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PRIMARY RENAL AMYLOIDOSIS: A CASE REPORT

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Abstract: Primary amyloidosis is the most common form of systemic amyloidosis. The morbidity arises from extracellular deposition of immunoglobulin light chain fibrils in some organs such as the kidneys, heart and bowel. Distinctive haematological and biochemical laboratory findings may help in early diagnosis. Here we present a 60-year-old lady with an generalized amyloidosis. On microscopy sections show small fragments of structure less material was seen giving orange-red appearance, when stained with Congo Red. Our comprehensive overview of this rare and often fatal disease aims to increase the awareness of AL type amyloidosis. This may facilitate earlier diagnosis and thus allow initiation of prompt and specific therapies, which are indispensable in order to improve disease prognosis.

Keywords: amyloidoses, AL amyloidosis, amyloidosis, Congo red

INTRODUCTION:

The amyloidoses are a group of disorders in which soluble proteins aggregate and deposit extracellularly in tissues as insoluble fibrils, causing progressive organ dysfunction[1,2]. Clinically evident renal involvement mainly occurs in AL or AA amyloidosis[3]. Renal involvement can also occur in some hereditary forms of amyloidosis, most commonly in AFib, AApoAI, AApoAII, ALys, and AGel, but it is rare in ATTR[4]. Amyloid fibrils can be identified in biopsy specimens both by their characteristic appearance on electron microscopy and by their ability to bind Congo red (leading to apple-green birefringence under polarized light) and thioflavine T (producing an intense yellow-green fluorescence)[5,6].

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CASE REPORT:

A female of 60 years of age presented to our hospital with mild anemia, facial oedema and mild azotaemia. She had prior history of an infection of the urinary tract which responded to antibiotics. A plain KUB x-ray film of the abdomen was performed. It showed a stag horn calculus in left kidney, overlying the middle and lower calyces with smooth edges. The excretion urography showed only faint excretion of contrast around the density. Both kidneys were approximately 12 cm long and its outline appeared smooth with no

loss of cortex. A follow up scan was done after one year. It revealed left kidney perinephritis and intrarenal pelvis. The left kidney was then opened and showed dilatation of the pelvicalyceal system and thickening of the pelvis extending into the pelvicalyceal junction. Renal biopsy was performed and the tissues were sent to pathology department for histopathological evaluation. On microscopy sections show small fragments of structure less material was

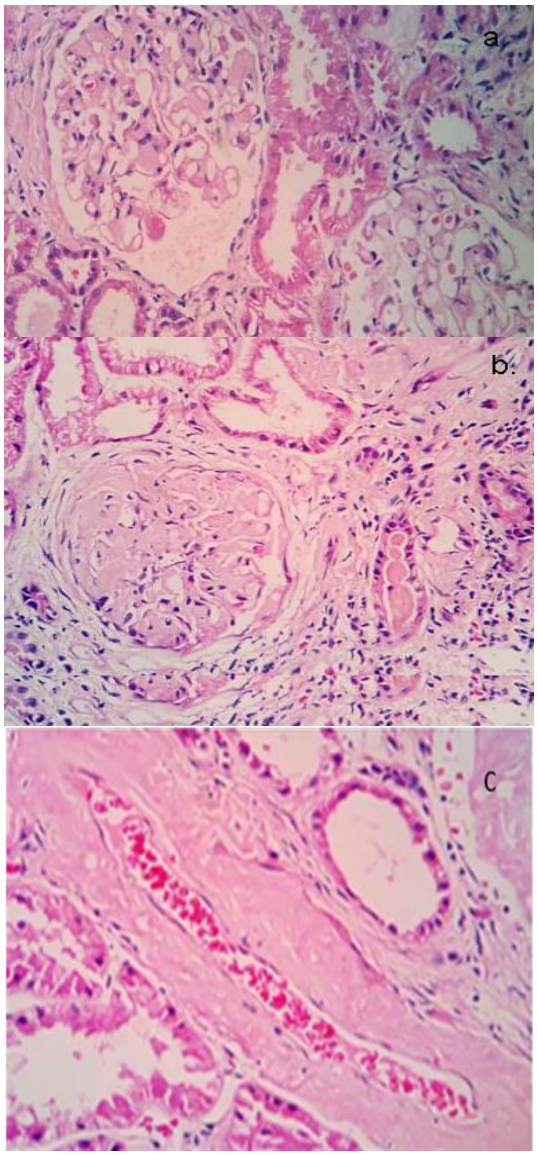

Fig.a,b Microphotograph showing glomeruli and thickend tubules showing deposition of amyloid in the basement membrane(H&E;40x).
Fig.c Microphotograph showing thickend tubules showing deposition of amyloid in the basement membrane(Congo red;40x)

seen giving orange-red appearance, when stained with Congo Red. (Fig. a,b,c) This confirmed the material deposited to be amyloid. The final diagnosis of renal amyloidosis was thus rendered. The patient was stable with regular haemodialysis for two years. Then she developed severe renal failure resulting in her death.

DISCUSSION:

Amyloidosis is an extremely rare entity that is difficult to diagnose. This is because of the unspecific early clinical manifestations of the disease. The hypothesis of amyloidosis is only considered when specific organ failure occurs. The average age at diagnosis is 64 years, and this condition occurs mainly in men. This is in contrast to our patient who is 60 years female. One of the first clinical manifestations of amyloidosis is macroglossia, which only occurs in approximately 10% of all cases. It is considered to be a pathognomonic sign of the disease. Our patient did not show macroglossia. The kidney is the organ most commonly involved in systemic AL amyloidosis which occurs in approximately 10–15% of patients with multiple myeloma. The vast majority of these patients present with nephrotic-range proteinuria and a glomerular predominance of renal amyloid deposits consisting of randomly oriented 8–10 nm thick fibrils which stain positive for Congo Red and may demonstrate classically described apple-green birefringence or show anomalous colours under a polarised light source.[8] Renal insufficiency typically occurs later. Our patient developed renal failure after two years. In the largest clinicopathologic series of 407 renal amyloidosis patients including the AL type, Said et al.[6] reported 97% had glomerular deposits, 56% had arteriolar and arterial deposits, 58% had interstitial deposits, and only 8% had tubular basement membrane deposits.[9] In the present study renal amyloid deposits were seen in the basement membrane.

CONCLUSION:

The reporting pathologist while examining renal biopsies should keep the possibility of amyloidosis in mind even in the absence of other clinical-manifestation.

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CONFLICT OF INTEREST:

Authors declared no conflict of interest