


Resumen.- OBJETIVO: La amiloidosis es una enfermedad caracterizada por el depósito de material hialino eosinófilo en distintos tejidos, siendo muy infrecuente la afectación vesical. Nuestra objetivo es dar a conocer un nuevo caso de amiloidosis vesical primaria de tipo AA y una revisión de la literatura al respecto.

MÉTODO: Varón de 66 años de edad que acude a consulta por hematuria intermitente de dos semanas de evolución, junto con urgencia y nicturia de 10-12 veces. La exploración física abdominal y genital resultó anodina así como el sedimento, urocultivo y citologías (microhematuria y fondo vesical, una mucosa con lesiones eritematosas- e inflamación). En cistoscopia se apreció, a nivel de trigono y fondo vesical, una mucosa de aspecto amiloide. Se completó el tratamiento con resección transuretral de la lesión vesical confirmando el diagnóstico de amiloidosis vesical tipo AA (propio de formas secundarias). El estudio de probable afectación sistémica fue normal.

RESULTADOS: Tras dos años de seguimiento el paciente se encuentra asintomático, sin evidencia de recidiva en las cistoscopias.

CONCLUSIONES: Las formas de amiloidosis vesical primaria de tipo AA son una patología muy infrecuente, con pocos casos descritos en la literatura urológica internacional. No obstante debemos tenerla en cuenta en el diagnóstico diferencial ante un paciente con hematuria y/o sintomatología urinaria persistente.

INTRODUCCIÓN

Amyloidosis is a disease characterised by deposition of eosinophilic hyaline material in different tissues. Urinary bladder involvement is uncommon with less than 200 cases of the primary form published in the literature. We present a new case of primary AA type amyloidosis of the urinary bladder (typical of secondary forms).

CASE REPORT

A 66-year-old male was seen in outpatient urology consultation with complaints of several weeks of intermittent haematuria with decreased urine calibre. In addition, he had intense nocturia 10-12 times per evening and occasional urgency. Physical examination of the abdomen and genitals was unremarkable. Urine sediment and blood tests were normal. Urine cytology studies were requested and revealed urothelial cells with no atypical cells and a moderate quantity of neutrophils and erythrocytes. Cystoscopy was performed and revealed yellowish erythematous lesions at the level of the vesicoureteric junction and the fundus. They were slightly elevated with green birefringence on polarised light. TUR of the bladder was later performed with the goal of completely resecting the lesion. The result of the pathology studies confirmed the biopsy findings and immunohistochemistry studies revealed AA type amyloid (typical of secondary forms).
antibodies and rheumatoid factor in search of a possible origin of the amyloidosis. All of the studies were normal. We can confirm that we are faced with a very uncommon case of AA type amyloidosis with exclusive manifestation, so far, of the bladder. DMSO instillation was not performed.

Two years after the intervention, the patient remains asymptomatic with normal endoscopic follow-up studies.

DISCUSSION

Amyloidosis is a disease characterised by extracellular deposition of eosinophilic hyaline (amyloid) material in different tissues. It can occur in any part of the urinary tract and has been described at the level of the kidney, renal pelvis, ureters, seminal vesicle, bladder as well as in the penis causing sexual dysfunction.

Two distinct types can be classified as: primary or idiopathic amyloidosis (AL), secondary amyloidosis (AA) and other forms such as hereditary forms, those associated with multiple myeloma, senile forms and focal forms which affect different organs by simulating tumours. Bladder involvement is uncommon, with approximately 160 cases of the primary form published in the literature (1) and around 30 cases of the secondary form. The latter are generally associated with rheumatoid arthritis (2,3,4).

There are histopathological differences depending on the type of amyloidosis. The primary forms predominantly affect the stroma and lamina propria which provokes a foreign-body reaction of giant cells towards the amyloid deposits. The secondary forms primarily affect the vascular and perivascular regions in the bladder submucosal which impedes adequate haemostatic vasoconstriction which may cause severe haematuria. Urinary irritation symptoms can also appear. Less frequently there is suprapubic pain, anterior pelvic mass and loss of renal function (5).

Cystoscopy findings can vary, with the appearance of haemorrhagic papillary findings with yellowish plaques in the submucosa (5). The diagnosis is confirmed with congo red stain and visualisation of the sample with polarised light. At the same time, the importance of immunohistochemistry technique in the diagnosis is due to its ability to distinguish between AL and AA type amyloid forms. Our case is peculiar in that it presented as primary AA type amyloidosis of the bladder, which is typical of secondary forms, making it necessary to rule-out the
presence of the disease in other areas. There are few cases with similar characteristics described in the literature (7,8,9).

The literature states that when amyloidosis of the bladder is primary, it can slowly evolve with progressive involvement of the entire bladder wall, causing a sensory neuropathy that behaves like a neurogenic bladder in the late stages with compromise of the upper urinary tract (10).

The treatment that has shown the best efficacy is transurethral resection of the lesion with or without bladder instillation of dimethyl sulfoxide (DMSO) in order to eliminate residual amyloid deposits (11) (50 ml instillations at 50% for 30 minutes repeated every 15 days). Rarely does massive haemorrhage require more aggressive treatment.

Close monitoring of these patients is necessary due to the possibility of disease recurrence.

CONCLUSIONS

Primary AA type amyloidosis of the bladder is a very uncommon pathology with few cases reported in the international urology literature. Nevertheless, we must keep it in mind in the differential diagnosis when faced with a patient with haematuria and/or persistent urinary symptoms.

REFERENCES AND RECOMMENDED READINGS

(*of special interest, **of outstanding interest)


