two cases of renal amyloidosis complicating non-specific aortoarteritis (Takayasu's arteritis) are presented. Histochemically the amyloid was potassium permanganate-sensitive in both cases (Wright et al., 1977). This means deposition of AA protein found in secondary amyloidosis. Mild proteinuria is a not uncommon finding in aorto-arteritis. This is often interpreted as being due to ischaemic or hypertensive renal disease. As kidney biopsy is not generally advised in such cases unless the proteinuria is significant, it is not possible to exclude amyloidosis in all instances.

Reactive systemic amyloidosis (RSA) has been known to occur as a result of various acute recurrent or chronic infections, inflammatory, or neoplastic disorders. Rheumatoid arthritis is reported to be the most frequent predisposing disease in the western hemisphere (Hubsy, 1975) whereas tuberculosis is the commonest predisposing illness of RSA in India (Chugh et al., 1981). Reactive systemic amyloidosis (RSA) has also been reported in ankylosing spondylitis, Reiter's syndrome, dermatomyositis, scleroderma, Behçet's syndrome, ulcerative colitis, Crohn's disease and, rarely, in systemic lupus erythematosus. More recently it is postulated that immunological stimuli act upon 'SSA' synthesizing cell to produce (amyloidogenic) SSA protein which undergoes proteolytic (lysosomal) cleavage to produce AA fibrils (Glenner, 1980).

Misra et al. (1959) observed amyloid in 2 cases of Takayasu's syndrome. However, both these cases had...
intercurrent tuberculosis. In both the cases presented here, even after careful investigations we failed to find any evidence of tuberculosis, bronchiectasis, syphilis, rheumatoid arthritis or intestinal pathology. Absence of monoclonal gammopathy in serum and light chains in the urine ruled out the possibility of multiple myeloma. Some autopsy data (Sen et al., 1973; Kinare, 1976) suggest an association between diseased segment of aortitis and local tuberculosis lymph nodes. Admittedly, lymphangiographic studies to delineate retroperitoneal and intrathoracic lymphadenopathy have not been done in our cases. However, there was no superficial lymphadenopathy in our cases which was recorded in the above-mentioned reports. Therefore, we are inclined to believe that there was no other primary disease, except the aorto-arteritis responsible for amyloidosis in the 2 patients reported here.

References


(Accepted 1 September 1983)
Renal amyloidosis and non-specific aorto-arteritis--a hitherto undescribed association.

Postgrad Med J 1984 60: 626-628
doi: 10.1136/pgmj.60.707.626

Updated information and services can be found at:
http://pmj.bmj.com/content/60/707/626

These include:

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/