**Background**

- Immunoglobulin G4 (IgG4)–related disease is a recently proposed clinical-pathologic entity characterized by fibroinflammatory lesions rich in IgG4-positive plasma cells and, often but not always, elevated serum IgG4 concentrations.
- Although definitive diagnosis requires histopathologic analysis, imaging plays an important role in demonstrating infiltration and enlargement of involved organs. Because IgG4-related disease usually shows a marked response to corticosteroid therapy, radiologists should be familiar with its clinical and imaging manifestations to avoid a delayed diagnosis and unnecessary surgical interventions.

**Learning objectives**

1. To illustrate imaging findings of IgG4-related sclerosing diseases affecting various abdominal organs including Liver, biliary tract, pancreas, retroperitoneum, kidneys, lymph nodes.
2. To inform the imaging features of IgG4-related diseases of abdomen which can be helpful to differentiate from many of tumorous or other inflammatory abdominal diseases.

**Imaging Findings/Procedure Details**

### IgG4-related Diseases of Abdomen and their Radiologic Mimics

<table>
<thead>
<tr>
<th>Organs</th>
<th>IgG4-related Diseases</th>
<th>Mimics</th>
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<tr>
<td>Pancreas</td>
<td>Autoimmune pancreatitis (AIP)</td>
<td>Focal form AIP - pancreatic malignancy</td>
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<td></td>
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<td>Diffuse form AIP - pancreatitis</td>
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<td>Liver</td>
<td>IgG4-related inflammatory pseudotumor, Hepatopathy</td>
<td>Malignant hepatic tumors</td>
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<td>Bilary tree</td>
<td>IgG4-related sclerosing cholangitis</td>
<td>Primary sclerosing cholangitis, cholangiocarcinoma</td>
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<tr>
<td>Kidney</td>
<td>Tubulointerstitial nephritis, Membranous glomerulonephritis, Inflammatory pseudotumors, chronic sclerosing pyelitis</td>
<td>Bilateral - lymphoma, metastases, Unilateral- RCC, transitional cell carcinoma, Wedge shape - infarct, pyelonephritis</td>
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<tr>
<td>Small bowel, mesentery</td>
<td>IgG4-related sclerosing mesenteritis</td>
<td>Lymphoma, carcinoid tumor, carcinomatosis</td>
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<td>Retropertion -eum</td>
<td>IgG4-related retroperitoneal fibrosis</td>
<td>Lymphoma, large-vessel vasculitis, syphilis, sarcoidosis-induced arthritis</td>
</tr>
<tr>
<td>Lymph nodes</td>
<td>IgG4-related Lymphadenopathy</td>
<td>Lymphoma, multicentric Castleman disease, disseminated malignancy</td>
</tr>
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</table>

### IgG4-related Autoimmune Pancreatitis

Specific findings: Capsulelike rim or halo of low attenuation surrounding pancreas, presumed to represent a fluid collection, a phlegmon, or fibrosis. Imaging: Hypoechoic at US, hypoattenuating at CT, midly hyperintense at T2-weighted MR imaging with distinctive delayed contrast enhancement.

- **Diffuse diseases:** a uniformly enlarged pancreas with sharp margin and loss of lobular contours, resulting in a featureless sausage like appearance
- **Focal disease:** focal enlargement of pancreas, resulting in a mass like appearance

### IgG4-related Autoimmune Pancreatitis

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### IgG4-related Sclerosing Cholangitis

Most common involved segment - the intrapancreatic portion of the CBD. Focal or diffuse bile duct wall thickening with stenosis and upstream dilatation.

A circular and symmetric rim of tissue encasing the bile duct wall with relatively smooth margins and homogenous enhancement in the delayed phase of CT and MRI.

- **Differential diagnosis** are primary sclerosing cholangitis, cholangiocarcinoma

### IgG4-related Renal Involvement

Five patterns of diseases; 1) bilateral round or wedge-shaped peripheral cortical lesions(m) 2) bilateral nodules in renal sinus 3) rim of soft tissue around the kidney 4) diffuse patchy involvement 5) diffuse wall thickening of renal pelvis CT - hypoattenuating during the arterial phase, becoming isoattenuating relative to the surrounding parenchyma during late phases.

MRI - low signal intensity on both T1- and T2-weighted MRI with mild enhancement on T1WI.

### IgG4-related Hepatopathy

- Most common - focal/ bilateral nodules in hepatic parenchyma.
- Hypoattenuating during the arterial phase, becoming isodense with late enhancement.

### IgG4-related Sclerosing Cholangitis

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### IgG4-related Kidney Disease

- Multiple round or wedge-shaped cortical nodules, - pyelonephritis, vascular insult, metastases, and lymphoma.
- Solitary round lesion, - RCC, TCC

### IgG4-related Disease

IgG4-related disease is a unique and distinct systemic disease that is frequently misdiagnosed as neoplastic and other inflammatory processes. The recognition of the typical imaging patterns in each abdominal organ can raise the suspicion of the disease, and the combination of radiologic, serologic, and histologic findings can lead to correct diagnosis, ensuring effective treatment.