

Localised retroperitoneal amyloidosis mimicking retroperitoneal fibrosis: a rare cause of obstructive uropathy

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ABSTRACT

Primary localised amyloidosis involving the retroperitoneum is a rare disease. We report a 71-year-old diabetic man who presented with generalised fatigue, malaise and elevated serum creatinine. Investigations confirmed obstructive uropathy secondary to a retroperitoneal mass behind the urinary bladder, causing extrinsic compression of both the ureters, resulting in bilateral hydroureteronephrosis. Following initial bilateral percutaneous nephrostomies to stabilise renal function, a computed tomography-guided biopsy of the pelvic lesion which was done, was suggestive of amyloidosis. We present this case due to the rarity of localised retroperitoneal amyloidosis as a cause of obstructive uropathy.

Keywords: amyloidosis, localised retroperitoneal amyloidosis, obstructive uropathy, retroperitoneum, retrovesical amyloid deposition

Singapore Med J 2009; 50(9): e332-e335

INTRODUCTION

Systemic amyloidosis represents a group of diseases characterised by an extracellular deposition of amyloid protein. The symptomatology depends on the organ and the region involved. Retroperitoneal amyloid deposition, even though reported in literature, is one of the rarer forms of presentation.⁽¹⁾ Even more uncommon is a localised involvement of the retroperitoneum causing obstructive uropathy. Various reports of retroperitoneal amyloidosis mimicking retroperitoneal fibrosis are available in the literature. We report the occurrence of a localised retrovesical amyloid deposition leading to obstructive uropathy.

CASE REPORT

A 71-year-old diabetic, normotensive man presented with right loin pain, weight loss and malaise of two months' duration. He also had moderate storage and voiding lower urinary tract symptoms for the past two years.



Fig. 1 US images show bilateral hydroureteronephrosis.

He had no haematuria. He was found to have a serum creatinine level of 3.1 mg/dL (272.8 mmol/L), with bilateral hydroureteronephrosis up to the bladder (Fig 1). The erythrocyte sedimentation rate (Westergren method) was 115 mm at one hour and the total leucocyte count was 8,600/mm³ with a predominant neutrophilia. He had no proteinuria. The general examination was normal. The digital rectal examination revealed a hard mass above the level of the prostate and distinctly separate from it. The upper limit of the mass could not be palpated. Rectal mucosa overlying the mass was normal.

Magnetic resonance (MR) imaging of the abdomen and pelvis showed a 13.7 cm × 7.6 cm irregular, well-defined hypointense lesion in the retrovesical region, predominantly on the left side. The mass compressed and encased the left ureter and iliac vessels, pushing the bladder to the right (Figs. 2a & b). The left obturator internus muscle was infiltrated. This lesion also invaded through the sciatic foramen into the gluteal region. After the bilateral percutaneous nephrostomy, his serum creatinine improved to 1.4 mg/dL (123.2 mmol/L). A computed tomography (CT)-guided biopsy (Fig. 3) of this lesion was carried out, and the routine haematoxylin and eosin stains showed fibrocollagenous tissue with deposits of homogeneous pale eosinophilic deposits (Fig. 4a) and focal foreign body reaction. This material was congophilic, and showed an apple-green birefringence on polarising microscopy (Fig. 4b) and fluorescence with thioflavine, suggestive of amyloidosis.

He was subsequently investigated for a possible

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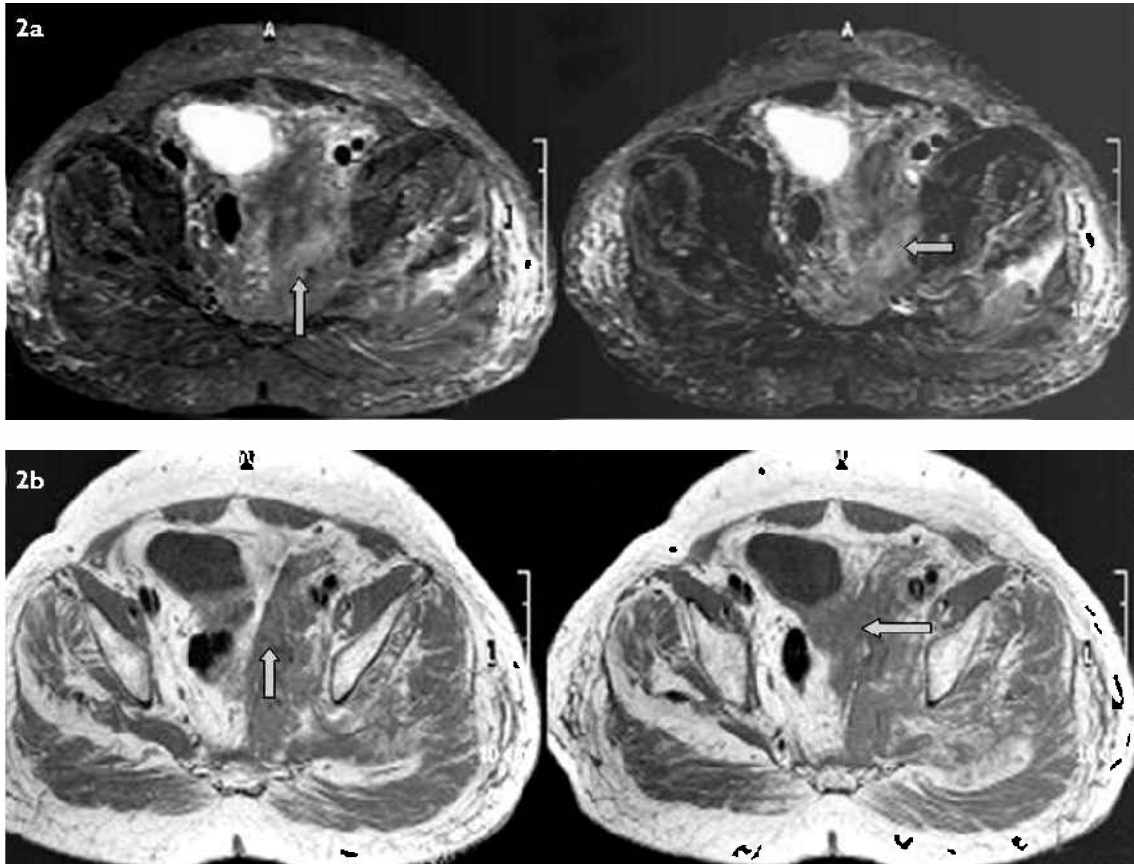


Fig. 2 (a) Axial STIR MR images show the pelvic mass (arrows). (b) Axial T1-W MR images show the pelvic mass (arrows) pushing the bladder to the right.

cause and type of amyloidosis. The urine sample was positive for Bence-Jones protein, using the immunoelectrophoretic method. Serum beta-2 microglobulin was 7.4 mg/L, which was three times the normal range. Serum beta-2 microglobulin was estimated using the immunotubidimetric assay, Spinreact Kit (Spinreact SA, Girona, Spain), and done on the Olympus AU™ series (Olympus Corporation, Tokyo, Japan). Hence, the possibility of amyloidosis secondary to multiple myeloma was considered. However, serum electrophoresis revealed only a generalised decrease in the gamma globulin (Fig. 5) with a total protein of 5.9 g/dL, and an albumin level of 3.4 g/dL.

Bone marrow biopsy showed a normocellular marrow with mild plasmacytosis (6%) with no evidence of myeloma. The above picture thus ruled out multiple myeloma as the cause of amyloidosis. The immunoglobulins IgG, IgA, IgM were 402 mg/dL, 36 mg/dL and 24 mg/dL, respectively. Serum-free kappa and lambda chains were 316 and 14.7, respectively, and the kappa/lambda ratio was 21.5, suggestive of amyloidosis secondary to the non-myeloma variant of plasma cell dyscrasias. The kappa and lambda estimations were done by the turbidimetric method, using the Freelite™ human

kappa and lambda free kits (The Binding Site Group Limited, Birmingham, UK) and done on the Olympus AU™ series. The 95th percentiles for kappa and lambda were, respectively, 3.30–19.40 mg/L and 5.71–26.30 mg/L. The normal kappa/lambda ratio for the kit was 0.26–1.65. He subsequently underwent bilateral antegrade double-J stenting and was started on chemotherapy with dexamethasone and melphalan. On follow-up at six months, urine Bence-Jones protein was negative, haemoglobin was 12.1 g/dL and serum creatinine was 1.2 mg/dL. Both stents were removed, and there was resolution of the hydronephrosis.

DISCUSSION

Systemic amyloidosis, also termed beta fibrillosis, is an all-encompassing term for a group of diseases, and is characterised by widespread extracellular deposition of insoluble amyloid protein.⁽²⁾ Amyloidosis may be primary or secondary. Primary amyloidosis is the immunological light-chain-type variety. Secondary amyloidosis occurs secondary to chronic infections like rheumatoid arthritis, tuberculosis or familial Mediterranean fever, and these have the amyloid A protein. There is also a variant which is common in the United States called the familial



Fig. 3 Axial CT image shows the mass on left side of the pelvis extending to the gluteal region.

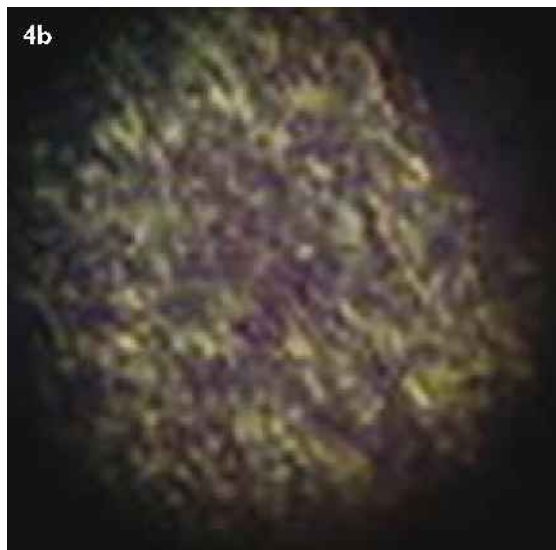
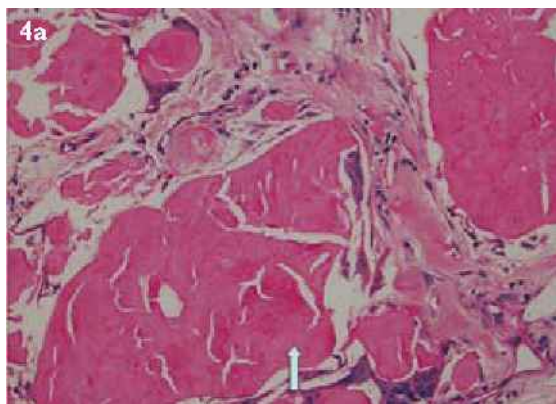


Fig. 4 (a) Photomicrograph shows amyloid deposition (Haematoxylin & eosin, $\times 100$). (b) Congo red-stained sections under the polarising microscope shows an apple-green birefringence.

transferrin type. The symptoms of amyloidosis depend on the organ that is most predominantly involved. The organs that are most commonly involved are the kidneys and heart.

Renal amyloidosis usually manifests with proteinuria, often in nephrotic range⁽³⁾ and is associated with profound

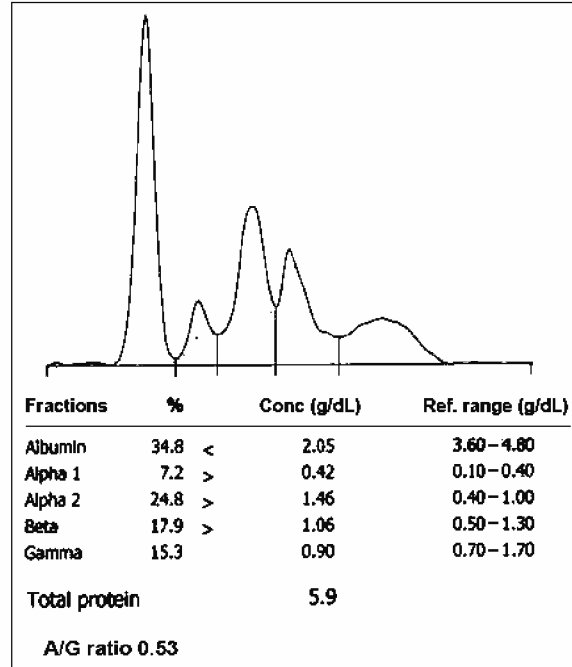


Fig. 5 Electrophoretic pattern shows decreased gamma globulins.

oedema and hypoalbuminaemia. The serum creatinine is usually normal in these patients. Renal dysfunction is the predominant manifestation of secondary amyloidosis. Glynn et al reported a diffuse involvement of the retroperitoneum by amyloid, mimicking retroperitoneal fibrosis.⁽⁴⁾ Renal cancer is the most common solid organ malignancy with amyloidosis, with as many as 3% of patients having autopsy evidence of amyloidosis.⁽⁵⁾ There have only been very few reported cases of retroperitoneal involvement of amyloidosis, either with renal cell carcinoma or with pancreatic malignancy.⁽⁶⁾ There have been a few reports of localised amyloidosis in the urinary tract, viz. the ureter and bladder. In the bladder, these resemble malignancy, and only histology can differentiate it from amyloidosis.^(7,8)

Our patient did not have any chronic inflammatory diseases, such as rheumatoid arthritis, tuberculosis or chronic osteomyelitis, that could have predisposed him to secondary amyloidosis. The initial presentation was very much like idiopathic retroperitoneal fibrosis. However, the retroperitoneal appearance of the plaque on MR imaging was not classical of retroperitoneal fibrosis, which normally begins around the L3–L4 vertebral bodies. Moreover, there was an absence of discrete retroperitoneal lymphadenopathy, which again pointed against an overt lymphoproliferative disorder. The diagnosis of amyloid was confirmed with Congo red staining. The treatment of primary amyloidosis is largely steroid-based with melphalan, and with autologous stem cell transplantation in a select group of non-responsive patients.⁽⁹⁾

In summary, localised retroperitoneal amyloidosis in the pelvis mimicking retroperitoneal fibrosis as a cause of obstructive uropathy, is a rare disease. To the best of our knowledge, this is the first case to be reported in the medical literature. This report stresses the need for an increased awareness of this rare entity among practising urologists. Prompt and appropriate treatment can then be instituted for the disease, which otherwise has a dismal prognosis.

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