

Case report & Review article



Immunoglobulin-g4(IgG4)-rich Pseudotumor of the Kidney – A case report and review of the literature

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ABSTRACT

IgG4-related disease is a newly identified fibroinflammatory condition characterized by tumefactive lesions, a dense lymphoplasmacytic infiltrate rich in IgG4-positive plasma cells and storiform fibrosis. Elevated serum IgG4 levels are frequently observed. IgG4-related disease affects predominantly middle-aged and elderly patients, with male predominance and responds favorably to steroids. The patients present with symptoms referable to the involvement of one or more sites either simultaneously or serially, usually in the form of mass lesions affecting various organs including pancreas, biliary tree, salivary glands, periorbital tissues, kidneys, lungs, retroperitoneum and lymph nodes. The case of a 61-year old male referred for therapy of a 6.6 cm tumorous lesion of the right kidney is presented. Histological assessment of the resected mass showed IgG4-rich sclerosing pseudotumor. The available literature is summarized, and diagnostic and therapeutic options for this condition are discussed. Consideration of a non-malignant disease may be reasonable even in cases of CAT scan findings suggestive of renal cancer.

Keywords: Interstitial nephritis; Immune complex; IgG4-related disease; Membranous glomerulonephritis; Membranous nephropathy; Renal mass

INTRODUCTION

IgG4-related disease (IgG4-RD) is a new disease entity characterized by high serum IgG4 levels, infiltration of IgG4-positive plasma cells, and fibrosis of various organs, such as pancreas, biliary tract, salivary and lacrimal glands, thyroid, lung, liver, kidney, prostate, aorta, retroperitoneum, and lymph nodes. So far, the pathogenesis of this disease, including the role of high IgG4 serum levels and the presence of IgG4-positive plasma cells as well as the molecular mechanisms involved in the upregulation of IgG4 class-switch recombination remains unclear.

Recent studies reported increased proportions of type 2 helper T (Th2) cells and regulatory T (Treg) cells

and increased production of Th2 and Treg cytokines in tissues of IgG4-related disease, resulting in sclerosing pancreatitis and cholangitis, sialadenitis, and tubulointerstitial nephritis. Moreover, high Th2 cell count and overproduction of Th2 cytokines have been described in peripheral blood of IgG4-RD, as well as in Treg cells [1].

Hyper-IgG4 disease is considered a systemic autoimmune disorder with a patchy or diffuse infiltration of the organs with IgG4 positive plasma cells along with fibrosis and tends to occur in middle-aged men.

The pancreas appears the most common organ involved. Patients usually present with obstructive jaundice, a pancreatic mass simulating pancreatic head cancer or symptoms of chronic pancreatitis. Individuals with IgG4-related disease may have involvement of many different organs and even present with lymphadenopathy. Renal involvement is very uncommon and mostly characterized by the presence of multiple renal nodules [1]. Here, we report a rare case of a male patient with a large solitary IgG4 pseudotumor of the kidney. The case is discussed based upon a thorough review of the literature.

CASE REPORT

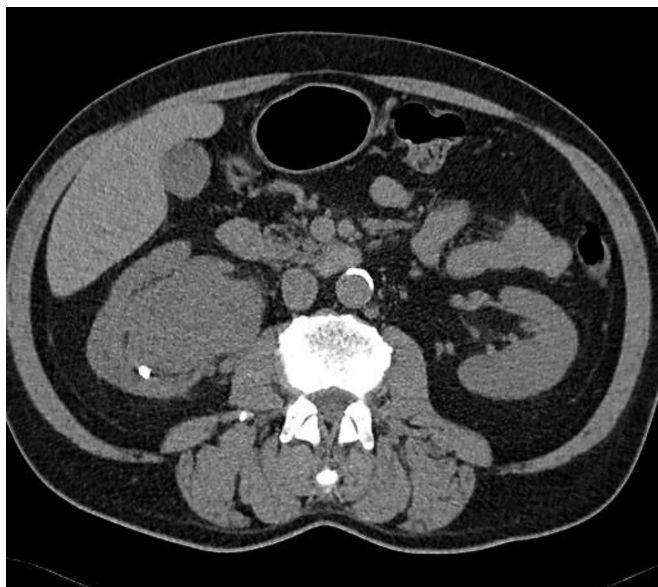
A 61-year-old obese male was seen by his family doctor with right-sided flank pain. Personal medical history comprised benign prostate enlargement (BPE), a tinnitus aurium and hyperuricemia. The patient had no history of pancreatitis. Since ultrasound revealed a large echogenous lesion in the right kidney a CAT scan was ordered (Fig. 1). CAT scan showed a 6.6 x 4.8 cm sized contrast enhancing mass in the right renal pelvis (Fig. 2). The contralateral kidney appeared normal in shape and function. Neither lymph node enlargement nor distant metastases were found, but an umbilical hernia, pancreatic lipomatosis and some diverticula of the sigmoid colon were reported. Subsequently, the patient was referred for surgical therapy. Preoperative serum creatinine was 0.56 mg/dL (normal range 0.4–0.9 mg/dL). As the diagnosis was rendered after resection of the mass, preoperative serum IgG4

level has not been measured. Based upon the diagnosis of a renal pelvic carcinoma, right side nephroureterectomy was performed. The procedure was performed without complications; the patient was discharged from hospital on day 7 after surgery. Postoperative serum creatinine was 1.1 mg/dL (calculated GFR 65 ml/min x 1.75 m²).

Fig. 1.



Fig. 2.



Four weeks later, the patient was readmitted for an incarcerated umbilical hernia. Herniotomy was performed.

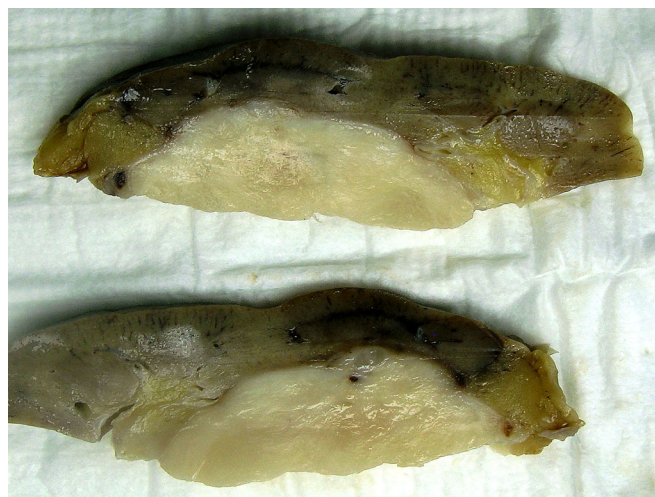
Five years after nephroureterectomy the patient is alive in good performance status without recurrence. Serum IgG4 levels are within normal range. A recent control visit yielded an asymptomatic small stone (6 mm diameter) in the left kidney.

Fig. 4. Histological features of renal IgG4-pseudotumor. **A:** the mass (right) is well demarcated from the surrounding renal tissue (left). **B:** Prominent storiform sclerosis entrapping mononuclear cell infiltrates. **C:** Predominance of plasma cells is seen at higher magnification. **D:** Very high numbers of IgG4-plasma cells (main image) with almost equal number of IgG-positive plasma cells (subimage) indicating a ratio of >0.8.

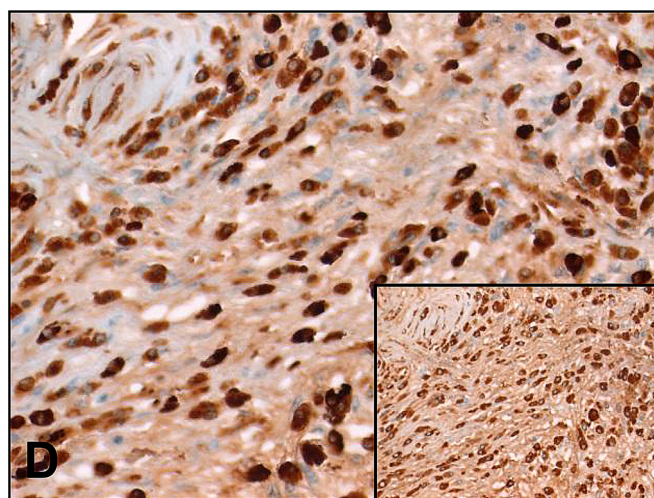
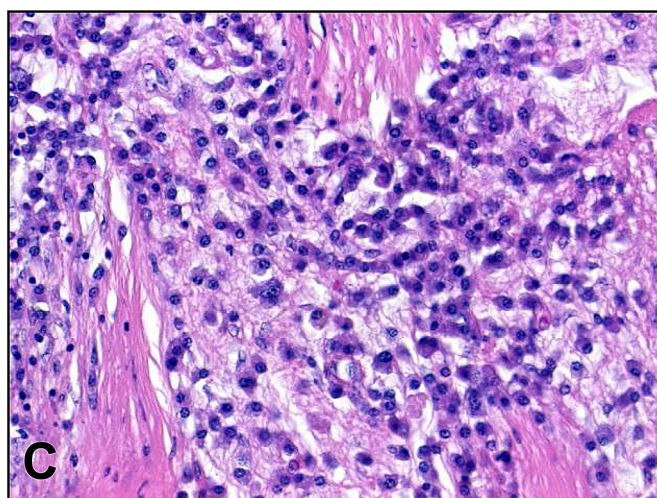
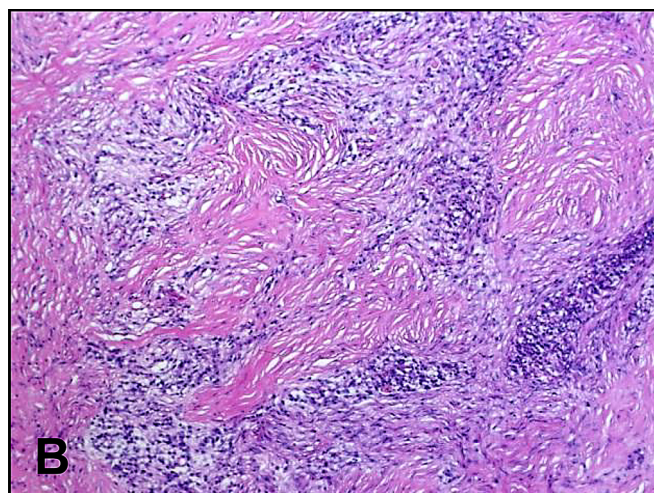
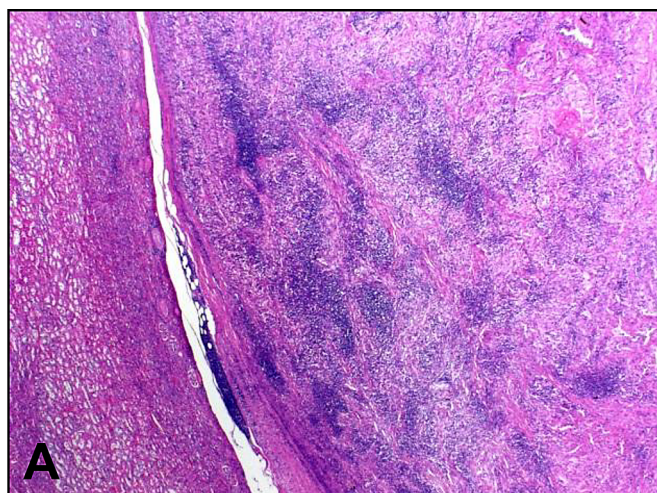
Gross and Microscopic Evaluation

Gross examination revealed a well-circumscribed but nonencapsulated firm whitish homogenous mass measuring 6.5 cm in maximum diameter. The tumor was confined to the kidney and largely protruding into the peripelvic fatty tissue (Fig. 3). Focal areas with myxoid appearance were noted, but no obvious necrosis or hemorrhage was observed.

Fig. 3. Gross specimen of renal IgG4-pseudotumor showed well circumscribed but non-encapsulated mass with homogeneous whitish cut-surface protruding into peripelvic fatty tissue.



Microscopic examination after thorough sampling revealed a well circumscribed dense lymphoplasmacytic infiltrates within highly sclerosed stromal background (Fig. 4A). The sclerosis showed distinctive storiform appearance with entrapment of inflammatory cells (Fig. 4B). At higher magnification, predominance of plasma cells was appreciated (Fig. 4C), but variable lymphoid aggregates occasionally forming follicle-like structures were seen as well. In other areas, a highly myxoid and strikingly paucicellular component was seen there was no evidence of obliterative phlebitis, necrosis, atypical stromal cells or multinucleated giant cells. Immunohistochemistry showed significantly elevated numbers of IgG4 positive plasma cells in excess of 200 cells/1 high power field with almost equal proportion of IgG4-positive and IgG-positive plasma cells (Fig. 4D) with a ratio of >0.8. All other relevant markers were negative (CDK4, MDM2, alpha smooth muscle actin, ALK1, desmin, Protein S100, pancytokeratin). No IgG4 positive plasma cells or similar lesions were present within the adjacent normal renal tissue. The morphologic and immunohistochemical findings were diagnostic of an IgG4-related fibrosclerotic inflammatory pseudotumor.



Immunological parameters:

Extensive immunological assessment was made post-operatively. Antibodies against mitochondria (AMA-M2), centromer B, histones, myositis (Jo-1), nucleosomes, Proliferating-Cell-Nuclear-Antigen (PCNA), polymyositis scleroderma antigen (PM-Scl), ribosomal-P-protein, ribonucleoprotein (RNP) 70, RNP A, RNP C, RNP/Sm, Ro-52

(52 kDa), scleroderma antigen (Scl)-70, Sm, and Sjögrens syndrome antigen (SS)-B were negative.

Positive serum levels were obtained for antinuclear antibodies (ANA) for Hep2 (1:320 (normal < 1:100) and SS-A nativ (60 kDa).

Postoperative complement and immunoglobulin levels are listed in table 1.

Table 1. Postoperative complement and immunoglobulin levels after removal of an IgG4

Parameter	Unit	Serum level	Normal range
Complement C3	mg/dl	119	79 - 152
Complement C4	mg/dl	25.3	16 - 47
IgG1	mg/l	5680	2800 - 8000
IgG2	mg/l	5760	1150 - 5700
IgG3	mg/l	855	240 - 1250
IgG4	mg/l	858	52 - 1250
IgA	mg/dl	439	82 - 453
IgE	U/ml	105	0 - 100
IgG	mg/dl	1380	751-1560
IgM	mg/dl	87.6	46 - 304

DISCUSSION

In 2003 Kamisawa et al. described eight patients with autoimmune pancreatitis associated with a lymphoplasmacytic infiltrate and fibrosis [3]. Other case series have expanded the spectrum of this disease and showed involvement of multiple organ systems in addition to the pancreas [4]. This condition has now been described in virtually every organ system: the biliary tree, salivary glands, periorbital tissues, kidneys, lungs, lymph nodes, meninges, aorta, breast, prostate, thyroid, pericardium, retroperitoneum, mediastinum and skin. Although the pancreas is the most common reported organ to be involved, many patients with IgG4-related disease without pancreatic involvement have been reported [3, 5]. IgG4-related disease may involve more than one organ, either simultaneously or sequentially. The histopathological features bear striking similarities across the involved organs. Disparate disorders such as Mikulicz's syndrome, Küttner's tumor multifocal fibrosclerosis, and eosinophilic angiocentric fibrosis are now considered to fall within this disease spectrum.

Multiple names have been proposed to describe this new disease, however a recent international consensus paper strongly recommends using the term IgG4-related disease for the overall condition and to avoid other potentially confusing nomenclature. [6, 7]. This term aptly recognizes the ubiquity of IgG4 within involved organs and the frequency of elevated serum IgG4 concentrations. The incidence is higher in older men, with a male to female ratio of 4 to 1. This disorder is more common in Asian patients than other ethnicities [8]. The typical presentation is with an incidental mass on imaging studies, symptoms related to the presence of a mass or end-organ dysfunction of the involved organ.

Immunoglobulin G4 (IgG4) is the least common of the four subclasses of immunoglobulin G (IgG), namely IgG1, IgG2, IgG3, and IgG4, normally constituting only 3% to 6% of the entire IgG fraction. The major differences among the subclasses lie in the composition and structure of the hinge region that has significant effects on the antigen binding and effector functions of the immunoglobulin molecules, resulting in different functional characteristics of each IgG subclass. For instance, IgG4, in contrast to the other IgG subclasses, does not activate complement and has only low affinity for target antigens. IgG4 is a T-helper 2 (Th2) cell-dependent isotype. Despite uncertainties about its normal function, IgG4 seems to play a significant role in allergic reactions, such as atopic eczema, bronchial asthma, and bullous skin lesions. IgG4 has not attracted the attention of pathologists and clinicians until the recent recognition of the syndrome of IgG4-related disease (IgG4-RD), a fibroinflammatory condition with a marked propensity to present as mass-forming lesions [7, 9].

The pathogenesis of IgG4-RD remains unclear, however, it has been reported that Th2 cytokines (IL-4 and IL-13) and Treg cytokine (IL-10) can induce IgG4- and IgE-specific class-switch recombination, and tumor growth factor (TGF)- β , a Treg cytokine, may induce tissue fibrosis [10].

In a series of 64 patients with IgG4-related disease, the most common extrapancreatic lesions were hilar lymphadenopathy; extra pancreatic bile duct lesions; lachrymal and salivary gland lesions and retroperitoneal fibrosis [8]. The kidneys may be involved in IgG4-related systemic disease [10, 11]. While most cases of IgG4-related disease involving the kidney have a history of prior pancreatic involvement, isolated kidney IgG4 disease is very rare [5, 12, 13]. The patients present with multiple renal nodules that may or may not be associated with impairment in renal function. On imaging studies, the most common findings are low attenuation cortical nodules, wedge shape lesions, or diffuse patchy areas [3, 11]. Most reported patients with renal IgG4-RD have been diagnosed with serologic studies showing a marked elevation of IgG4 and subsequent radiographic imaging.

Diagnosis

Prodromal characteristic features of IgG4-RD are:

- Elevation of serum IgG4 present in 60-70% of the patients
- and/or infiltration of different organ by IgG4 positive plasma cells along with tissue fibrosis
- Obliterative phlebitis.

The clinical features vary with the involved organ. The typical pancreatic IgG4 disease, for example, may present as obstructive jaundice, pancreatic mass or chronic pancreatitis.

In case reports or short series of autoimmune pancreatitis (AIP), it has been shown that pancreatic and extrapancreatic tissues involved with IgG4-RD show 18F-FDG uptake. For this reason, it has been suggested that FDG-PET/CT could be useful at diagnosis to distinguish IgG4-related pancreatitis from other pancreatic pathologies. Moreover, it has been suggested that this exploration could be useful for the evaluation of disease distribution, guiding minimally invasive tissue diagnosis to support the diagnosis of IgG4-RD, highlighting conditions for which steroid therapy is indicated, monitoring response to therapy, or detecting a relapse [14, 15].

Over the past decade, diagnostic criteria of IgG4-RD and the necessity to have tissue biopsy to confirm the diagnosis have been debated [16, 17]. Meanwhile, the International Association of Pancreatology (IAP) does no longer request tissue biopsy for the diagnosis of IgG4/type 1 autoimmune pancreatitis [2].

Core needle biopsy is becoming more common in the era of nephron preserving procedures like for control of the outcome of radio frequency ablation (RFA) and cryotherapy, or, in surveillance of the small renal masses (SRM). Needle biopsy is also considered in cases of doubtful diagnosis. Until today, there are no data available supporting the use of needle biopsy as diagnostic measure in patients suspected to suffer from IgG4-RD.

An algorithmic diagnostic approach has been suggested by Kawano et al. [18] in which IgG4 kidney disease is suspected if there is evidence of previous kidney injury (proteinuria, hematuria, and elevated N-acetyl- β -D-glucosaminidase, β 2- microglobulin, and/or α 1-micro-

globulin excretions in urinalysis) along with characteristic radiologic findings with either elevated serum IgG level, hypocomplementemia, or elevated serum IgE level.

Pathology

An international symposium on IgG4-related disease in 2011 in Boston, MA, was organized to provide practicing pathologists with a set of guidelines for the diagnosis of IgG4-related disease [19]. The experts concluded that the lesions histologically suggestive of IgG4-related disease based on these guidelines will often fulfill organ-specific criteria, in which histology is a key component. Discrepancies, if any, should be resolved in an interdisciplinary setting and with further evaluation, including a rebiopsy if necessary.

According to the Boston consensus for cases with tissue available, histologic criteria supporting the diagnosis of IgG4/type 1 autoimmune pancreatitis require at least three out of the following four conditions [20].

1. Periductal lymphoplasmacytic infiltrate
2. Obliterative phlebitis
3. Storiform fibrosis, and
4. Abundant IgG4 plasma cells (more than 10 per high-power field).

Although the combination of histopathological features and immunohistochemical stain results can provide strong supportive evidence for the diagnosis of IgG4-related disease, careful correlation with the clinical scenario and imaging characteristics of a particular patient is suggested. Thus, for the interpretation of pathology/biopsy results alone, the wording 'histologically suggestive of IgG4-related disease' is proposed [5, 14, 21]. However, this suggestion is of only limited support for a therapeutic decision.

IgG4-related kidney disease (IgG4-RKD) is the term used to refer to any pattern of renal involvement by IgG4-related disease (IgG4-RD). As with other medical kidney diseases, IgG4-RKD can be described in terms of changes to the different "compartments" in the kidney:

- the tubules
- the interstitium
- the glomeruli
- and the vessels.

In the kidney, IgG4-RD manifests most commonly as tubulointerstitial nephritis (TIN), which may be mass forming and detected on radiographic examination. Glomerular disease, in particular, membranous glomerulonephritis (MGN), may also be seen in IgG4-RD, with or without concurrent IgG4-related TIN (IgG4-TIN). A lesion of the arteries, IgG4-related plasma cell arteritis, has also been observed. The kidney may also be affected by extrarenal manifestations of IgG4-RD, including ureteral inflammatory pseudotumor or retroperitoneal fibrosis [18].

The size range reported for IgG4 pseudotumors has been variable with the largest described masses up to 8

cm in maximal dimension [11]. The main pathologic finding in IgG4-related pseudotumor of the kidney includes a patchy or diffuse tubulointerstitial nephritis associated with storiform fibrosis [12]. The infiltrating cells are mainly lymphocytes, plasma cells, and eosinophils. Immunohistochemical studies for IgG4 are critical to make the diagnosis as they highlight the significant increase in IgG4 positive plasma cells in the interstitium with sparing of the glomeruli. Glomerulopathy is uncommon in IgG4 disease [12].

The majority of patients respond to glucocorticoids, particularly in early stages of disease. In some subsets of organ disease (eg, pancreatitis), glucocorticoid responsiveness has been considered one diagnostic criterion for the disorder. Two major presentations of this condition, which often affects more than one organ, are type 1 autoimmune pancreatitis (IgG4-related pancreatitis) and salivary gland disease; the latter may present as salivary gland enlargement or as sclerosing sialadenitis (formerly termed "Mikulicz disease" and Küttner's tumor, respectively) [22].

CONCLUSIONS

Benign renal lesions with inflammatory background are rare and represent app. 1% of kidney tumors. Differential diagnostic considerations include interstitial pyelonephritis of other cause (chronic pyelonephritis related to infection), xanthogranulomatous pyelonephritis, and drug related pyelonephritis; the latter is of particular interest as it can often have prominent eosinophils. In addition, benign lesions of the renal pelvis are even less frequent.

The current treatment of choice for IgG4-related disease is corticosteroids. In the few case reports and case series of renal involvement by IgG4 disease, the renal masses have been shown to decrease in size on therapy, with some disappearing and others recurring after cessation of therapy [10, 12]. In patients presenting with impaired renal function, corticosteroids have been reported to improve renal function after four weeks of therapy and decrease in serum IgG4 levels [12].

Neither biopsy nor further laboratory tests were made in this patient before nephrectomy. Reasons for this were misleading findings in CAT scan strongly suggestive of malignant disease with the renal lesion as a solitary manifestation. In addition, biopsy may yield incorrect findings due to a sampling error the idea of confirming the diagnosis by needle biopsy was abandoned. Finally, the patient suffered from colic-like pains, which required a timely intervention. Nevertheless, it may be discussed if a correct diagnosis and subsequent corticosteroid treatment could have changed the course of disease and might have prevented nephrectomy. In consequence, consideration of a non-malignant disease may be reasonable even in cases of CAT scan findings strongly suggestive of renal cancer.

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