

RETROPERITONEAL FIBROSIS: A SINGLE CENTER EXPERIENCE WITH MULTIDISCIPLINARY APPROACH

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Introduction: Retroperitoneal fibrosis is a rare disease characterized by the presence of inflammation and fibrous tissue in the retroperitoneum. The etiology is unclear in two-thirds of patients, in the remaining cases it can be triggered by infections, tumors, drugs, surgery and radiotherapy. An association between IgG4-related disease and retroperitoneal fibrosis is supported by the literature of the last years.

Materials and methods: We describe our cohort of retroperitoneal fibrosis composed by 16 patients. The data were subjected to statistical analysis. We refer to statistically significant as P-value < 0,05. We also try to explore the possible link between IgG4-related disease and retroperitoneal fibrosis.

EPIDEMIOLOGY	
PATIENTS	16
MALE : FEMALE	11:05
MIDDLE AGE	59,2±10,9
HISTORY OF CANCER	18,75%
SMOKER OR EX	50%
HISTORY OF ATOPY	31,25%

Tab 1: epidemiological features of retroperitoneal fibrosis patients

COMORBIDITY	
IMPAIRED RENAL FUNCTION	14
HYPERTENSION	6
CARDIOVASCULAR DISEASE	3
AUTOIMMUNE/REUMATOLOGIC	5

Tab 2: main comorbidities detected in retroperitoneal fibrosis

Kidney involvement: At the time of the diagnosis, the mean estimated glomerular filtration rate (eGFR) was 54.2±27.6 ml/min and 14 patients had an impaired renal function. Six patients there was also hydronephrosis and worst renal function (mean eGFR 32.2±16.9), so they needed ureteral stent. After resolution of urinary dilatation there was a recovery (mean eGFR 56.9±23), resulting in serum creatinine similar to the patients who did not need stenting. Only 1 patient of these required dialysis for hyperkalemia before ureteral stent.

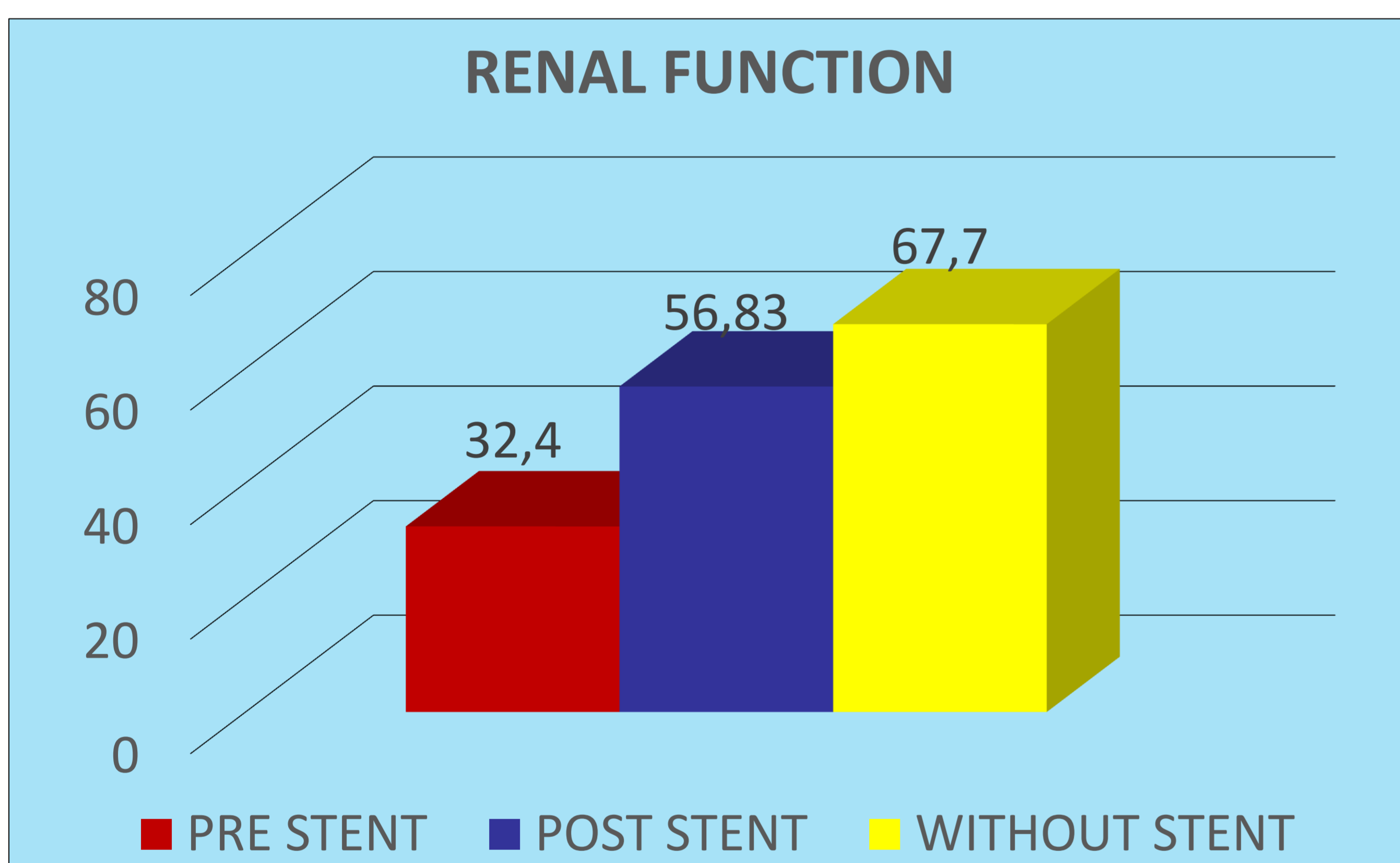


Fig 1: eGFR of patients with obstructive nephropathy (before and after stent) and not.

DIAGNOSIS	
CT	10
HISTOLOGY	3
ULTRASONOGRAPHY	2
MRI	1

Tab 3: first radiological confirmation of retroperitoneal fibrosis

Clinical features: the values of IgG4/IgG ratio were linked with direct correlation (p=0,009) with extraperitoneal manifestations. Seven patients underwent PET. There were pathologic enhancement in retroperitoneum and periaortic for all of them.

SERUM FEATURES	
LOW C3-C4	12,50%
ANA POSITIVE	43,75%
EOSINOPHILIA	6,25%
IGG4/IGG >8%	75%
IGG	1638±1000
HIGH IGG	31,25%
IGG4	391,6±585,5
HIGH IGG4	50%

Tab 4: data collected at the time of diagnosis.

EXTRAPERITONEAL	
PANCREAS	25%
SALIVARY GLANDS	6,25%
LYMPHNODES	18,75%
LIVER	31,25%
BRAIN	6,25%
EYE	12,50%
THYROID	6,25%

Tab 5: inflammatory extraperitoneal involvement.

Conclusion: almost of the patients result in impaired renal function. Kidney involvement do not seem to be linked to obstructive uropathy, because after stenting, eGFR is similar in patients with or without stent. This evidence is showed in a small number of patients, so other evaluations have to be performed to understand renal disease and physiopathology of RF.

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