

## IgG4 Related Kidney Disease with Extra-Renal Involvement Demonstrated on FDG PET/CT

### Abstract

Immunoglobulin G4 (IgG4)-related kidney disease is a relatively rare clinical entity and usually occurs as an extra-pancreatic manifestation of IgG4-related autoimmune pancreatitis. We describe here the imaging findings of a patient who presented with recurrent multiorgan IgG4-related disease, involving bilateral kidneys/ureters, proximal small bowel, and multiple abdominal and extra-abdominal lymph nodes.

**Keywords:** 2-Fluoro-2-deoxy glucose, immunoglobulin G4, immunoglobulin G4-related kidney disease, inflammatory, kidney, positron emission tomography/computed tomography

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### Case History

A 52-year-old male with a history of immunoglobulin G4 (IgG4)-related biliary stricture, treated with steroids in 2017, presented with increased stool frequency and weight loss for 2 months. Blood reports revealed normal total white blood cell count with increased differential eosinophil counts - 13% (normal <6%), absolute eosinophil count - 1190 cells (normal <450 cells/cumm), normal lactate dehydrogenase - 142 IU/L (normal <450 IU/L), and elevated serum IgG4 subclass - 353 mg/dL (normal range for >18 years old: 3–201 mg/dL). Routine urine analysis does not reveal any infections/proteinuria, and urine cytology was negative malignancy. Colonoscopy-guided biopsy was suggestive of colitis. In view of known history of IgG4-related disease, a whole-body 18F-2-fluoro-2-deoxy glucose (FDG) positron emission tomography/computed tomography (PET/CT) was done which revealed FDG uptake in the diffuse thickening of the pelvis of bilateral kidneys [Figure 1a-g, yellow arrows, pelvis of left kidney -  $SUV_{max}$  - 11.32] and bilateral proximal ureter [white arrows, Figure 1e, left ureter -  $SUV_{max}$  - 4.45] with smooth and patent urinary lumen [Figure 1b-d, red arrows]. On delayed post-intravenous (IV) furosemide, delayed PET/CT images showed soft tissue density in the bilateral

pelvis without any delayed contrast retention [Figure 1f, yellow arrow heads] and with persistent FDG uptake in the bilateral pelvis/ureteric wall [Figure 1g, white arrow head]. In addition, FDG PET/CT showed increased FDG uptake in the multiple enlarged mesenteric/retroperitoneal/pelvic nodes (with surrounding mesenteric fat stranding) and mild wall thickening in the proximal small bowel and proximal large bowel [Figure 2a-c, yellow arrows]. Extra-abdominal increased FDG uptake was also noted in the bilateral tonsils and enlarged bilateral axillary and cervical nodes [Figures 1a and 2d]. In view of patient's history, current imaging, and blood investigations, diagnosis of IgG4-related kidney disease (IgG4-RKD) relapse was established without need for re-biopsy. The patient was treated steroids which subsequently showed good clinical improvement.

### Discussion

IgG4-RKD is a relatively newly recognized disease after many reports associated its occurrence in patients with autoimmune pancreatitis.<sup>[1,2]</sup> Early recognition of the disease is important as it is a condition associated with dramatic response to IV steroids.<sup>[3]</sup> Imaging features, best described on CT, are the characteristics and often the first recognized manifestation of this disease. Most common pattern reported on CT appears to be multiple rounded low-density lesions in the bilateral renal

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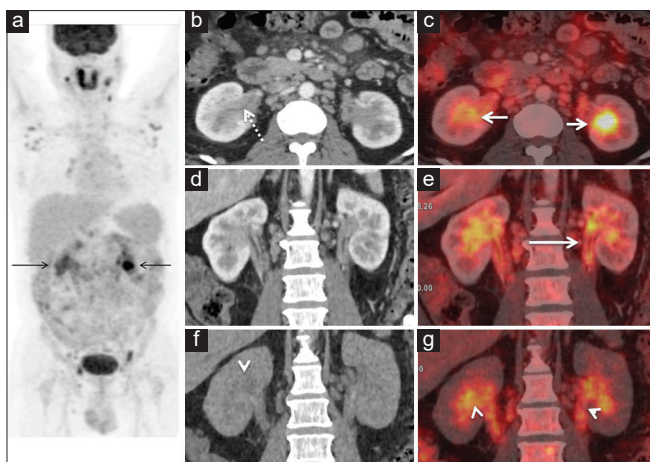
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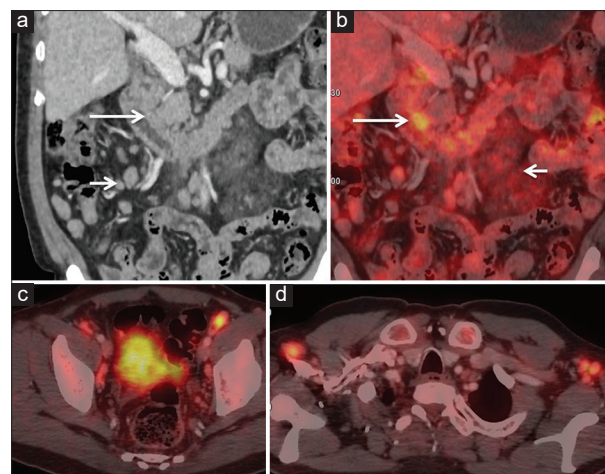
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**Figure 1:** (a) Whole-body 2-fluoro-2-deoxy glucose positron emission tomography MIP image showing increased 2-fluoro-2-deoxy glucose uptake in the bilateral kidneys (black arrows), bilateral palatine tonsils, and bilateral cervical, axillary, and pelvic nodes. (b and c) Trans-axial computed tomography and positron emission tomography/computed tomography images showing enhancing soft tissue thickening in the bilateral renal pelvis (small white arrows) with smooth intraluminal urinary lumen (dashed white arrow). (d and e) Coronal computed tomography and positron emission tomography/computed tomography images showing bilateral pelvis and proximal ureteric thickening (long white arrow). (f and g) Delayed computed tomography and positron emission tomography/computed tomography images showing no contrast retention on computed tomography and persistent accumulation of 2-fluoro-2-deoxy glucose in the bilateral renal pelvis and proximal ureters (white arrow heads) suggestive of inflammation

cortex. Other less common patterns reported are diffuse enlargement of both kidneys or diffuse thickening of the pelvis with smooth intraluminal surface (as seen in our case), or rarely, the disease may present as a solitary hypovascular kidney mass. These characteristic radiology appearances along with serological profile can help differentiate IgG4-related kidney disease with other mimicking systemic illness (such as pyelonephritis, lymphoma, sarcoidosis, vasculitis, renal malignancies, and Castleman's disease).<sup>[4,5]</sup>

IgG4-related fibroinflammatory disease may virtually involve every other organ including pancreas (most common), liver, bile ducts, stomach, salivary glands, thyroid, kidneys, and colon.<sup>[6-8]</sup> Due to the same reason, the clinical symptoms are variable/nonspecific, often leading to delay in diagnosis and thereby threatening organ function. Growing evidence over the past half-decade favors the use of PET/CT as a one-stop shop for imaging IgG4-related diseases for diagnosis, guiding biopsy, and treatment response evaluation.<sup>[9,10]</sup> Detecting extra-renal involvement at baseline appears to be the most important indication of using FDG PET/CT in IgG4-RKD.<sup>[11,12]</sup> Appropriate use of PET/CT should be guided by the serum IgG4 levels, as the probability of detecting multiorgan disease is >80% if the serum IgG4 levels are two times the upper limit of normal (i.e., >270 mg/dL, normal limit ≤135 mg/dL).<sup>[13]</sup> Another potential use of FDG PET/CT in this disease would be in avoiding invasive tissue diagnosis, especially in recurrent disease setting.



**Figure 2:** (a and b) Coronal computed tomography and positron emission tomography/computed tomography images showing 2-fluoro-2-deoxy glucose uptake in the thickened duodenal wall (long white arrows) with adjacent mesenteric fat stranding and multiple enlarged mesenteric nodes (short white arrow). (c and d) Trans-axial positron emission tomography/computed tomography images showing increased 2-fluoro-2-deoxy glucose uptake in the enlarged pelvic and axillary nodes

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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