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Case Report

Xanthogranulomatous Pyelonephritis and Amyloidosis

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Keywords

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Abstract

The association of systemic amyloidosis and xanthogranulomatous pyelonephritis is extremely uncommon with fourteen cases described in the literature. We present the case of a 68-year-old woman who underwent nephrectomy for an inflammatory mass in her non-functioning left kidney. Histopathological examination of the specimen revealed, in addition to a xanthogranulomatous pyelonephritis, deposits of amyloid in the interstitial blood vessels and in the perirenal adipose tissue. Our purpose is to describe this case, given the unusual association of these entities.

ABBREVIATIONS

XGP: Xantho Granulomatous Pyelonephritis; **AA:** Amyloid A; **SAA:** Serum Amyloid A; **CT:** Computerized axial Tomography

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGP) is the inflammatory sequel of chronic supurative renal infections and usually develops in an obstructed kidney in which portions of the renal parenchyma are transformed into a xanthomatous and suppurative inflammatory mass [1]. It has a common association with *Proteus* or *Eschericia coli* infection, although *Pseudomonas* species have also been implicated [2]. In the other hand, Amyloid A (AA) amyloidosis is probably the most common type of amyloidosis worldwide and its deposits are composed mainly of the serum amyloid A (SAA) protein (an apolipoprotein of high density that serves as a dynamic acute phase reactant) [3]. It may occur in either sporadic or familial settings, developing in association with an enhanced and prolonged inflammation that leads to a sustained upregulated production of SAA and, subsequently, to incomplete degradation, misfolding, and deposition in the tissues [4]. Both of these entities are relatively frequent pathologies with innumerable cases being reported, however, their association is extremely rare [2].

CASE PRESENTATION

A 68-year-old woman presented with abdominal pain, weight loss and anemia. On computerized axial tomography (CT) the left kidney was enlarged with pyelocalyceal dilation and calcifications. It was consistent with an inflammatory process which extended to the next adipose tissue and affected the wall of the sigmoid colon. Gross pathological examination revealed a 9,5x6x5 cm kidney, weighing 396g. The renal pyramids were filled with a purulent material and one large staghorn calculi. Several intrarenal abscesses were present. The perirenal fat was involved (Figure 1). Light microscopy revealed extensive xanthogranulomatous inflammation with abundant foamy macrophages, plasma cells and neutrophils (Figure 2). The residual renal parenchyma had some sclerotic glomeruli, atrophic tubules and interstitial fibrosis. The interstitial blood vessels walls and the perinephric fat showed deposits of a homogeneous eosinophilic material suggestive of amyloid, which was Congo red positive (Figure 3). Evaluation of the stained slides under polarized light revealed greenish birefringence and immunohistochemically it was A-amyloid positive. Also, the Congo red-stained slides were examined under fluorescent microscopy and the amyloid deposits showed red fluorescence. Finally, a diagnosis of xanthogranulomatous pyelonephritis with AA amyloidosis was made.After further review of the patient's clinical history, she had any symptoms suggestive of amyloidosis in any other location.

DISCUSSION

XGP is a severe, chronic renal parenchymal infection

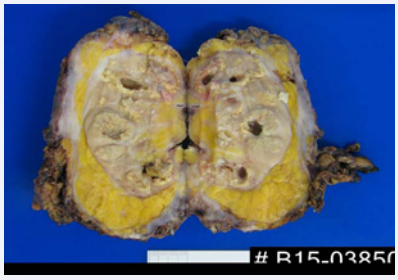

Figure 1 Grossly, the renal pyramids were filled with a purulent material and one large staghorn calculi.

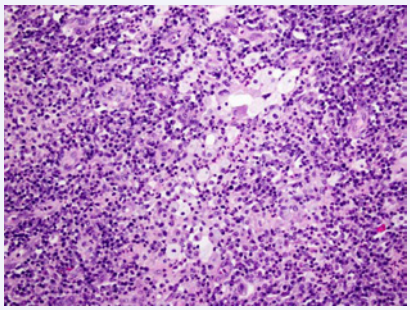

Figure 2 The renal parenchyma was extensively replaced by xanthogranulomatous inflammation with abundant foamy macrophages, plasma cells and neutrophils (H-E 20X).

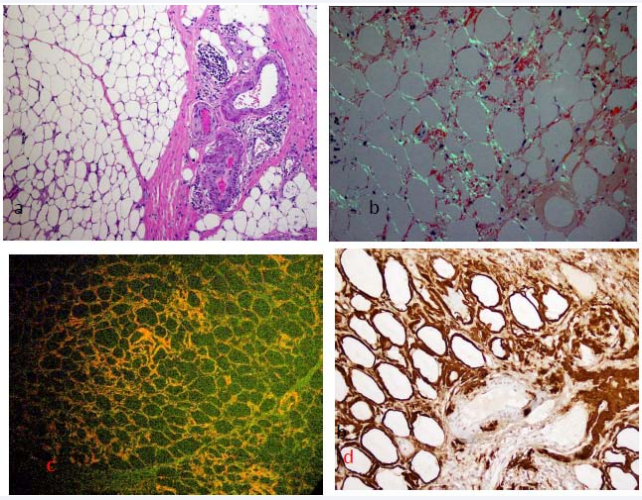

Figure 3 a: the blood vessels walls and the perinephric fat showed deposits of a homogeneous eosinophilic material suggestive of amyloid (HE 10x); b: the Congo red stain was positive showing under polarized light greenish birefringence; c: the Congo red-stained slides were examined under fluorescent microscopy and the amyloid deposits showed red fluorescence; d: Immunohistochemically it was A-amyloid positive.

characterized by renal suppuration and collecting system obstruction. Although XGP may involve male or female patients at any age, the disease is predominantly encountered in middle-aged women. It treatment consists of nephrectomy and antibiotics [5,6]. Secondary AA amyloidosis is a relatively frequent entity in which the renal involvement is almost a rule. Its *underlying* main causes are chronic inflammatory processes, specially rheumatoid arthritis, tuberculosis, leprosy, osteomyelitis, syphilis, bronchiectasis and decubitus ulcers, among others [2,7]. It may also occur in some tumors as renal cell carcinoma and Hodgkin's lymphoma [2]. Even though XGP is indeed a chronic inflammatory process, it is not a common underlying cause of AA amyloidosis. On the contrary, the association between XGP and AA amyloidosis is exceptional. To the best of our knowledge, till date there have been fourteen cases reported in the literature. The first case was described by Querfeld et al. in 1986 [8]. Among the cases described so far, this association is more frequent in female adults (M:F ratio, 2.75:1) and spans all age groups (4-76 years). Nephrolithiasis, weight loss and abdominal pain were the most common clinical manifestations (all of them present in our patient) [2,5,7-14]. Our patient did not have any chronic inflammatory conditions or neoplastic processes known to be associated with amyloidosis. She did not have nephrotic syndrome or relevant clinical manifestations related to the amyloidosis either, but she did have mild proteinuria. In the majority of cases previously reported, there was clinical remission of amyloidosis after removal of the renal lesion [2]. We cannot comment on the further course of the renal function as our patient was lost to subsequent follow up.

Table 1:

CASE	AUTHOR	YEAR	AGE	GENDER
1	Querfeld et al	1986	8	Male
2	Garber et al	1989	61	Female
3	Lauzurica et al	1991	38	Female
4	Lauzurica et al	1991	67	Female
5	Akhtar et al	1992	44	Male
6	Noyan et al	1995	6	Male
7	Mazuecos et al	1996	51	Female
8	Işlek et al	1998	4	Male
9	Rivera et al	1998	74	Female
10	Almirall et al	2001	70	Female
11	Bilbao et al	2006	51	Female
12	Val Bernal et al	2007	76	Female
13	Val Bernal et al	2007	54	Female
14	Punia et al	2015	25	Female
15	Areán et al	2015	68	Female

In conclusion, we have presented this case in order to obtain a better recognition of XGP as an underlying cause of AA amyloidosis. We consider that, although this association of events is not frequent according to the literature, pathologists must remember to look for amyloid deposits whenever a diagnosis of XGP is made, or when persistent proteinuria develops during the follow up of XGP cases.

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