

## Case Report

# An unusual presentation of atrial myxoma with haematuria and proteinuria

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### Abstract

Myxomas are uncommon primary cardiac tumours, usually affecting the left atrium. We describe an unusual presentation of cardiac myxoma with asymptomatic proteinuria and haematuria. Surgical excision of the tumour resulted in complete resolution of the urinary abnormalities. The production of antiendothelial cell antibodies and interleukin-6 by cardiac myxomas may be relevant as these substances have been implicated with the development of renal injury and proteinuria.

**Keywords:** atrial myxoma; interleukin-6; proteinuria

### Background

Cardiac myxomas are the most common benign cardiac tumour usually found in the left atrium [1]. They usually present with at least one of the classical triad of obstructive cardiac, embolic and constitutional symptoms [1]. Due to their position in the left atrium, the most common presentation is with mitral valve obstruction and cardiac symptoms, such as dizziness, dyspnoea that may improve on recumbency, cough, pulmonary oedema and congestive cardiac failure [1]. Myxomas are also commonly associated with embolization of arteries in the central nervous system, kidney and extremities [1]. Constitutional symptoms such as fever, anorexia, weight loss, malaise, myalgia and arthralgia are also frequent [1]. We describe an unusual presentation of atrial myxoma with microscopic haematuria and significant non-nephrotic-range proteinuria.

### Case report

A 54-year-old man was found to have microscopic haematuria and proteinuria on two consecutive urine tests as part of routine surveillance for hypertension. The patient had past medical history of essential hypertension and gout. His regular medications were amlodipine 5 mg once daily and allopurinol 100 mg once daily. On his first nephrological assessment, he was well with no specific complaints. Blood pressure was found to be 160/76 mmHg and

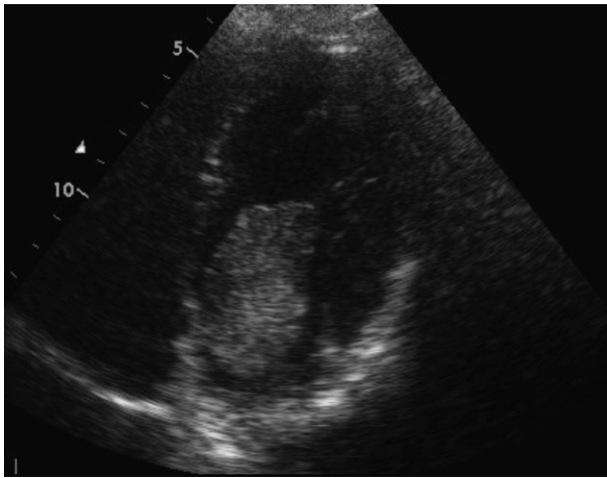
physical examination was normal. Urinalysis confirmed microscopic haematuria with 25 erythrocytes per millilitre (Siemens Diagnostics Clinitek Multistix). Urinary protein-to-creatinine ratio (PCR) at referral and on repeat testing in clinic were elevated at 127.3 and 104.8 mg/mmol, respectively. Serum creatinine was 114  $\mu\text{mol/L}$  and the estimated glomerular filtration rate was 69 mL/min/1.73 m<sup>2</sup> using the Modification of Diet in Renal Disease equation. An electrocardiogram showed voltage criteria for left ventricular hypertrophy and an echocardiogram was therefore arranged to assess end organ damage caused by hypertension. Echocardiography revealed a mobile mass in the left atrium, measuring  $\sim 5 \times 2$  cm in size and attached to the atrial septum (Figure 1). The mass prolapsed through the mitral valve during systole but did not cause any significant haemodynamic compromise.

The patient was admitted as an emergency and transferred to a local cardiothoracic centre, where a gelatinous mass was removed during an uneventful operation and shown to be a benign myxoma with small groups of basophilic tumour cells in a stroma of loose, partially haemorrhagic connective tissue.

Follow-up after the operation showed resolution of the proteinuria that prompted the original referral. On repeat testing 4 months after resection, urinary PCR was found to be 18.4 mg/mmol.

### Discussion

The presentation of atrial myxoma with proteinuria and haematuria in the absence of any other clinical signs or symptoms is very unusual. Surgical excision of the tumour resulted in complete resolution of the haematuria and proteinuria and therefore a renal biopsy was not justified. One previous case report by Kalra *et al.* [2], also described a patient with myxoma presenting with right heart failure and nephrotic range proteinuria with no evidence of intrinsic renal pathology. As in our case, surgical removal of the myxoma led to resolution of the proteinuria, suggesting that the myxoma may be responsible for such urinary abnormalities. Other recognized causes of proteinuria in patients with cardiac myxomas include membranous



**Fig. 1.** Parasternal long axis (four chamber) view of echocardiogram, showing a mobile mass in the left atrium attached to the atrial septum.

nephropathy [3], amyloidosis [4], interstitial nephritis [5] and cryoglobulinaemia [6].

Patients with myxomas have been shown to have detectable antiendothelial cell antibodies (AECA) in peripheral circulation [7]. This heterogeneous group of antibodies has been found in various vasculitic and connective tissue conditions and has a variety of actions, including endothelial cell activation, apoptosis and cytotoxicity [8]. It is presumed that myxomas cause local activation of the immune system, allowing the production of such autoantibodies by bystander activation. In animal models, the development of AECA caused renal injury and proteinuria [8].

Cardiac myxomas also secrete interleukin-6 (IL-6) [9], which may have a role in the development of renal injury and proteinuria. IL-6 is a pro-inflammatory cytokine, which causes fever, induces the production of acute phase proteins in hepatocytes and promotes differentiation of B cells, as well as activating macrophages, T cells and B cells. Population-based studies in type 1 diabetes have correlated elevated IL-6 levels with the development of gross proteinuria [10]. In addition, using a mouse model of lupus nephritis, Cash *et al.* [11] have shown that IL-6 deficiency protects against the development of renal disease.

The overproduction of IL-6 by myxomas is also responsible for the constitutional symptoms that are present in 90% of patients. Common laboratory findings in patients with myxomas include normocytic anaemia, leucocytosis, thrombocytopenia, low complements, elevated erythrocyte sedimentation rate, acute phase reactants and immunoglobulins [12]. In addition, patients may have Raynaud's phenomenon and skin rashes, such as erythematous or livedoid macules and papules or telangiectasia [12]. As a result of these abnormalities and the prolonged multisystemic symptoms, cardiac myxomas could imitate connective

tissue disease, such as systemic lupus erythematosus and vasculitis such as polyarteritis nodosa [1]. Secondary infection of myxomas may also cause similar symptoms [13].

Myxomas, although uncommon, are an important consideration in the differential diagnosis of many renal conditions. These include vasculitis, amyloidosis, renal infarction and asymptomatic urinary abnormalities. These symptoms and signs may improve with removal of the tumour. The production of AECA and IL-6 by cardiac myxomas may be relevant as these substances have been implicated with the development of renal injury and proteinuria.

#### Acknowledgements.

Author's contributions. J.B. wrote the original manuscript. C.W. physician responsible for care of patient. L.K. revised original manuscript.

*Conflict of interest statement.* None declared.

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*Received for publication: 19.10.10; Accepted in revised form: 23.11.10*