

# Primary amyloidosis of urinary bladder

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## SUMMARY

**Six cases of primary amyloidosis of the urinary bladder are reported. This is a rare clinical entity and is of interest to the urologist because it is usually confused with carcinoma. Conservative treatment is usually adequate, though long-term follow up is recommended.**

## INTRODUCTION

Primary localized amyloidosis of the urinary bladder is a rare entity. Seventy-two cases have been reported in the English literature<sup>1</sup>. This condition is of clinical interest because, both in symptoms and gross appearance, it resembles an infiltrating neoplasm. Accurate diagnosis depends on biopsy of the bladder lesion. This usually responds to conservative management, i.e., observation or transurethral resection. A series of six cases is described together with their follow-up data.

## PATIENTS AND METHODS

This study is based on clinico-pathological analysis of six patients (five men and one woman) with primary amyloidosis of urinary bladder seen at St Mary's Hospital (Portsmouth, UK) from 1966 to 1993. The age of the patients at presentation was 53 years to 77 years (mean 67 years).

The diagnosis in all the six cases was made histologically from the tissue obtained by biopsy of the bladder lesion. These tissues were routinely formalin fixed, paraffin embedded and stained with haematoxylin and eosin. Special stains, e.g. Congo red were used once the diagnosis of amyloid was suspected and was confirmed using polarized light.

## RESULTS

The clinical findings are summarized in Table 1. Four patients presented with haematuria and two had abdominal

pain associated with it. Intravenous urography in all cases was normal. Cystoscopy in two cases revealed elevated areas and in one case lesion appeared as a large papilliferous growth. The suspected diagnosis on cystoscopy in all cases was neoplasm.

Five cases underwent biopsy of the lesion. Transurethral resection of the papilliferous growth was performed in one case. Light microscopy of the tissue submitted revealed typical features of localized amyloidosis<sup>2</sup>. These showed extracellular aggregates of homogenous eosinophilic material, predominantly in the lamina propria and inner half of the muscle. The deposits gave a red reaction with congo red and showed apple-green birefringence with cross polarized light. Minor deposits were present in vessel walls and there was a focal foreign body giant cell reaction to scanty granular deposit of amyloid.

One patient died of cancer of stomach 9 years after diagnosis of amyloidosis. Another patient is 3 years post diagnosis and well, and one has been diagnosed 3 months ago and is asymptomatic. Three of the cases were lost to follow-up.

## DISCUSSION

The aetiology of amyloidosis is unknown<sup>3</sup>. Immunomechanisms are considered to play a certain role in the production of experimental amyloidosis.

In the cases of bladder amyloidosis a pathogenetic mechanism can be postulated<sup>4</sup>. Chronic and recurrent mucosal and submucosal inflammation leading to chronic cystitis causes an influx of lymphoplasmacellular elements, one of which becomes monoclonal. The monoclonal proliferates and secretes an aberrant type of light polypeptide chain which is amyloidogenic<sup>5</sup>. By means of lysosomal proteolysis of phagocytic cells or other

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Table 1 Clinical summary

Case	Age/sex	Previous GU history	Presenting complaint	Cystoscopy	Diagnosis	Treatment	Follow-up
1	73/M	N/A	N/A	N/A	Neoplasm	Biopsy	—
2	66/M	N/A	PH	Submucosal elevations	? Bilharzia	Biopsy	—
3	77/F	Nil	PH	Nodular area LUO	? Neoplasm	Biopsy	Persistent amyloidosis at 6 & 7 years. Died at 9 years. Ca stom.
4	67/M	Mumps orchitis Prostatitis Epididymitis	Haematuria Abdominal pain	Red patch, right upper quadrant	? Neoplasm ? Inflammation	Biopsy	3 years, well
5	65/M	Nil	N/A	N/A	? Neoplasm	Biopsy	N/A
6	53/M	Haematuria	Haematuria S/P pain	Large papilliferous growth-bladder base	? Neoplasm	Resection and fulguration	Diagnosed 2/12 ago

GU=Genitourinary; PH=painless haematuria; LUO=left ureteral orifice

physiochemical means, these light chains are formed into amyloid fibrils and deposited as aggregates in the bladder tissue.

The cases described here showed typical clinical and gross features of localized amyloidosis of bladder with lesions often resembling neoplasm. All patients were managed on conservative treatment. Those who could be followed up were asymptomatic.

Some authors have used intravesical instillation of dimethylsulphoxide to prevent recurrent haematuria successfully<sup>6</sup>. Oral colchicine has avoided the need for cystectomy in another patient with uncontrolled haematuria<sup>7</sup>. Some patients even required partial or total cystectomy with ileal loop diversion<sup>8</sup>.

If the inconvenience to the patient is insignificant, conservative therapy is preferable and majority of the patients can be managed in this way. However, long-term follow up is recommended.

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