Kidney involvement in patients with type 1 autoimmune pancreatitis

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IgG4-RKD is present in 27.4% of patients with AIP type 1 with male gender predominance.
In cases of early diagnosis and cortisone treatment, the clinical course is mild in most of the cases.
Regular laboratory control of renal function should be a part of the follow-up of patients with AIP type 1.

Background: Autoimmune pancreatitis (AIP) type 1 is a special form of chronic pancreatitis with a strong lymphocytic infiltration as the pathological hallmark and other organ involvement (OOI). IgG4-related kidney disease (IgG4-RKD) was first reported as a complication or an extra pancreatic manifestation of AIP in 2004. The aim of the present study was to determine the frequency and the clinical impact of kidney lesions observed in patients with AIP type 1.

Materials and method

We performed a single-centre retrospective study on a prospectively collected cohort of patients with histologically proven or highly probable diagnosis of AIP according to the ICDC classification.

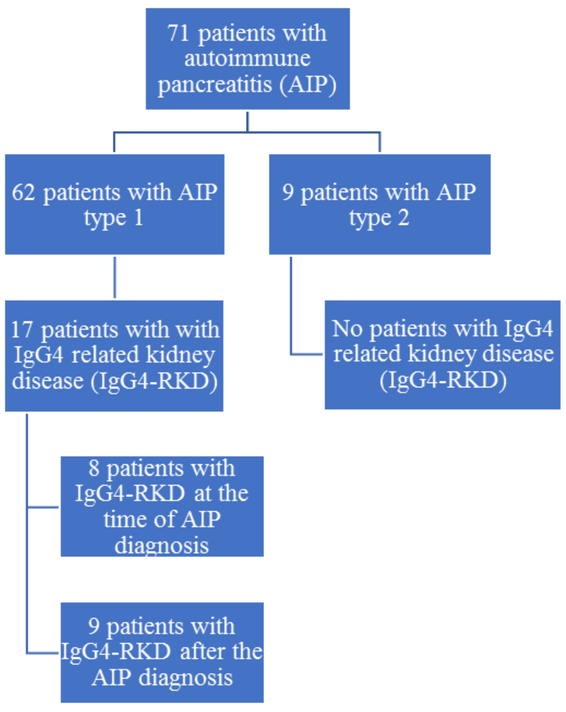

Figure: Flowchart of patients

Results: Seventy-one patients with AIP were evaluated. AIP type 1 was diagnosed in 62 (87%) patients. Kidney involvement was present in 17 (27.4%) patients with AIP type 1; 15 (88.2%) males and 2 (11.8%) females. Laboratory and/or imaging signs of kidney involvement were presented at the time of AIP diagnosis in 8 (47.1%) patients. In other patients, the onset of kidney involvement occurred between 4 months and 8 years following diagnosis. At the time of the diagnosis of kidney involvement, 8 (47.1%) patients showed elevated creatinine, and 9 (52.9%) patients showed normal serum creatinine. None of the patients were treated with dialysis.

ender	Age	Treatment	OOI (other than kidney/pancreas)	Imaging	Type of kidney involvement	Unilateral vs bilateral involvement	Onset of kidney involvement
M	74	steroids, surgery	cholangitis vasculitis (aorta) retroperitoneal fibrosis	CEMR	multiple lesions	bilateral	6 y after AIP
F	73	steroids	cholangitis Sjögren's Syndrome enlarge mediastinal LN	CECT CEMR	multiple lesions	bilateral	3 m after AIP
M	52	steroids, biliary stent	cholangitis	CECT MR w/o c	multiple lesions	bilateral	synchronous
M	49	steroids, surgery	cholangitis hepatitis enlarge abdominal LN	CECT CEMR	soft tissue in the perinephric space, diffuse swelling	bilateral	6 m after AIP
M	60	steroids	cholangitis	CEMR	multiple lesions	unilateral (left)	11 m after AIP
M	57	steroids, azathioprine	cholangitis enlarge abdominal LN vasculitis (aorta)	CECT CEMR	solitary lesion	unilateral (right)	synchronous
M	42	steroids	cholangitis	CECT CEMR	multiple lesions	bilateral	synchronous
F	39	none	cholangitis lung involvement	CECT CEMR	multiple lesions	bilateral	synchronous
M	39	none	cholangitis	CECT CEMR	multiple lesions	bilateral	synchronous
M	73	none	cholangitis	CEMR	multiple lesions	bilateral	synchronous
M	68	steroids	none	CECT CEMR	multiple lesions	bilateral	synchronous
M	68	steroids, surgery	cholangitis	CECT CEMR	focal thinning of renal cortex	bilateral	synchronous
M	85	biliary stent	cholangitis lung involvement vasculitis (aorta)	CECT CEMR	multiple lesions	bilateral	synchronous
M	71	steroids	cholangitis	CECT*	multiple lesions	bilateral	2 y after AIP
M	65	steroids	cholangitis vasculitis (aorta)	CEMR	multiple lesions	bilateral	8 y after AIP
M	52	none	cholangitis	CEMR	multiple lesions	bilateral	4 y after AIP
M	64	none	cholangitis	CEMR	solitary lesion	unilateral (right)	synchronous

Table 1: Demographic, clinical and radiological characteristics of individual patients

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