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## PICTORIAL ESSAY

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# Immunoglobulin G4-related Systemic Disease: Pictorial Review of the Pancreatic and Extrapaneatic Manifestations

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### ABSTRACT

*Immunoglobulin G4-related systemic disease (IgG4-RSD) is a newly recognised and emergent condition, which was initially thought to manifest only as autoimmune pancreatitis. As the knowledge about this disease grows, it is now known that it also manifests in various extrapancreatic organs. Some of the appearances can mimic neoplasms leading to unnecessary surgery and anxiety. This pictorial review presents the characteristic radiological features of IgG4-RSD in various organs with different imaging modalities. We hope to raise awareness of this new disease entity and assist the differentiation of lesions due to IgG4-RSD from other pathologies.*

*Key Words: Autoimmune diseases; Cholangitis, sclerosing; Immunoglobulin G; Pancreatitis; Retroperitoneal fibrosis*

## 中文摘要

### 免疫球蛋白G4相關的系統性疾病：胰腺和胰腺外其他組織臨床表現的圖文回顧

陳慧儀、梁凱瑩、陳澤宗

免疫球蛋白G4相關的系統性疾病（IgG4-RSD）是一種新近提出並不斷發展的疾病，這種疾病最初認為僅表現為自身免疫性胰腺炎。伴隨對這種疾病的深入了解，目前認為IgG4-RSD也會涉及胰腺外的其他器官，其中一些表徵與腫瘤很相似，因而導致了不必要的手術及焦慮。本圖文回顧展示了IgG4-RSD在不同器官中影像學的不同特徵，希望借此提高人們對這種新疾病的認識，從而協助分辨IgG4-RSD及其他疾病。

### INTRODUCTION

Immunoglobulin G4-related systemic disease (IgG4-RSD) — also known as hyper-IgG4 disease, IgG4-related sclerosing disease, and IgG4-related disease —

is a recently recognised and emergent condition. It is characterised by diffuse infiltration of IgG4-positive plasma cells and CD4-/CD8-positive T lymphocytes along with fibrosis and obliterative phlebitis leading to

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Submitted: 10 Apr 2014; Accepted: 15 Jul 2014.

organ dysfunction.<sup>1,2</sup>

It was initially thought to manifest only as autoimmune pancreatitis (AIP). However, involvement of many other extrapancreatic organs, including the biliary tree, retroperitoneum, salivary glands, lymph nodes,<sup>3</sup> kidneys,<sup>4</sup> and lungs<sup>5</sup> has been reported. IgG4-RSD usually affects middle-aged and elderly patients with a male predominance.<sup>6</sup> Over 90% of patients exhibit elevated serum IgG4 levels, which reflect increased disease activity.<sup>7</sup> It also has a favourable response to steroid therapy.<sup>8-11</sup> Unfortunately, massive infiltration of the organs by inflammatory cells can lead to tumefactive enlargement of the affected organs, which may mimic malignancy and lead to unnecessary anxiety and surgery.<sup>12,13</sup> Recognition of typical imaging findings of multiorgan involvement of the disease can help differentiate IgG4-RSD from malignancy and improve the diagnostic accuracy for IgG4-RSD.

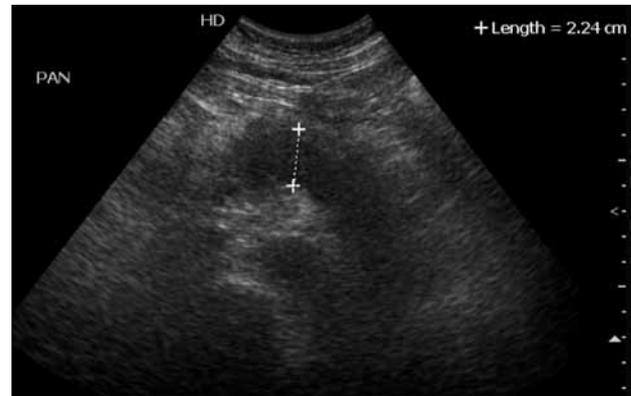
This pictorial review presents the characteristic radiological features of IgG4-RSD in various organs with different imaging modalities. We hope to raise awareness of this new disease entity and assist the differentiation of lesions due to IgG4-RSD from other pathologies.

### IMMUNOGLOBULIN G4-RELATED AUTOIMMUNE PANCREATITIS

AIP has been estimated to account for 1.95%<sup>1</sup> to 5.4%<sup>14</sup> of patients with chronic pancreatitis. It commonly manifests as obstructive jaundice with no or only mild abdominal pain, weight loss, and recent onset of diabetes (usually type II) in elderly patients.<sup>15-17</sup>

On imaging, the affected area of the pancreas appears hypoechoic on ultrasonography (USG) [Figure 1], homogeneously hypodense on computed tomography (CT), and T1 hypo- and mildly T2 hyperintense on magnetic resonance imaging (MRI).<sup>17</sup> Variable contrast enhancement patterns have been described.<sup>18</sup> Unlike chronic alcoholic pancreatitis, calcification and pseudocyst are rarely found in AIP.<sup>18</sup> In endoscopic retrograde cholangiopancreatography, segmental or diffuse irregular narrowing of the main pancreatic duct is commonly found.<sup>19</sup>

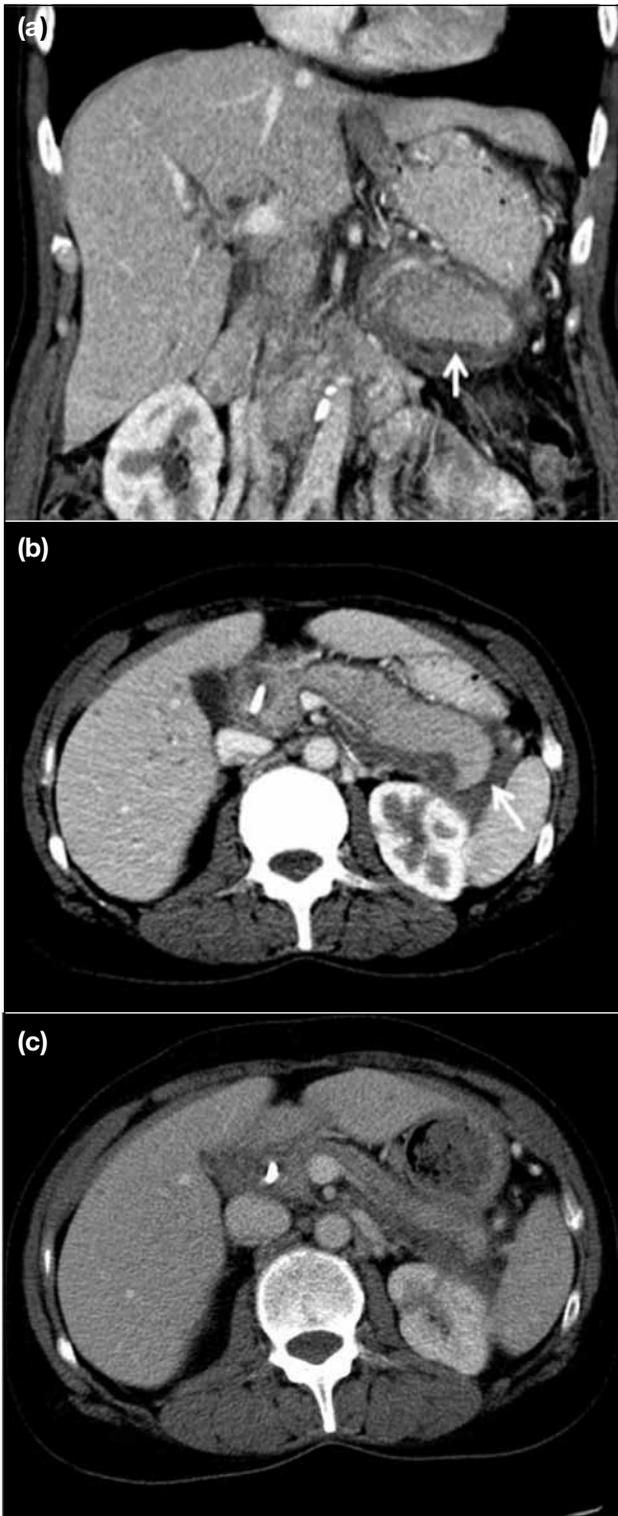
There are two main types of AIP: the diffuse and focal type. The diffuse type is more common,<sup>16</sup> which appears as diffuse enlargement of pancreas with sharp margins, loss of lobular contour, and absence of pancreatic clefts.



**Figure 1.** Diffuse autoimmune pancreatitis in a 62-year-old woman. A transabdominal ultrasound scan shows diffusely swollen and hypoechoic pancreas.

The diffuse type of AIP can resemble acute pancreatitis but the distinctive absence of peri-pancreatic fat stranding, fat necrosis, and fluid collection as well as the presence of a capsule-like rim or halo help to differentiate between the two conditions (Figure 2).<sup>20</sup> This capsule-like rim or halo around the pancreatitis is mainly due to lymphoplasmacytic inflammatory infiltrates and fibrotic change, which sometimes demonstrates delayed contrast enhancement.<sup>16</sup> A diffusely enlarged pancreas can also be found in malignant lymphoma, plasmacytoma, metastases, and diffuse infiltrative pancreatic carcinoma.<sup>16</sup> The pancreas, in most of these conditions however, shows heterogeneous attenuation with an irregular contour which is in contrast with that seen in AIP.<sup>16</sup>

The focal type often presents as a solitary mass in the proximal pancreas, which may mimic pancreatic carcinoma (Figure 3).<sup>12</sup> The homogeneous enhancement of the pancreas, and absence of encasement of mesenteric vessels or local invasion of the adjacent organs or distant metastasis help to differentiate AIP from pancreatic carcinoma.<sup>18</sup> Also, parenchymal atrophy proximal to the mass, commonly seen in pancreatic carcinoma, is usually not observed in AIP.<sup>12</sup> This is probably because the acinar parenchyma in AIP, although atrophic, is replaced with fibrous tissue. Hence, the overall size of the pancreas does not usually change.<sup>21</sup> Moreover, in patients with segmental narrowing of the main pancreatic duct due to AIP, the main pancreatic duct proximal to the segmental narrowing usually shows minimal or no dilatation which is in contrast to the significant dilatation of the main pancreatic duct proximal to the narrowed segment seen



**Figure 2.** Diffuse autoimmune pancreatitis in a 53-year-old woman. (a) Coronal and (b) axial contrast-enhanced computed tomography (CT) shows a diffusely swollen, homogeneous, enhancing pancreas surrounded by a hypodense halo (arrow). Peri-pancreatic stranding, fat necrosis, and fluid collection are typically absent. (c) Axial contrast-enhanced CT performed after steroid treatment shows marked decrease in swelling of the pancreas.



**Figure 3.** Focal, autoimmune pancreatitis in a 63-year-old man which mimics pancreatic cancer. A contrast-enhanced axial computed tomography image shows ill-defined, slightly low-attenuation area in the head of pancreas (arrows). Note the absence of encasement of mesenteric vessels and local invasion of the adjacent organ.

in pancreatic carcinoma.<sup>12,18</sup> The fibro-inflammatory infiltrates aggregate around medium and large ducts in the pancreas, and the varying degree of fibrosis around the pancreatic duct probably explains why the duct is minimally dilated or much less dilated as compared with the ductal dilatation seen in pancreatic carcinoma.<sup>16</sup>

In the spectrum of IgG4-RSD, established clinical diagnostic criteria are only available for AIP. These are based on several factors including imaging, serological, and histological findings as well as presence of extra-pancreatic involvement and response to corticosteroid therapy. According to the Japanese Pancreas Society, the criteria for diagnosing AIP must include typical imaging findings with either serological or histological findings.<sup>22</sup> Kim et al<sup>23</sup> from Korea revised the Japanese criteria by adding ‘response to steroids’ into the inclusion criteria but the presence of imaging findings remains essential. The HISORt criteria proposed by the Mayo Clinic suggest that the diagnosis of AIP can be confirmed if one or more of the aforementioned five factors are present.<sup>24</sup> More recently, the 14th Congress of the International Association of Pancreatology proposed other criteria, which are similar to those proposed by the Mayo Clinic with ‘response to steroids’ being optional. Levels 1 and 2 were also introduced based on the diagnostic reliability of each feature.<sup>25</sup>

## EXTRAPANCREATIC MANIFESTATIONS

### Biliary Tract

IgG4-RSD involvement of bile ducts presents as IgG4-related sclerosing cholangitis (IgG4-SC), which is found in up to 92.5% of patients with AIP.<sup>26</sup> In IgG4-SC, the affected segments of the biliary tree demonstrate wall thickening<sup>27</sup> and stricture.<sup>18</sup> The most commonly involved segment is the intrapancreatic segment of the common bile duct (Figure 4).<sup>16</sup> Less commonly multifocal intrahepatic biliary strictures are involved which mimic primary sclerosing cholangitis (PSC). This can be differentiated from IgG4-SC by cholangiography as band-like stricture with beaded or pruned-tree appearances; diverticulum-like formation are only found in PSC<sup>28</sup> while long stenosis, segmental stricture, and a long stricture with pre-stenotic dilatation are significantly more common in IgG4-SC. However, when focal biliary stricture occurs at the hilar region, especially in the presence of soft-tissue mass, IgG4-SC can resemble hilar cholangiocarcinoma (Klatskin

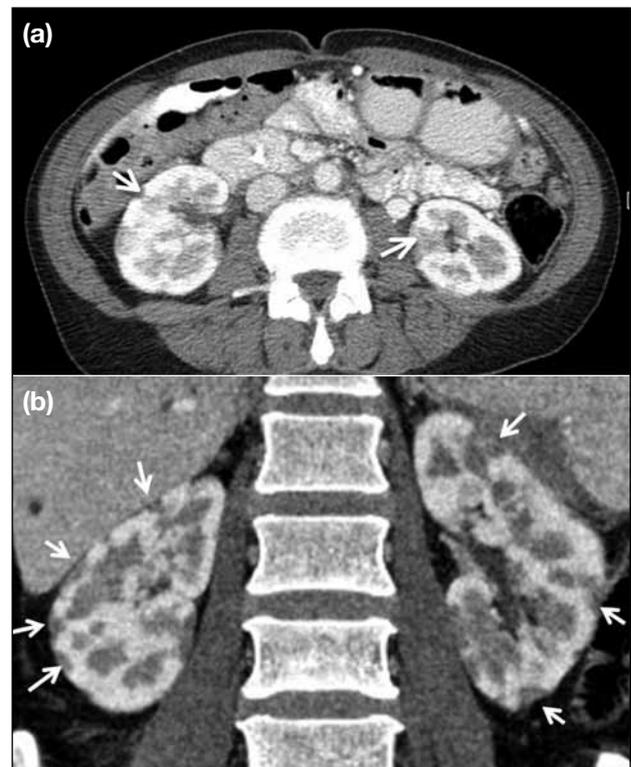
tumour)<sup>29</sup> and biopsy remains the mainstay for differentiating the two conditions.

### Kidney

Renal involvement is found in 35% of cases of IgG4-RSD. Multiple hypoenhancing parenchymal lesions on contrast-enhanced CT are the most common findings. These lesions can appear as small peripheral cortical nodules, round or wedge-shaped lesions (Figure 5) or diffuse, patchy involvement.<sup>4</sup> The wedge-shaped parenchymal lesions mimic pyelonephritis or vascular infarcts<sup>30</sup> while multiple, peripheral cortical nodules or diffuse patchy involvement can mimic metastases,<sup>31</sup> lymphoma,<sup>32</sup> or Wegener's granulomatosis.<sup>33,34</sup> IgG4-RSD can also involve the renal sinus or renal pelvis wall with soft tissue masses (Figure 6) or thickening of renal pelvis wall. This can imitate lymphoma or urothelial tumour.<sup>4</sup> Hence, it is important to recognise that these renal lesions may be secondary to IgG4-related disease, particularly when there is evidence of pancreatic abnormality.



**Figure 4.** An endoscopic retrograde cholangiopancreatography image shows focal narrowing of the intrapancreatic segment of the common bile duct (arrow) in a 53-year-old woman with autoimmune pancreatitis.



**Figure 5.** Immunoglobulin G4-related renal involvement in a 52-year-old woman. (a) Axial and (b) coronal contrast-enhanced computed tomography images show multiple, bilateral, small, wedge-shaped, hypoenhancing, cortical lesions in the kidneys (arrows).



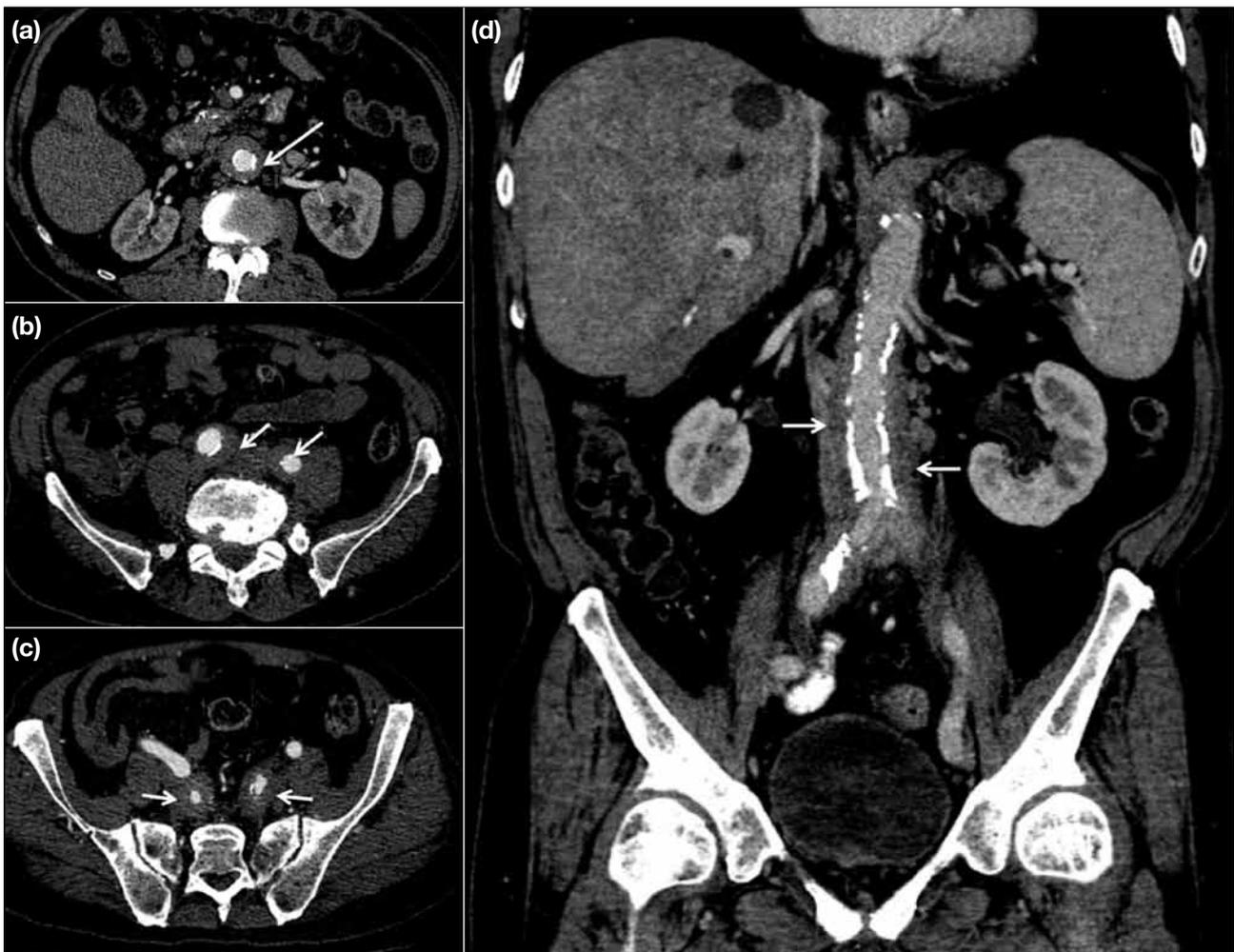
**Figure 6.** Immunoglobulin G4-related renal involvement in a 64-year-old man. A contrast-enhanced computed tomography scan shows soft tissue mass around the renal sinus and renal pelvis wall (arrow).

### Retroperitoneum

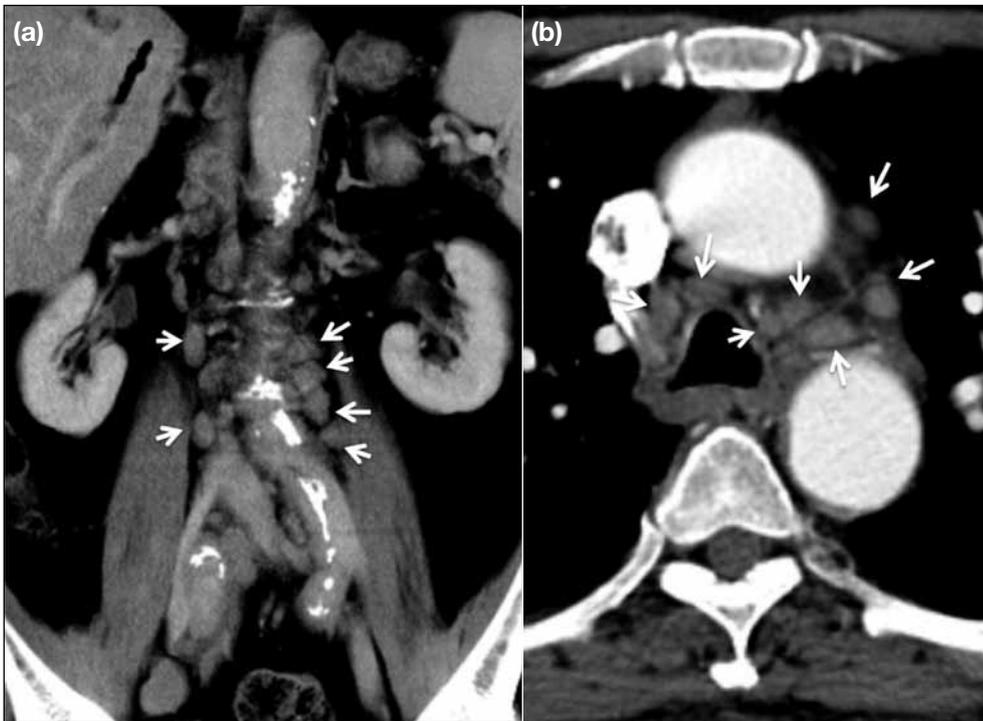
IgG4-RSD manifests in the retroperitoneum as retroperitoneal fibrosis (RPF), which is found in 12.5%<sup>35</sup> to 20%<sup>27</sup> of patients with the condition. It resembles RPF resulting from other causes and is believed to be responsible for more than two-thirds of the cases of the idiopathic form of RPF.<sup>36</sup> Three imaging patterns have been described, depending on the site of involvement: (1) a peri-aortic or arterial mass involving the connective tissue around the abdominal aorta or its first branches (Figure 7); (2) a peri-ureteral mass; and (3) a plaque-like mass that broadly involves the retroperitoneum.<sup>37</sup>

### Lymph Node

Lymph node involvement is found in 80.4%<sup>35</sup> of



**Figure 7.** Immunoglobulin G4-related retroperitoneal fibrosis in a 75-year-old man. Axial contrast-enhanced computed tomography images show enhancing soft tissue thickening (arrows) around the (a) aorta, (b) bilateral common iliac, and (c) internal iliac arteries. (d) A coronal image shows diffuse soft tissue thickening along the aorta and its branches.

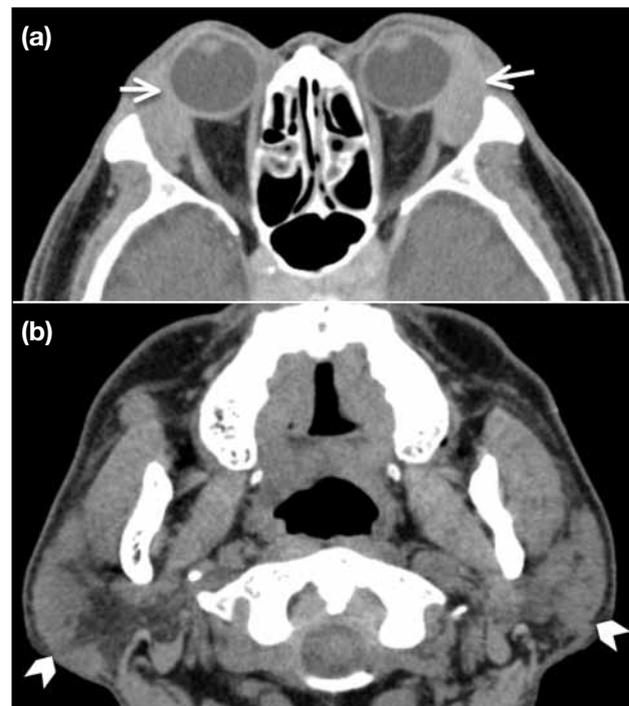


**Figure 8.** Immunoglobulin G4-related abdominal and mediastinal lymphadenopathy in a 75-year-old man. (a) A coronal contrast-enhanced computed tomography (CT) scan shows para-aortic lymphadenopathy (arrows). (b) An axial contrast-enhanced CT scan shows right paratracheal and aortopulmonary lymphadenopathy (arrows).

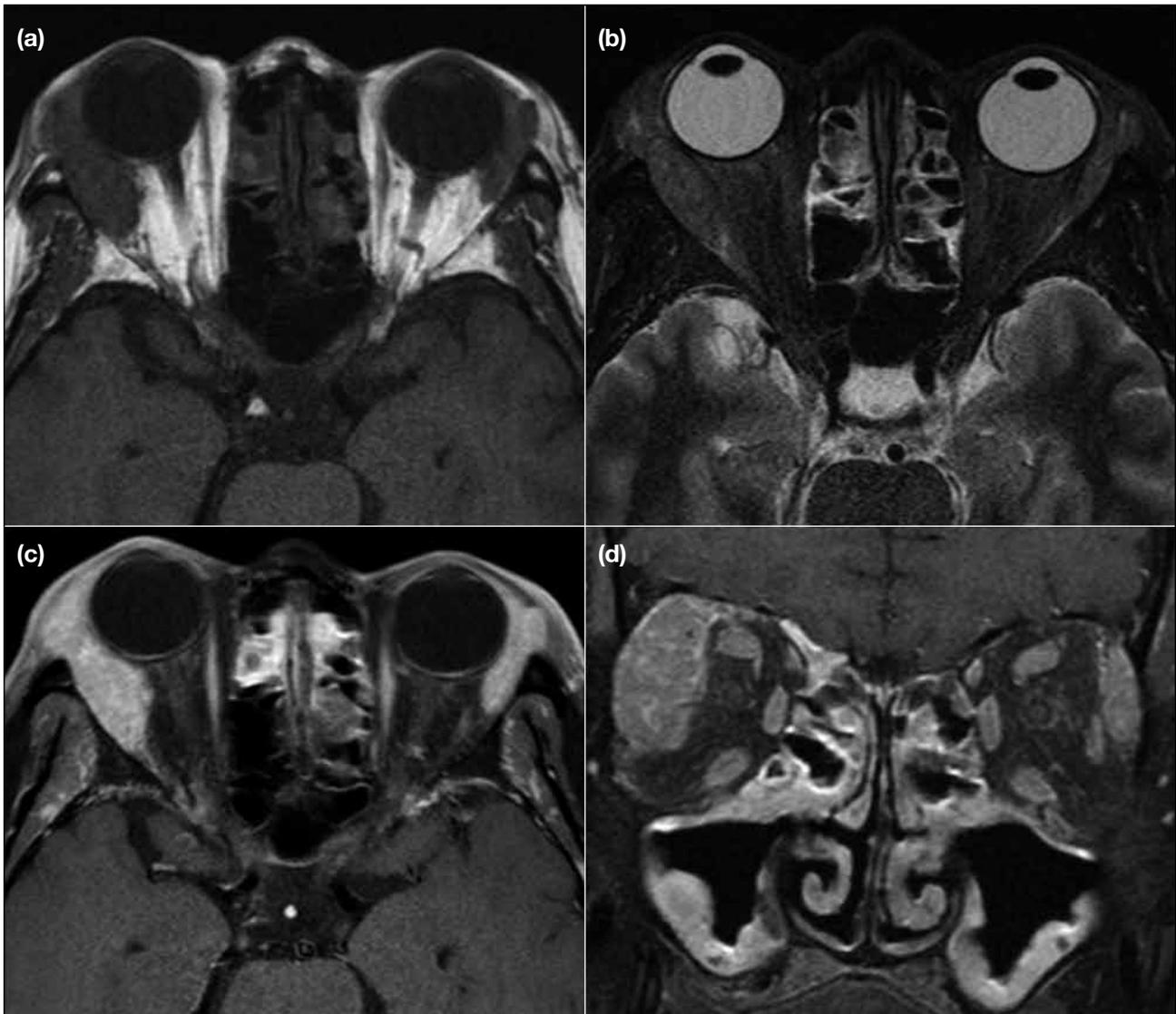
patients and may even be the initial manifestation. It can manifest as generalised lymphadenopathy or localised disease adjacent to a specific, affected organ. The size of the lymph nodes varies and may be as large as 2 cm (Figure 8).<sup>35</sup> Lymphadenopathy associated with IgG4-RSD is not easily distinguished from lymphoma, nodal metastasis, and reactive lymph node enlargement. Only the coexistence of lymphadenopathy and pancreatic lesions may help to differentiate.<sup>27</sup>

### Orbit

In the orbits, IgG4-RSD has been found to affect lacrimal glands in 12.5%<sup>35</sup> of patients with AIP. The patients present with dacryoadenitis which manifests as bilateral diffuse enlargement of the lacrimal glands showing homogeneous enhancement on CT (Figure 9) and MRI (Figure 10). On USG, bilateral hypoechoic nodular areas with increased vascularisation can also be found (Figure 11).<sup>38</sup> Lacrimal gland involvement is often seen in association with salivary gland lesion as in Mikulicz's disease, which is also considered part of the spectrum of IgG4-RSD (Figure 10).<sup>39</sup> Less commonly, extraocular muscles can also be affected in IgG4-RSD, manifesting as enlargement of the muscle bellies as well as the tendinous insertion. The latter helps in



**Figure 9.** Mikulicz's disease in a 75-year-old man. (a) An axial, contrast-enhanced computed tomography (CT) scan shows diffuse, symmetrical swelling of the lacrimal glands with homogeneous enhancement (arrows). (b) A contrast-enhanced CT scan demonstrates concomitant diffuse swelling of bilateral parotid glands (arrowheads).



**Figure 10.** Magnetic resonance images of a 64-year-old man with immunoglobulin G4-related dacryoadenitis. (a) Axial T1-weighted, (b) T2-weighted, (c) post-gadolinium fat-suppressed T1-weighted and (d) coronal post-gadolinium fat-suppressed T1-weighted images show bilateral asymmetrical enlargement of lacrimal glands which are homogeneously hypointense on T1, mildly hyperintense on T2 and show diffuse contrast enhancement.

distinguishing IgG4-RD from Graves' ophthalmopathy (Figure 12).<sup>37</sup>

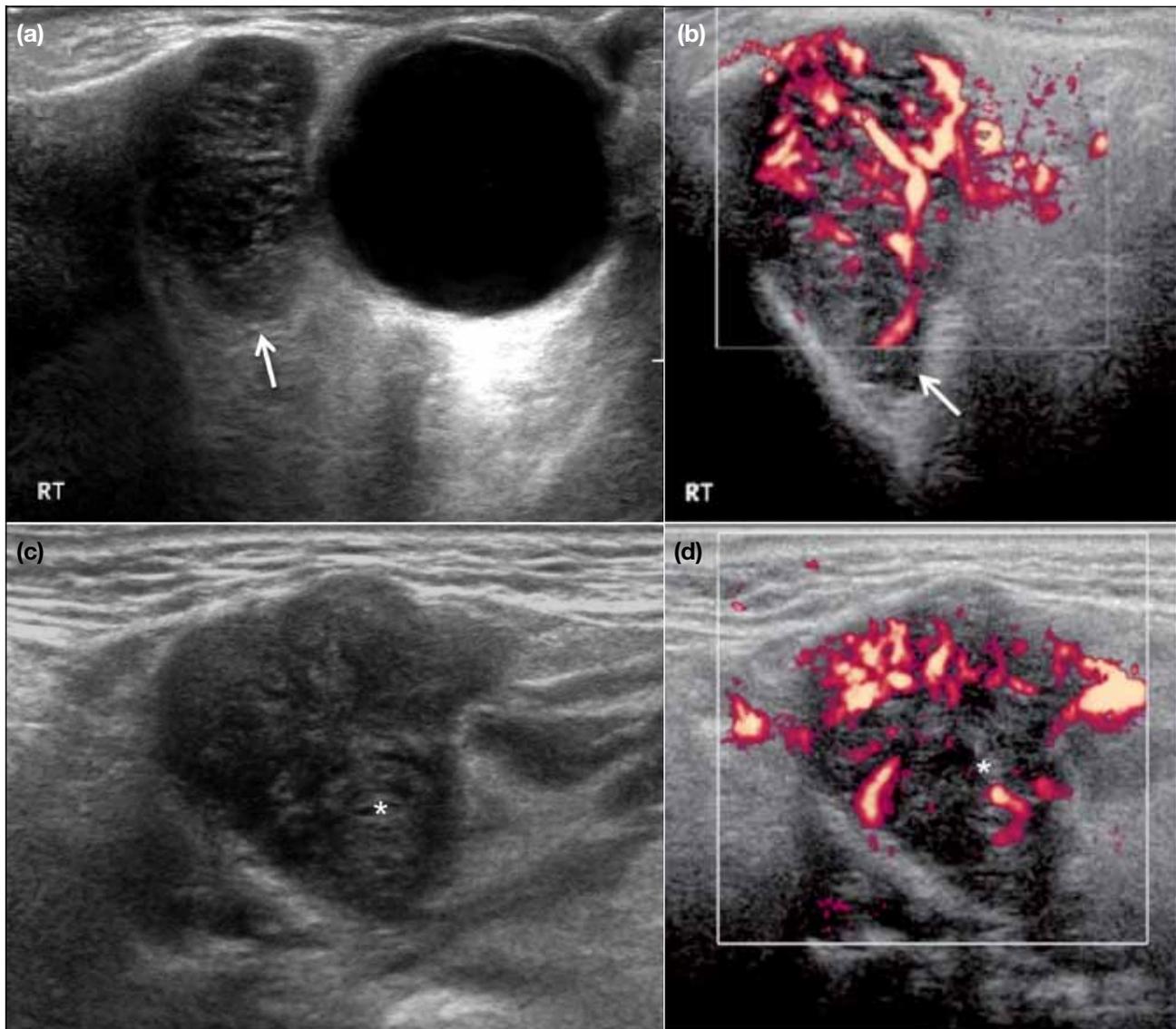
### Salivary Gland

The salivary gland involvement is common and found in 25.9% of patients with AIP.<sup>35</sup> They commonly present with bilateral diffuse swelling of the glands, which appear enlarged and homogeneously enhanced on CT (Figure 9) and MRI. Similar to lacrimal gland involvement, the lesions present as bilateral, hypoechoic nodular areas with considerable vascularisation on USG (Figure 11).<sup>38</sup> However, these findings are non-specific

and viral infection (mumps), lymphoma, Sjögren's syndrome, and sarcoidosis should also be considered in the differential diagnosis. Less commonly, IgG4-RSD can present with isolated involvement of the salivary glands and mimic a neoplasm. Sclerosing sialadenitis (Küttner's tumour), which is part of the spectrum of IgG4 RSD, can present as a firm, unilateral submandibular gland swelling and imitate a neoplasm.<sup>40</sup>

### Lungs

Pulmonary involvement in IgG4-RSD has been reported in 13% of patients with AIP.<sup>41</sup> The imaging



**Figure 11.** Mikulicz's disease in an 87-year-old man. Ultrasonography images of the (a) right lacrimal gland (arrow) and (c) right submandibular gland (asterisk) demonstrate diffuse swelling with nodular hypoechoic areas with (b and d) increased vascularisation.

findings are categorised into four major subtypes<sup>5</sup>: (1) solitary or multiple solid nodular type (Figure 13); (2) round-shaped, ground-glass opacity type; (3) alveolar interstitial type which includes honeycombing, bronchiectasis, and diffuse ground-glass opacities; and (4) bronchovascular type, which is characterised by thickening of the bronchovascular bundles and interlobular septa. These findings may be difficult to differentiate from lung tumour or metastases (solid nodular), sarcoidosis (bronchovascular pattern), bronchioloalveolar carcinoma (round area with ground-glass opacification), and interstitial lung disease (bronchiectasis, honeycombing, diffuse ground-glass changes); thus, biopsy is usually required for a definitive

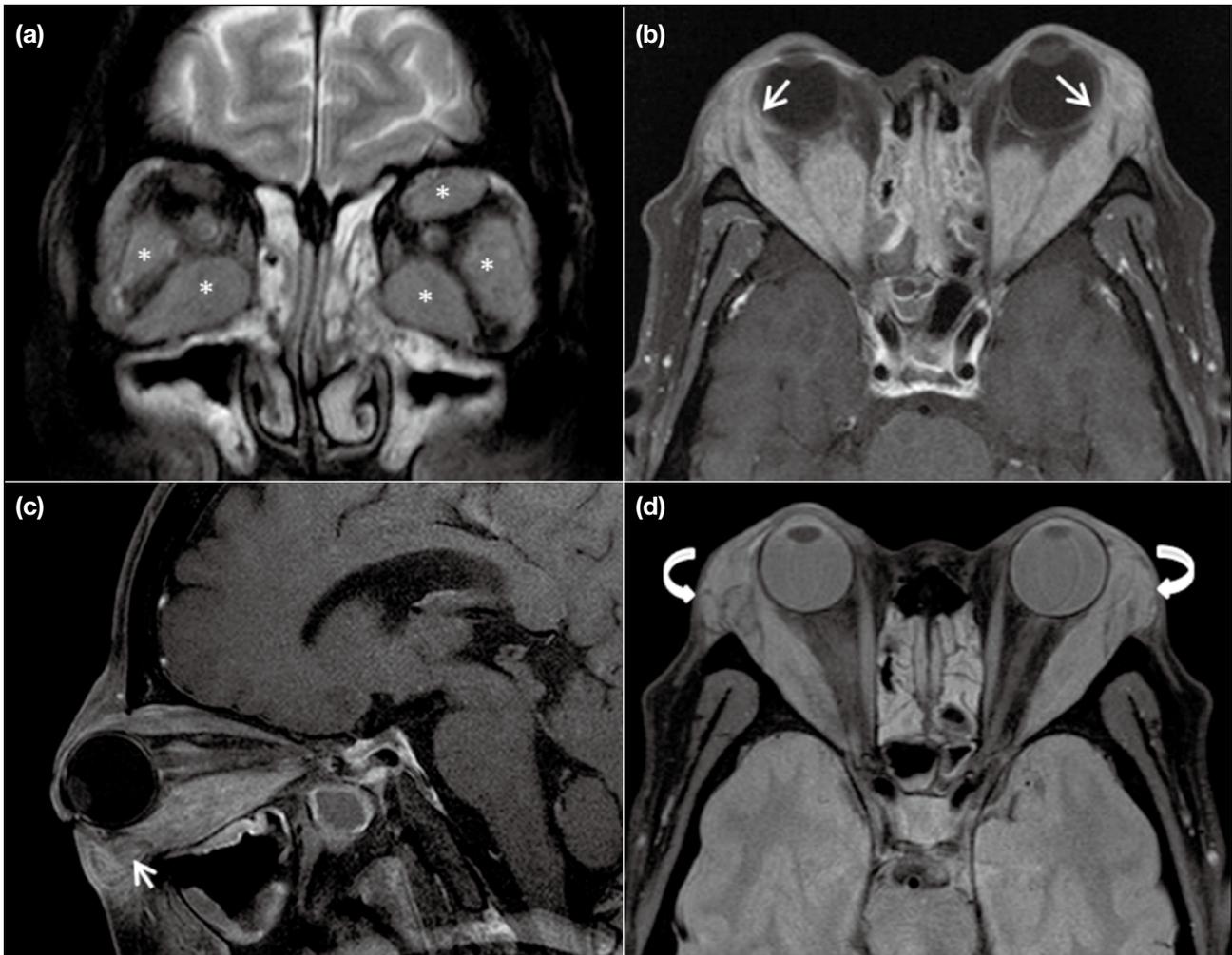
diagnosis.<sup>17</sup>

### Other Organ Manifestations

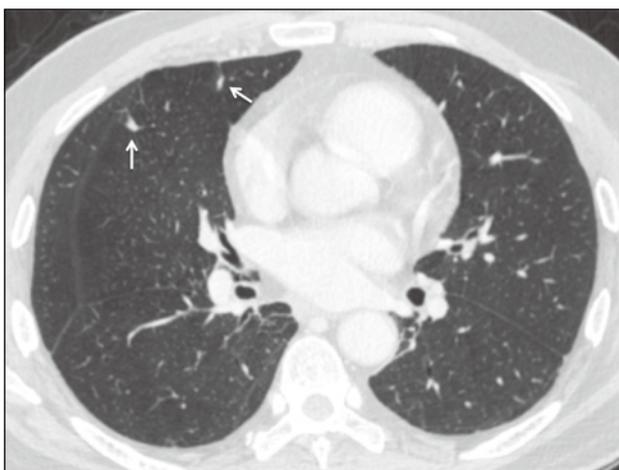
Manifestations of IgG4-RSD in many other organs including the gallbladder, liver, breast (pseudotumour), prostate, pericardium (constrictive pericarditis), skin, ears, nose, paranasal sinuses, thyroid, peripheral nerve (perineural inflammation), pituitary gland and meninges have been reported, and the list of organs associated with the disease is still growing.<sup>37</sup>

### CONCLUSION

Besides AIP, IgG4-RSD has a broad spectrum of extra-pancreatic manifestations. It is important to recognise



**Figure 12.** Immunoglobulin G4 (IgG4)-related lacrimal and extraocular muscle involvement in a 53-year-old woman. (a) A coronal T2-weighted magnetic resonance (MR) image demonstrates marked swelling of the extraocular muscles (asterisks). (b) Axial and (c) sagittal contrast-enhanced T1-weighted MR images show bilateral proptosis with diffuse extraocular muscle swelling. The involvement of the tendonous insertion (arrows) helps to differentiate IgG4-related disease from Graves' ophthalmopathy. (d) An axial proton density-weighted MR image shows concomitant enlargement of bilateral lacrimal glands (curved arrows).



**Figure 13.** Immunoglobulin G4-related pulmonary involvement in a 65-year-old man. Two solid nodules (arrows) are noted in the anterior aspect of right middle lobe.

the possible multiorgan involvement of the disease to facilitate early diagnosis and prompt initiation of treatment, and avoid unnecessary investigations or interventions.

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