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REVIEW ARTICLE

Infrequent organ involvement of IgG4-related diseases: a literature review

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Abstract

IgG4-related disease (IgG4-RD) is a chronic and systemic disease that can involve multiple organs. The most commonly involved organs include the salivary glands, orbital adnexal structures, paranasal sinus, thyroid, lungs, breasts, aorta, pancreas, biliary ducts, kidneys, retroperitoneum, lymph nodes, prostate, pituitary, and endocranium. Due to increased disease research, several new site-specific nuances of IgG4-RD have been described. The authors have reviewed the recent literature and briefly summarize the infrequent organ involvement of IgG4-RD.

Keywords IgG4-related disease · Infrequent organ involvement · Literature review · Systemic disease

IgG4-related disease (IgG4-RD) is a chronic and systemic disease with unknown causes, characterized by the infiltration of IgG4+ plasma cells together with distinct storiform fibrosis in involved lesions and significantly increased serum IgG4 levels. In 2010, an *Autoimmunity Reviews* article entitled "The birthday of a new syndrome: IgG4-related diseases constitute a clinical entity" was published, demonstrating that this new disease was internationally recognized by experts [1]. Due to the short duration of understanding of the disease, investigators summarized the most commonly involved organs, including the salivary glands, orbital adnexal structures, paranasal sinus, thyroid, lungs, breasts, aorta, pancreas, biliary ducts, kidneys, retroperitoneum, lymph nodes, prostate, pituitary, and endocranium [2]. However, due to increased disease research, several new site-specific nuances of IgG4-RD have been described. The authors have reviewed the recent literature and briefly summarize the infrequent organ involvement of IgG4-RD.

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Bibliographic databases were searched including PubMed and Embase, from inception to May 2017 with no language restrictions, supplemented with manual reference searches. As the disease and type of manifestations that we were looking for were infrequent organ involvement of IgG4-RD, and most rare cases were presented in the form of case reports, we set the search key words: immunoglobulin G4-related disease OR IgG4-related disease OR IgG4-RD; case report. Case reports describing commonly involved organs as mentioned above like salivary glands, orbital adnexal structures, paranasal sinus, etc. were excluded. In order to avoid the omission of literatures, we also referred to a number of reviews and treatises. The classification of subtitles is mainly classified according to the general system and location of the human body, but because of the complex characteristics of the disease, it does not strictly follow any regular system division.

Chest and diaphragm

The most common manifestations of IgG4-related chest and diaphragm diseases are interstitial pneumonia, which sometimes involves the lung parenchyma. Uncommon manifestations were also found in the literature, including bronchial wall thickening, bilateral pleural effusion, chylothorax, low tracheal masses, pneumonia during the puerperium, thymus enlargement, and middle-posterior mediastinal lesions. Hayashi M et al. reported a case of IgG4-RD with marked thickening of the bronchial wall, cavitating lung disease, and

hardening and swelling of the salivary glands [3]. The diagnosis was supported by a histopathologic examination of the bronchial mucosa biopsy specimen. Jinnur PK et al. reported a 60-year-old man who presented with hemoptysis and cavitating lung disease with clinical, laboratory, and histopathologic findings compatible with IgG4-RD [4]. Ishida et al. [5] reported an IgG4-related pleural disease presenting as a massive bilateral effusion supported by pathologic evaluation of the pleural biopsy specimen. Kato et al. [6] also reported a case of IgG4-related pleuritis with chylothorax, suggesting that IgG4-RD can cause chylothorax. Kobraei et al. [7] reported a patient with IgG4-related airway disease who presented with a low tracheal mass and was treated with tracheal resection and reconstruction. Wang et al. [8] reported a 24-year-old woman diagnosed with IgG4-related lung disease in the puerperium who developed suspected IgG4-related pneumonia and was effectively treated with corticosteroids. In addition to pulmonary lesions, mediastinal involvement was also reported. Kazantseva et al. [9] reported a case of IgG4-RD that mimicked lymphoma in a 59-year-old woman with generalized lymphadenopathy. The patient died due to bilateral pneumonia, and an autopsy revealed a tumor-like mass in the thymus, which was shown to be IgG4-related thymus enlargement. Maki et al. [10] reported a case of IgG4-RD with a middle-posterior mediastinal lesion that reduced following steroid treatment.

Cardiovascular system

The most common manifestation of IgG4-related vasculitis is aortic peripheral inflammation, especially around the abdominal aorta. Uncommon manifestations were also described in the literature, including inflammation in and around the coronary artery, constrictive pericarditis, pulmonary arterial hypertension, pseudotumor on the mitral and aortic valves, isolated thoracic aortic aneurysm, involvement of the superior vena cava and the atrial septum of the heart, and small-vessel vasculitis. Coronary artery involvement is relatively rare, but the symptoms are more severe. Jinnur PK et al. reported coronary findings in a patient with multi-organ IgG4-RD. Computed tomography of the chest demonstrated nodular and periarterial soft tissue thickening along the proximal coronary artery segments [11]. Patel NR et al. reported a case of sudden cardiac death due to coronary artery involvement by IgG4-RD. An autopsy was performed, revealing multi-organ involvement by IgG4-RD, including the coronary arteries. The inflammation and fibrosis, combined with concomitant atherosclerotic disease, resulted in severe stenosis and thrombosis of the coronary arteries, which led to cardiac hypoperfusion, myocardial infarction, and, ultimately, sudden cardiac death [12]. Atallah et al. [13] reported an IgG4-RD case with effusive-constrictive pericarditis and tamponade. Yanagi H et al. also reported a patient with constrictive pericarditis caused by IgG4-RD whose diastolic function improved with corticosteroid therapy [14]. Ishida et al. [15] reported a 22-year-old woman with generalized lymphadenopathy and symmetrical swelling of the lacrimal and submandibular glands diagnosed with IgG4-RD, whose echocardiography and right heart catheterization revealed a high mean pulmonary arterial pressure. Shirai et al. [16] also reported an IgG4-RD patient in pulmonary arterial hypertension who received long-term epoprostenol treatment. Besik J et al. reported a 64-year-old woman, with an extensive inflammatory IgG4-related pseudotumor on her mitral and aortic valves, who underwent replacement of both affected valves [17]. Maleszewski et al. [18] reported two cases of IgG4-RD of the aortic valve. Nunez Fernandez et al. [19] reported IgG4-RD with isolated thoracic aortic aneurysm. Song et al. [20] reported a 55-year-old woman with IgG4-RD involving the superior vena cava and the atrial septum of the heart who received a permanent pacemaker because of frequent sinus pauses and syncope. Alba et al. [21] reported a case of small-vessel vasculitis involving muscle, peripheral nerves, and the kidneys with prominent IgG4-positive plasma cell infiltrates, which expands the spectrum of vascular involvement in the context of IgG4-RD.

Urogenital system

The most common manifestations of IgG4-RD involving the urogenital system include tubulointerstitial nephritis and glomerular lesions. Uncommon manifestations were also found in the literature, including renal pelvic mass, renal hilum mass, thickness of the ureter wall, testicular inflammation, and postmenopausal uterine enlargement. Zhang et al. [22] reported a case of a renal pelvis mass that mimicked urothelial carcinoma. Ramasamy et al. [23] reported a patient with a large infiltrative mass centered on the right renal hilum with biopsy demonstrating IgG4-RD and who experienced spontaneous resolution after 6 months. Lei et al. [24] reported a 66-year-old Asian man with severe left hydronephrosis because of marked thickness of the left ureter wall, who was diagnosed with IgG4-related kidney disease mimicking a malignant ureteral tumor. Hayakawa et al. [25] reported a 70-year-old woman with circumferential wall thickening in the right middle ureter. A possible diagnosis of segmental ureteritis due to IgG4-RD was established, and the patient improved with steroid therapy. de Buy Wenniger et al. [26] reported a case of a patient with IgG4-related testicular inflammation and postulated that IgG4-RD in the urogenital tract was not restricted to kidney disease and prostatitis but may also affect the testes. Ohkubo H et al. reported a 68-year-old woman with post-menopausal uterine enlargement. A total hysterectomy was performed, and IgG4-RD was confirmed by immunohistochemical findings. This was the first report of IgG4-RD involving the uterus [27].

Nervous system

Compared to other organs, IgG4-RD involvement in the nervous system is relatively rare; hypertrophic pachymeningitis and hypophysitis were the most frequently reported conditions. Infrequent manifestations include hypertrophic pachymeningitis with brain parenchymal invasion, intracranial pseudo-tumors, autoimmune hypophysitis rapidly converting to an empty sella, autoimmune hypophysitis with ntracranial multi-organ involvement, bilateral intracranial optic nerve and chiasmal involvement, trigeminal nerve involvement, and extensive peripheral nerve involvement. Li et al. [28] reported a case of IgG4-related hypertrophic pachymeningitis at the falx cerebri with brain parenchymal invasion but without extracranial involvement. Okano et al. [29] reported a case of a 62-year-old man who presented with visual disturbance and quadrantanopia of the right eye and who had experienced autoimmune pancreatitis 7 years previously. He was diagnosed with intracranial pseudotumors associated with IgG4-RD and improved with oral corticosteroid medication. Nakasone et al. [30] reported a case of autoimmune hypophysitis with rapid conversion to an empty sella and an immediate decrease of the serum IgG4 level in only 12 days. Kanoke et al. [31] reported three cases of autoimmune hypophysitis with intracranial multi-organ involvement. Behbehani et al. [32] reported a case of IgG4-RD with bilateral intracranial optic nerve and chiasmal involvement. Katsura et al. [33] reported a patient with IgG4-RD in the trigeminal nerve, who was diagnosed histopathologically, without involvement of any of the common sites. Fujii et al. [34] reported a patient with IgG4-RD and extensive peripheral nerve involvement that progressed from localized IgG4-related lymphadenopathy, suggesting that IgG4-RD may affect many organs through peripheral nerve involvement. Soussan et al. [35] reported a 55-year-old woman with IgG4-related diffuse perineural disease involving the right optic nerve and orbital muscles and a diffuse infiltration involving the lumbodorsal and sacral nerve roots.

Gastrointestinal tract and mesentery

IgG4-RD can involve the gastrointestinal tract and the mesentery; however, this manifestation is not common. Lopes J et al. reported a case of a 23-year-old man who presented with dysphagia, weight loss, and recurrent esophageal strictures. Endoscopic ultrasound with esophagogastroduodenoscopy revealed a mass present in the distal esophagus. Then, the patient was diagnosed with autoimmune esophaitis as part of the IgG4 spectrum of diseases [36]. Oh et al. [37] also reported a case of IgG4-related esophaitis. Na et al. [38] reported a case of gastric nodular lesions caused by IgG4-RD. Frydman et al. [39] presented an acute gastric-pericardial fistula secondary to IgG4-RD that required urgent operative management. Hasosah et al. [40] reported a 7-year-old child with a history of recurrent abdominal pain and fever, who was found to have an intra-abdominal mass suspicious for malignancy. A tissue biopsy revealed a diagnosis of sclerosing mesenteritis associated with IgG4-RD. Kim et al. [41] reported a case of IgG4-related sclerosing mesenteritis that occurred in conjunction with Crohn's disease. Laco et al. [42] reported a case of a 65-year-old woman with enterocolic lymphocytic phlebitis (ELP), who was then diagnosed with definite IgG4-RD, presuming that a subset of ELP is a manifestation of IgG4-RD. Wong et al. [43] reported a 46-year-old woman with IgG4-RD of the small bowel presenting with necrotizing mesenteric arteritis and a solitary jejunal ulcer.

Skin

IgG4-related skin diseases often affect the head and neck region, primarily manifesting with painless plaques, itching erythema, or subcutaneous nodules. Ise et al. [44] reported a patient with IgG4-RD, autoimmune pancreatitis, and skin lesions on the trunk and limbs, whose skin lesions responded well to oral prednisolone. Kempeneers et al. [45] described a 73-year-old white man who presented with two infiltrated, erythematous nodules on his abdomen. The histopathological characteristics were highly suggestive of IgG4-related cutaneous disease. Kondo et al. [46] presented a 53-year-old healthy factory worker with small nodules of similar size, shape, and location on both ears, who was diagnosed with IgG4-related skin disease without multiple organ involvement. Iwata et al. [47] reported a case that was first considered pseudolymphoma based on the histopathological analysis but was finally diagnosed as IgG4-related skin lesions. They then performed immunostaining on past potential cases of IgG4-RD and found that of the 32 cases of cutaneous pseudolymphomas, two cases (6.3%) satisfied IgG4-RD histopathological diagnostic criteria. This finding suggested that the clinical presentations and histopathological features of skin involvement in IgG4-RD are analogous to cutaneous B-cell pseudolymphoma; thus, careful identification is required.

Neck

IgG4-related thyroiditis is the most frequently reported organ involvement in the neck. Cheuk et al. [48] reported four cases of IgG4-related idiopathic cervical fibrosis with infiltrative, firm cervical masses. McKinnon et al. [49] reported a case of IgG4-RD presenting as a solitary neck mass. Tirelli et al. [50] described the case of a patient with bilateral neck masses and bilateral neck abscesses who underwent bilateral neck incision for drainage of the abscesses on two occasions, followed by a neck dissection after a third recurrence. Immunohistochemistry of the specimen revealed IgG4-RD.

Bone and joint

Few reports have described IgG4-related bone and joint disease. Bittencourt et al. [51] reported a case of IgG4-RD involving the temporal bone, with a soft tissue density mass in the bone found by computed tomography, which was finally confirmed by immunohistochemical analysis. Windisch et al. [52] presented a male patient with this disease involving the lymph nodes and possibly the joints and kidneys, with the symptom of joint swelling involving fingers, wrist, and ankle joints. Tomiyama et al. [53] reported a 71-year-old man with swelling of the right shoulder and left wrist joints who was diagnosed with IgG4-RD and successfully treated with corticosteroid therapy, suggesting that erosive arthritis may occur in patients with IgG4-RD.

Eyes, ears, and mouth

IgG4-RD usually affects periorbital tissues, including the lacrimal gland, extraocular muscles, and periorbital soft tissue of the submandibular and parotid glands, which is known as Mikulicz's disease. Ohno et al. [54] reported a case of IgG4-RD involving the sclera. Prayson [55] reported an IgG4-RD patient presenting as uveitis. London et al. [56] reported two cases of adult-onset asthma and periocular xanthogranuloma (AAPOX) that fulfilled all of the diagnostic criteria for definite IgG4-RD, demonstrating a strong relationship between IgG4-RD and AAPOX syndrome. Schiffenbauer et al. [57] reported a case of IgG4-RD presenting as recurrent mastoiditis. Khurram et al. [58] reported a case with lesions involving the tongue and palatine tonsil resembling oral squamous cell carcinoma. Khoo et al. [59] presented a case of supraglottic IgG4-related plasma cell granuloma successfully treated with oral corticosteroids. Reder et al. [60] reported three cases of IgG4-RD with laryngopharyngeal involvement. Shaib et al. [61] described two male patients with an ongoing medical history of a predominant laryngeal focus, who initially received the diagnoses granulomatous polyangiitis (GPA) and Behcet's disease and were finally diagnosed with IgG4-RD. Wilsher [62] reported a case of IgG4-RD involving the thyroglossal duct.

Table 1 Summary of the common and uncommon manifestations of IgG4-RD

Region	Common manifestations as recognized	Uncommon manifestations as reported
Chest and diaphragm	Interstitial pneumonia; lung parenchyma	Bronchial wall thickening; bilateral pleural effusion; chylothorax; low tracheal masses; pneumonia during the puerperium; thymus enlargement; middle-posterior mediastinal lesions
Cardiovascular system	Aortic peripheral inflammation	Inflammation in and around the coronary artery; constrictive pericarditis; pulmonary arterial hypertension; pseudotumor on the mitral and aortic valves; isolated thoracic aortic aneurysm; involvement of the superior vena cava and the atrial septum of the heart; small-vessel vasculitis
Urogenital system	Tubulointerstitial nephritis; glomerular lesions	Renal pelvic mass; renal hilum mass; thickness of the ureter wall; testicular inflammation; Post-menopausal uterine enlargement
Nervous system	Hypertrophic pachymeningitis; hypophysitis	Hypertrophic pachymeningitis with brain parenchymal invasion; intracranial pseudotumors; autoimmune hypophysitis rapidly converting to an empty sella; autoimmune hypophysitis with intracranial multi-organ involvement; bilateral intracranial optic nerve and chiasmal involvement; trigeminal nerve involvement; extensive peripheral nerve involvement
Gastrointestinal tract and mesentery	No common manifestations	Autoimmune esophaitis; gastric nodular lesions; acute gastric-pericardial fistula; sclerosing mesenteritis; enterocolic lymphocytic phlebitis; necrotizing mesenteric arteritis and a solitary jejunal ulcer
Skin	Painless plaques, itching erythema, or subcutaneous nodules in head and neck region	Skin lesions on the trunk and limbs; infiltrated, erythematous nodules on abdomen; small nodules on both ears; mimic pseudolymphoma
Neck	Thyroiditis	Infiltrative, firm cervical masses; solitary neck mass; bilateral neck masses and bilateral neck abscesses
Bone and joint	No common manifestations	A soft tissue density mass in temporal bone; erosive arthritis
Eyes, ears, and mouth	Mikulicz's disease	Scleral involvement; uveitis; adult-onset asthma and periocular xanthogranuloma (AAPOX); recurrent mastoiditis; lesions involving the tongue and palatine tonsil; supraglottic plasma cell granuloma; laryngopharyngeal involvement; thyroglossal duct involvement
Other rare organ involvement	No common manifestations	Hepatic angiomyolipoma (AML); sclerosing angiomatoid nodular transformation (SANT) of the spleen; coexistence with POEMS (polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, and skin changes) syndrome; coexistence with Rosai-Dorfman disease

Other rare organ involvement

Agaimy and Markl [63] reported a case of hepatic angiomyolipoma (AML) with a possible diagnosis of IgG4-RD. Kim et al. [64] reported a case of sclerosing angiomatoid nodular transformation (SANT) of the spleen associated with IgG4-RD. Nishihara et al. [65] described a case of a 57-year-old man who developed bilateral hand and foot numbness, followed by leg weakness and skin pigmentation, who was diagnosed with POEMS syndrome(polyneuropathy, organomegaly, endocrinopathy, monoclonal protein, and skin changes) accompanied with IgG4-related autoimmune pancreatitis, suggesting that these two rare disorders may share common mechanisms. de Jong et al. [66] reported a case of an elderly male referred with pulmonary opacities and extensive mediastinal lymphadenopathy suspicious for lung cancer, who was found to have Rosai-Dorfman disease in a lymph node and concomitant IgG4-RD in the lungs and kidney. Zhao et al. [67] also reported a case of extranodal Rosai-Dorfman disease involving the appendix and mesenteric nodes with a possible relationship to IgG4-RD.

As a synthesis of the reports, we summarized the common and uncommon manifestations of IgG4-RD in Table 1. In conclusion, we found that IgG4-RD can involve almost any organ and can mimic many other diseases, especially malignant tumors. In addition, many other diseases can mimic IgG4-RD, especially lymphoma, which was most frequently reported. When the clinical manifestations are complicated, a careful differential diagnosis must be performed, and the diagnosis of IgG4-RD should be considered. Due to the limited knowledge of this disease and the limited ability to search literature, the authors believe that this paper has many shortcomings and loopholes. Further research and summaries are needed to refine the spectrum of IgG4-RD.

Compliance with ethical standards

Disclosures None.

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