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REVIEW ARTICLE

Multimodalities Imaging of Immunoglobulin 4-Related Cardiovascular Disorders

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ARTICLE HISTORY

Received: August 05, 2018
Revised: January 07, 2019
Accepted: January 08, 2019

DOI:
10.2174/1573403X15666190117101607

Abstract: Immunoglobulin 4 (IgG4)-related systemic disease (IgG4-RSD) is a systemic inflammatory disease characterized by elevation of serum IgG4. IgG4-RSD can affect any organ in the body, and the list of organs associated with this condition is growing steadily. IgG4-related cardiovascular disease affects the coronary arteries, heart valves, myocardium, pericardium, aorta, pulmonary and peripheral vessels. Echocardiography is the most commonly used non-invasive imaging method. Computed tomography angiography (CTA) can assess aortitis, periarteritis and coronary aneurysms. Coronary CTA is fast, offers high spatial resolution and a wide coverage field of view. Cardiac magnetic resonance imaging (CMR) offers a comprehensive evaluation of the cardiovascular system including cardiac function, extent of myocardial fibrosis, characterise cardiac masses with different pulse sequences and guide to further treatment. Fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) can provide important information about the extent of disease, the presence of active inflammation and the optimum biopsy site. In general, the role of diagnostic imaging includes establishing the diagnosis, detecting complications, guiding biopsy and documenting response to therapy.

Keywords: IgG4-RSD, cardiovascular complications, coronary aneurysm, aortitis, computed tomography angiography, CMR.

1. INTRODUCTION

Immunoglobulin 4-related systemic inflammatory disease (IgG4-RSD) is a newly recognized disorder, characterized by infiltration of IgG4 positive plasma cells and fibrosis in systemic organs with elevation of serum IgG4 levels [1]. The term IgG4-RSD defines several clinical entities once regarded as entirely separate diseases, and the list of organs associated with this condition is growing steadily [2]. The presence of numerous IgG4+ plasma cells within affected tissue is the gold standard for diagnosis of IgG4-RSD. The organs associated with IgG4-RSD include pancreas, biliary tree, kidney, lung, lymph nodes, meninges, aorta, breast, prostate, thyroid glands, pericardium and skin. IgG4-related cardiovascular disease affects the coronary arteries, heart valves, myocardium, pericardium, aorta and peripheral vessels [3-6]. Different imaging methods, including echocardiography, computed tomography (CT), cardiac magnetic imaging resonance (CMR), fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) and coronary angiography, have been used for early disease detection, diagnosis and follow-up of complications [7]. Echocardiography is the most common non-invasive imaging method, whereas CTA can assess aortitis, periarteritis and coronary aneurysms. CMR offers a comprehensive evaluation of the cardiovascular system, including cardiac function, the extent of fibrosis, disease-related complications and a guide to further treatment. FDG PET/CT can provide important information on the severity of the disease, presence of active inflammation and the optimum biopsy site [8]. An inflammatory or infectious process can cause abnormal FDG uptake in the aortic wall, and a nonspecific protocol is needed for assessing vasculitis. Metabolic activity of vasculitis is well appreciated on PET images, but morphological assessment is limited because of low spatial resolution. The purpose of this review is to describe various cardiovascular disorders related to IgG4 and the role of different imaging techniques in the early detection, diagnosis and management of complications.

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2. CORONARY ARTERY STENOSIS

IgG4-related vascular inflammation takes place predominantly in the adventitial regions. Adventitial lymphocyte infiltrates are known to play a role in atherosclerotic lesion formation. Although, currently there is limited evidence, IgG4-related perivascular infiltration may play a role in atherogenesis of IgG4-related coronary artery lesions. Coronary stenosis is induced by tumorous formation surrounding the coronary artery or periarterial soft tissue thickening along the coronary artery (Fig. **1**) [6, 7].

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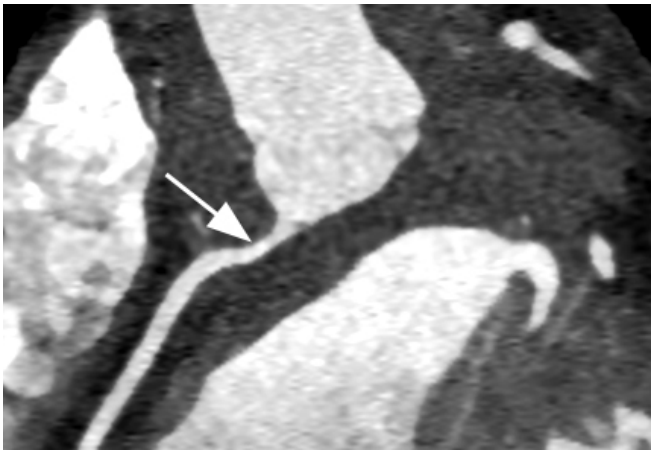

Fig. (1). CT coronary angiogram (multiplanar reformatting image, MPR) image shows diffuse encasement of the aorta and mild proximal right coronary artery stenosis (arrow) with encasing mildly enhancing soft tissue mass.

It has been suggested that the severity of IgG4-periadventitial inflammation may not necessarily parallel coronary artery luminal narrowing, as previous reports described cases of IgG4-related coronary periarteritis without causing even mild stenosis at the site of the coronary artery when surrounded by several periadventitial soft tissue cuffs (Fig. **2**) [8]. Stenosis can be detected easily by coronary artery CTA; however, invasive (conventional) coronary angiography is the gold standard technique for the final diagnosis of the severity of coronary stenosis and aneurysms. Coronary artery Stenosis can be detected easily by coronary artery CTA. Several expert consensus documents on the use of coronary CTA for excluding CAD in symptomatic patients with reference to many studies have reported high negative predictive value [9, 10], however, the predictive value depends heavily on disease prevalence, the prevalence of coronary artery stenosis in patients with IgG4 is unknown. Furthermore, coronary artery calcification may also alter the diagnostic performance of coronary CTA [11, 12], CTA is ineffective for ruling out obstructive CAD in patients with severe coronary artery calcification and in patients with high probability of CAD [13], another alternative for non-invasive test for assessment of CAD is the coronary MR angiography, Coronary MR angiography had a sensitivity of 78%-82% and a specificity of 915-96% in detection of coronary artery disease of at least 50% identified at conventional coronary angiography [14, 15]. However, a lengthy imaging time has been considered a major disadvantage of coronary MR angiography and the test was not completed in 8%-13% of patients with owing to drift of diaphragm position during imaging acquisition [16]. Subsequently, invasive (conventional) coronary angiography is the gold standard technique for the final diagnosis of the severity of coronary stenosis and aneurysms.

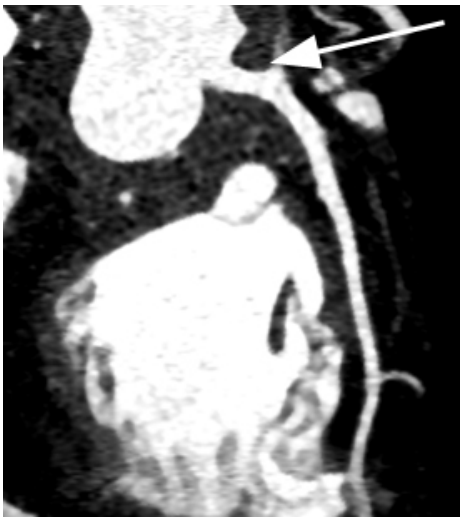

Fig. (2). MPR image of CT coronary angiogram encasement of the proximal aorta and left main coronary artery with soft tissue mass with no significant stenosis (arrow).

3. CORONARY ARTERY ANEURYSM

IgG4-related inflammation is known to play a role in arterial wall remodeling and aneurysmal formation. Ikutomi *et al*. reported a 75-year-old female with a history of autoimmune periarteritis diagnosed by elevated serum IgG4. Coronary angiography showed severe diffuse stenosis of the left anterior descending coronary artery, focal stenosis of the proximal left circumflex and an aneurysmal dilatation of the right coronary artery. CTA showed two focal tumoral lesions surrounding the middle and distal portions of the RCA [17]. Tanigawa *et al*. reported a smaller coronary artery aneurysmal dilatation in patients with clinically-diagnosed IgG4-RSD. More recently, Urabe *et al*. reported a case of acute myocardial infarction, which was caused by intra-coronary thrombosis at the site of a coronary aneurysm with a maximum diameter of 21mm. CTA showed that most of the epicardial coronary artery aneurysmal dilatation was surrounded by a tumor-like lesion [18]. Coronary CTA in patients with IgG4-related coronary artery disease is an excellent modality, as it can depict the spectrum of coronary artery disease in cases such as stenosis, soft tissue formation, tumor-like lesion, coronary ectasia and aneurysmal formation. Other imaging modalities for depiction of coronary artery aneurysms (CAAs) include coronary MR angiography, transthoracic echocardiography, and angiographic cardiac catheterization. Assessment of CAAs include evaluation of its shape and structure including morphology (fusiform, secular) aneurysmal dilation, wall calcification, luminal thrombosis, and any significant stenosis. The description should note whether the CAA is single or multiple as well as the name of precise segmental coronary artery involved [19]. Of all the different causes of CAAs, atherosclerotic disease is the most common cause resulting in at least 50% of CAAs in adults, and the most common cause of the CAAs in children is Kawasaki disease [20].

4. MYOCARDIAL LESIONS

Several case reports described that patients with IgG4-RSD may present with cardiac masses. Pathologically, IgG4-related cardiac tumors consist of spindle-shaped cells within a myxoid stroma. The stroma is infiltrated with lymphocytes, plasma cells and eosinophils, and IgG4-positive plasma cells are abundant. Kouzu and colleagues reported a case with a mass in the right ventricular outflow tract [21], and Ku-

sunose *et al*. reported a case with IgG4, which revealed a mass near the inferolateral wall of the left ventricle [22]. These masses were treated with corticosteroids, and some tumors were resected surgically. The diagnoses were established with echocardiography. CMR may identify masses more accurately than echocardiography and provide more reliable diagnoses. CMR is an important tool in the evaluation of cardiac masses, T1-weighted, T2-weighted, and gadolinium-enhanced sequence allowed for more tissue characterization [23], cine gradient echo image is used to assess functional effects of the masses [24]. Cardiac CT may be used to evaluate cardiac masses, particularly, when CMR is not available or contraindicated, Cardiac CT offers fast examination, high spatiotemporal resolution, and wide field of view and provides detailed evaluation of cardiac mass and adjacent structures [25].

5. PERICARDIAL LESIONS

IgG4-related constrictive pericarditis and IgG4-positive plasma cell infiltration into the pericardium are the underlying mechanisms reported in several cases, and patients may present with right-sided heart failure in advanced cases [26]. Morita *et al*. reported a case with pericardial effusion, where an open biopsy specimen from the pericardium revealed IgG4 positive plasma cells [27]. Mori *et al*. reported the case of a 65-year old female with pericardial involvement associated with autoimmune pericarditis. Chest CT imaging showed pericardial thickening, which resolved with corticosteroid therapy [28]. Pericardial and pleural effusion sometimes accompanies pericarditis, which could lead to a fatal outcome by causing pericardial tamponade or respiratory insufficiency. Echocardiography detects IgG4-related constrictive pericarditis as a highly echogenic thickening of the pericardium and pericardial effusion. CT and CMR imaging, as well as FDG PET/CT can also establish the diagnosis (Fig. **3**). All patients with IgG4-related constrictive pericarditis were treated with pericardiectomy, and in some cases, this was followed by corticosteroid therapy [29].

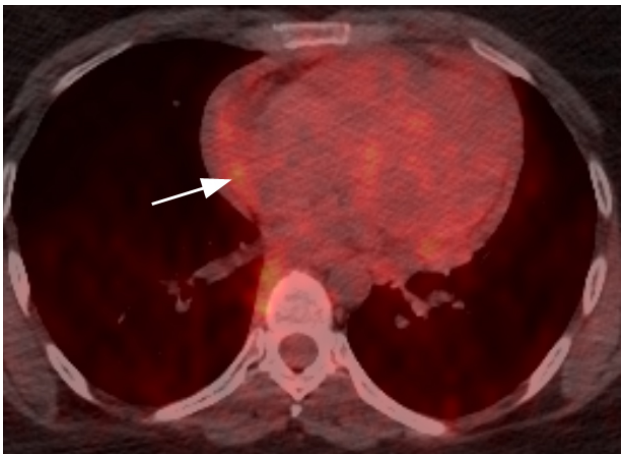

Fig. (3). Axial fused image of FDG PET-CT shows circumferential thinking of the pericardium with patchy area of FDG uptake (arrow), a finding that is consistent with mild pericardial effusion and acute pericarditis.

6. VALVULAR HEART LESIONS

Infiltration of IgG4-positive plasma cells often disturbs heart valve function causing stenosis or regurgitation. Maleszewski reported two cases of IgG4-related aortic stenosis, in which neither of the patients had any known non-cardiac manifestation of IgG4 at the time of surgery or any other explanation for the severe aortic stenosis, but both cases developed clinically silent IgG4 pancreatitis [30], Another case report by Yamauchi and colleagues described aortic regurgitation and heart block in a 59-year-old female. The excised lesion revealed a high ratio of IgG4-positive to IgG-positive plasma cells, providing a definitive diagnosis of IgG4 disease [31]. IgG4 infiltration sometimes forms a tumor on valve leaflets, and valve replacement has been selected as the most appropriate treatment. Echocardiography is the primary imaging modality for the diagnosis and management of patients with valvular heart disorders. Transthoracic echocardiography (TTE) and transoesophageal echocardiography (TEE) allow early and accurate diagnosis of valvular involvement severity (Fig. **4**). In the last 20 years, CMR has emerged as an alternative non-invasive modality without ionizing radiation to assess patient's valvular heart disease. CMR provides images of valve anatomy and allows quantitative evaluation of stenosis and regurgitation, in addition, CMR determines the consequences of the valve lesions, including ventricular dilatation, pressure overload, and alteration of systolic function [32].

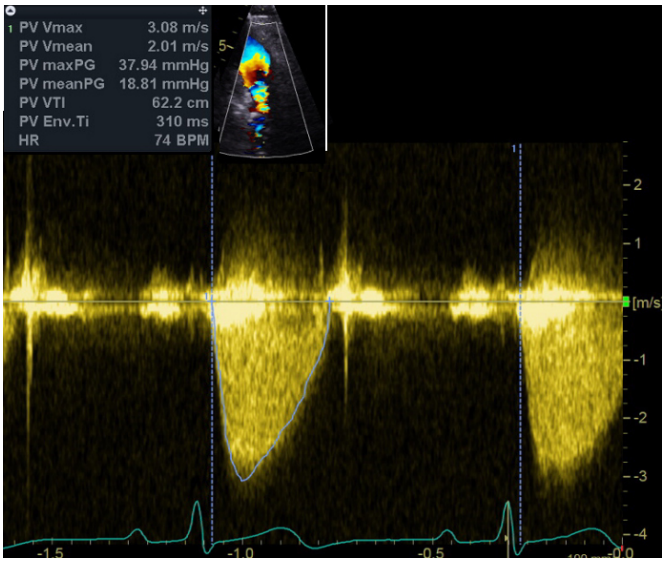

Fig. (4). Color flow Doppler of the pulmonary valve showed an area of systolic high-grade turbulence at the level of the pulmonary valve (small coloured insert), with a peak velocity of 3.08 meter/second and maximum pulmonary pressure gradient of 37.44 mm/HG consistent with moderate pulmonary valve stenosis. *(The color version of the figure is available in the electronic copy of the article).*

7. AORTIC LESIONS

The three pathologic diagnostic criteria for IgG4-related aortitis and periaortitis include: 1) An overall history consistent with aortitis and/or periaortitis not readily explained by

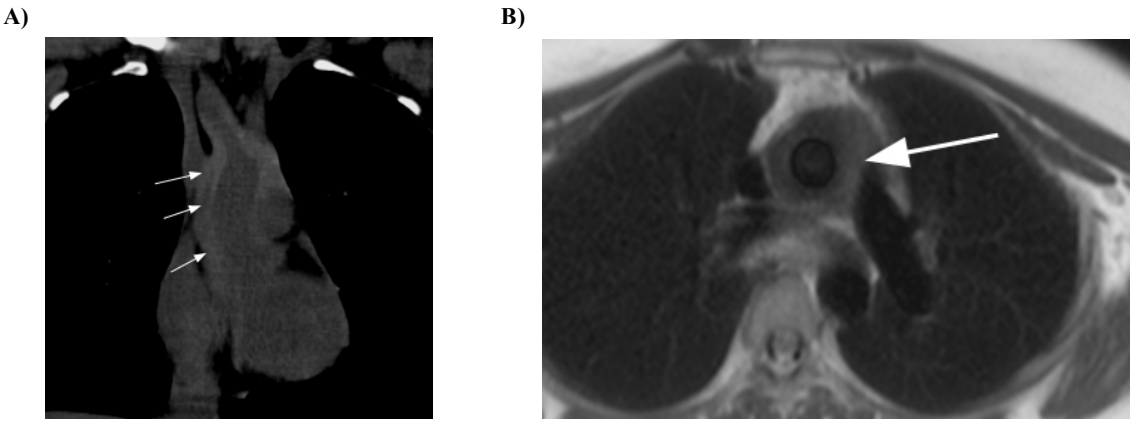

Fig. (5). A: Coronal non-contrast enhanced CT image of the chest demonstrates diffuse circumferential thickening of the wall of the ascending thoracic aorta (arrows). **B**: axial T1 weighted image MR image shows severe wall thickening of the ascending thoracic aorta (arrow).

another process, such as atherosclerosis; 2) At least 50% of the plasma cells stain for IgG4 and; 3) Plasma cells present in at least three fields magnified to at least 400x [33]. The three major components of chronic periaortitis are IgG4-related retroperitoneal fibrosis, IgG4-related abdominal aneurysm and IgG4-related perianeurysmal fibrosis [34]. IgG4-related periaortitis predominantly occurs in the infrarenal abdominal aorta and affects patients with later IgG4-RSD onset and those with highly active disease status. Periaortitis can progress to aortic rupture during or after corticosteroid therapy [35]. The clinical presentation of IgG4-related aortic disease can be very nonspecific, leading to diagnostic delay. Common symptoms are poorly localized lower back pain, lower abdominal and flank pain, leg edema and hydronephrosis from ureteral obstruction related to retroperitoneal fibrosis. Typically, CTA reveals vessel wall thickening and non-smooth soft tissue thickening around the aorta (Fig **5A**). Magnetic resonance angiogram (MRA) reveals vessel wall thickening and edema (Fig. **5B**). FDG PET/CT typically shows FDG uptake in the aortic wall during active inflammation (Fig. **6**). Nunez *et al*. reported a case with IgG4-associated ascending thoracic aortic aneurysmal changes. Biopsy of the periaortic mass detected plasma and other inflammatory cells and immunohistochemically demonstrated that more than 50% of the plasma cells were positive for IgG4 [36].

8. PULMONARY ARTERY STENOSIS

Pulmonary artery stenosis (Fig. **7**) and pulmonary hypertension have been reported in previous case studies. Perugino *et al*. reported a 69-year-old female who was presented with pulmonary hypertension diagnosed by echocardiogram. Chest CT revealed a 4 cm x 3 cm soft tissue mass encasing the left pulmonary artery, a soft tissue mass encasing the vocal cords and wall thickening of the thoracic aorta. On PET/CT, both soft tissue masses and the aortic wall were FDG-avid. The pulmonary artery involvement in IgG4-RSD is considered to be primarily a large vessel vasculitis [37]. Ishida *et al*. reported a 22-year-old woman diagnosed with IgG4-RSD who presented with pulmonary arterial hypertension (PAH) by echocardiography. This was the first case diagnosed with PAH related to IgG4 and was treated successfully with corticosteroids. The pathophysiological mechanism underlying the development of PAH remains unknown; however, it has been suggested that obliterative vasculitis of pulmonary arteries due to an immunological mechanism may have a role in the pathogenesis of PAH associated with IgG4-RSD [38].

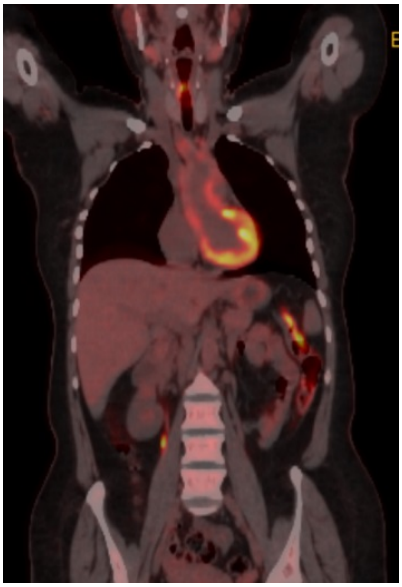

Fig. (6). Coronal fused FDG PET-CT image shows diffuse mild FDG uptake in the ascending thoracic aorta consistent with active inflammatory peri aortitis.

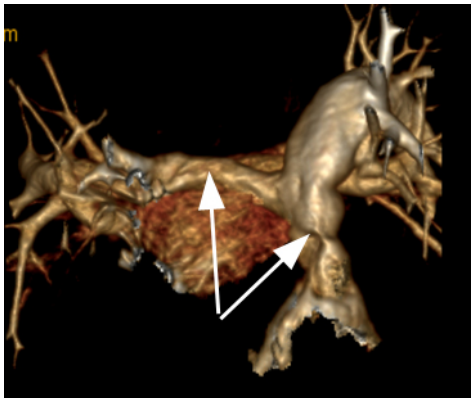

Fig. (7). Volume rendering image of the CT pulmonary angiogram shows severe narrowing of the proximal main and right pulmonary artery (arrows).

9. CAROTID ARTERIES

Carotid arteries involvement with IgG4-RSD has been reported in several reports. Perugino *et al.* reported a 67-year old man with a history of IgG4-RSD presented with recurrent episodes of right-sided amaurosis fugax, MRA revealed moderate to severe stenosis of the right common carotid artery [37]. The patient underwent successful carotid endarterectomy and pathological examination was consistent with focal involvement of IgG4-RSD. Barp A *et al.* reported a very unusual case of IgG4-RSD in a 67-year old man presented with carotid dissection and surgical exploration showed tight stenosis of the left internal carotid artery with large pseudoaneurysm. Histological examination was consistent with IgG4-RSD (Fig. **8**) [39]. demonstrates carotid Doppler ultrasound of a 61-year old female with IgG4-RSD with total occlusion of the left internal carotid artery.

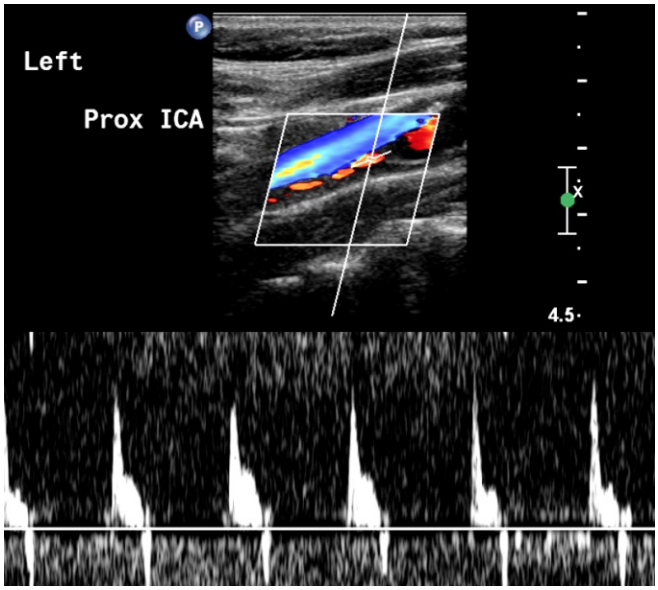

Fig. (8). Color Doppler ultrasound showed complete occlusion of the left internal carotid artery with the reversal of diastolic flow. *(The color version of the figure is available in the electronic copy of the article).*

10. PERIPHERAL ARTERIAL DISEASE

Compared to aortic disease, peripheral arterial diseases are very rarely detected; most of the morphological changes in IgG4-associated peripheral arterial diseases are aneurysmal changes. IgG4-related peripheral arterial lesions display pathological features similar to other IgG4-related vascular lesions. Aneurysmal changes have been reported in renal, splenic, femoral and popliteal arteries [40-42]. In addition to aortic and peripheral arterial disease, small vessel systemic vasculitis has been reported with IgG4-RSD [43].

SUMMARY

IgG4-RSD is a fibro-inflammatory disorder that can affect any organ in the body. The various cardiovascular clinical manifestations of the disease may include coronary artery stenosis, aneurysm, myocardial diseases, pericarditis and pericardial effusion, valvular heart disease, aortic, pulmonary and peripheral vascular disease. Different imaging techniques, including echocardiography, CT, CMR, and FDG PET/CT have been used in the management of patients with IgG4-RSD. The role of diagnostic imaging includes establishing the diagnosis, detecting complications, guiding biopsy, and documenting response to therapy.

Echocardiography is the most commonly used method of assessing cardiac function, valvular heart involvement and other cardiac involvements such as pericardium and myocardium. Coronary CT angiography is the most reliable non-invasive technique for assessing coronary artery stenosis, aneurysms and aortic and peripheral vascular disease. CMR offers a comprehensive evaluation of the cardiovascular system including cardiac function, the extent of fibrosis, disease-related complications, and a guide to further treatment. FDG PET/CT can provide important information about the severity of the disease, the presence of active inflammation and the optimum biopsy site.

CONSENT FOR PUBLICATION

Not applicable.

FUNDING

None.

CONFLICT OF INTEREST

The author declares no conflict of interest, financial or otherwise.

ACKNOWLEDGEMENTS

Declared none.

REFERENCES

[1] Hamano H, Kawa S, Horiuchi A, *et al*. High serum IgG4 concentrations in patients with sclerosing pancreatitis. N Engl J Med 2001; 344(10): 732-8.

[2] Khosroshahi A, Deshpande V, Stone JH. The clinical and pathological features of igg(4)-related disease. Curr Rheumatol Rep 2011; 13(6): 473-81.

[3] James OG, Christensen JD, Wong TZ, Borges-Neto S, Koweek LM. Utility of FDG PET/CT in inflammatory cardiovascular disease. Radiographics 2011; 31(5): 1271-86.

[4] Kasashima S, Kawashima A, Endo M, *et al*. A clinicopathologic study of immunoglobulin G4-related disease of the femoral and popliteal arteries in the spectrum of immunoglobulin G4-related periarteritis. J Vasc Surg 2013; 57(3): 816-22.

[5] Agaimy A, Weyand M, Strecker T. Inflammatory thoracic aortic aneurysm (lymphoplasmacytic thoracic aortitis): A 13-year-experience at a German Heart Center with emphasis on possible role of IgG4. Int J Clin Exp Pathol 2013; 6(9): 1713-22.

[6] Keraliya AR, Murphy DJ, Aghayev A, Steigner ML. IgG4-related disease with coronary arteritis. Circ Cardiovasc Imaging 2016; 9(3): e004583.

[7] Matsumoto Y, Kasashima S, Kawashima A, *et al*. A case of multiple immunoglobulin G4-related periarteritis: A tumorous lesion of the coronary artery and abdominal aortic aneurysm. Hum Pathol 2008; 39(6): 975-80.

[8] Urabe Y, Fujii T, Kurushima S, Tsujiyama S, Kihara Y. Pigs-in-a-blanket coronary arteries: A case of immunoglobulin G4-related coronary periarteritis assessed by computed tomography coronary angiography, intravascular ultrasound, and positron emission tomography. Circ Cardiovasc Imaging 2012; 5(5): 685-7.

[9] Taylor AJ, Cerqueira M, Hodgson JM, *et al*. ACCF/SCCT/ACR/AHA/ASE/ASNC/NASCI/SCAI/SCMR 2010 appropriate use criteria for cardiac computed tomography. A report of the American College of Cardiology Foundation Appropriate Use Criteria Task Force, the Society of Cardiovascular Computed Tomography, the American College of Radiology, the American Heart Association, the American Society of Echocardiography, the American Society of Nuclear Cardiology, the North American Society for Cardiovascular Imaging, the Society for Cardiovascular Angiography and Interventions, and the Society for Cardiovascular Magnetic Resonance. J Am Coll Cardiol 2010; 56(22): 1864-94.

[10] Budoff MJ, Achenbach S, Blumenthal RS, *et al*. Assessment of coronary artery disease by cardiac computed tomography: A scientific statement from the American Heart Association Committee on Cardiovascular Imaging and Intervention, Council on Cardiovascular Radiology and Intervention, and Committee on Cardiac Imaging, Council on Clinical Cardiology. Circulation 2006; 114(16): 1761-91.

[11] Pundziute G, Schuijf JD, Jukema JW, *et al*. Impact of coronary calcium score on diagnostic accuracy of multislice computed tomography coronary angiography for detection of coronary artery disease. J Nuclear Cardiol 2007; 14(1): 36-43.

[12] Ong TK, Chin SP, Liew CK, *et al*. Accuracy of 64-row multidetector computed tomography in detecting coronary artery disease in 134 symptomatic patients: Influence of calcification. Am Heart J 2006; 151(6): 1323.e1-6.

[13] Miller JM, Rochitte CE, Dewey M, *et al*. Diagnostic performance of coronary angiography by 64-row CT. N Engl J Med 2008; 359(22): 2324-36.

[14] Weber OM, Martin AJ, Higgins CB. Whole-heart steady-state free precession coronary artery magnetic resonance angiography. Magn Reson Med 2003; 50(6): 1223-8.

[15] Sakamoto A, Nagai R, Saito K, *et al*. Idiopathic retroperitoneal fibrosis, inflammatory aortic aneurysm, and inflammatory pericarditis--retrospective analysis of 11 case histories. J Cardiol 2012; 59(2): 139-46.

[16] Jahnke C, Paetsch I, Nehrke K, *et al*. Rapid and complete coronary arterial tree visualization with magnetic resonance imaging: Feasibility and diagnostic performance. Eur Heart J 2005; 26(21): 2313-9.

[17] Ikutomi M, Matsumura T, Iwata H, *et al*. Giant tumorous lesions (correction of legions) surrounding the right coronary artery associated with immunoglobulin-G4-related systemic disease. Cardiology 2011; 120(1): 22-6.

[18] Tanigawa J, Daimon M, Murai M, Katsumata T, Tsuji M, Ishizaka N. Immunoglobulin G4-related coronary periarteritis in a patient presenting with myocardial ischemia. Hum Pathol 2012; 43(7): 1131-4.

[19] Jeudy J, White CS, Kligerman SJ, *et al*. Spectrum of coronary artery aneurysms: From the radiologic pathology archives. Radiographics 2018; 38(1): 11-36.

[20] Cohen P, O'Gara PT. Coronary artery aneurysms: A review of the natural history, pathophysiology, and management. Cardiol Rev 2008; 16(6): 301-4.

[21] Kouzu H, Miki T, Mizuno M, *et al*. Inflammatory myofibroblastic tumor of the heart. Overlap with IgG4-related disease? Circ J 2014; 78(4): 1006-8.

[22] Kusunose K, Hotchi J, Takagawa Y, *et al*. Serial imaging changes during treatment of immunoglobulin G4-related disease with multiple pseudotumors. Circulation 2015; 131(21): 1882-3.

[23] Patel R, Lim RP, Saric M, *et al*. Diagnostic performance of cardiac magnetic resonance imaging and echocardiography in evaluation of cardiac and paracardiac masses. Am J Cardiol 2016; 117(1): 135-40.

[24] Sparrow PJ, Kurian JB, Jones TR, Sivananthan MU. MR imaging of cardiac tumors. Radiographics 2005; 25(5): 1255-76.

[25] Rajiah P, Kanne JP, Kalahasti V, Schoenhagen P. Computed tomography of cardiac and pericardiac masses. J Cardiovasc Comput Tomogr 2011; 5(1): 16-29.

[26] Sugimoto T, Morita Y, Isshiki K, *et al*. Constrictive pericarditis as an emerging manifestation of hyper-IgG4 disease. Int J Cardiol 2008; 130(3): e100-1.

[27] Morita T, Izawa A, Hamano H, *et al*. Significant pericardial involvement of immunoglobulin G4-related disease. Ann Thorac Surg 2014; 98(2): e47-9.

[28] Mori K, Yamada K, Konno T, *et al*. Pericardial involvement in IgG4-related disease. Int Med (Tokyo, Japan) 2015; 54(10): 1231-5.

[29] Misawa Y. Immunoglobulin G4-related cardiovascular diseases. Ann Thorac Cardiovasc Surg 2017; 23(6): 281-5.

[30] Maleszewski JJ, Tazelaar HD, Horcher HM, *et al*. IgG4-related disease of the aortic valve: A report of two cases and review of the literature. Cardiovasc Pathol 2015; 24(1): 56-9.

[31] Yamauchi H, Satoh H, Yamashita T, *et al*. Immunoglobulin G4-related disease of the heart causing aortic regurgitation and heart block. Ann Thorac Surg 2013; 95(6): e151-3.

[32] Cawley PJ, Maki JH, Otto CM. Cardiovascular magnetic resonance imaging for valvular heart disease: Technique and validation. Circulation 2009; 119(3): 468-78.

[33] Stone JR. Aortitis, periaortitis, and retroperitoneal fibrosis, as manifestations of IgG4-related systemic disease. Curr Opin Rheumatol 2011; 23(1): 88-94.

[34] Ozawa M, Fujinaga Y, Asano J, *et al*. Clinical features of IgG4-related periaortitis/periarteritis based on the analysis of 179 patients with IgG4-related disease: A case-control study. Arthritis Res Ther 2017; 19(1): 223.

[35] Kasashima S, Kawashima A, Kasashima F, *et al*. Immunoglobulin G4-related periaortitis complicated by aortic rupture and aortoduodenal fistula after endovascular AAA repair. J Endovasc Ther 2014; 21(4): 589-97.

[36] Nunez Fernandez MJ, Fachal Bermudez C, Duran Munoz D, *et al*. IgG4-related disease and isolated thoracic aortic aneurysm. VASA Zeitschrift fur Gefasskrankheiten 2014; 43(5): 385-9.

[37] Perugino CA, Wallace ZS, Meyersohn N, Oliveira G, Stone JR, Stone JH. Large vessel involvement by IgG4-related disease. Medicine 2016; 95(28): e3344.

[38] Ishida M, Miyamura T, Sato S, Kimura D, Suematsu E. Pulmonary arterial hypertension associated with IgG4-related disease. Intern Med (Tokyo, Japan) 2014; 53(5): 493-7.

[39] Barp A, Fedrigo M, Farina FM, *et al*. Carotid aneurism with acute dissection: An unusual case of IgG4-related diseases. Cardiovasc Pathol 2016; 25(1): 59-62.

[40] Sarac M, Marjanovic I, Bezmarevic M, Zoranovic U, Petrovic S, Mihajlovic M. An aortoduodenal fistula as a complication of immunoglobulin G4-related disease. World J Gastroenterol 2012; 18(42): 6164-7.

[41] Danlos FX, Daoued-Keffi F, Rohmer J, *et al*. IgG4-related disease associated with renal microaneurysms and polycythaemia. Rheumatology (Oxford, England) 2016; 55(2): 380-2.

[42] Tajima M, Hiroi Y, Takazawa Y, *et al*. Immunoglobulin G4-related multiple systemic aneurysms and splenic aneurysm rupture during steroid therapy. Hum Pathology 2014; 45(1): 175-9.

[43] Alba MA, Milisenda J, Fernandez S, *et al*. Small-vessel vasculitis with prominent IgG4 positive plasma cell infiltrates as potential part of the spectrum of IgG4-related disease: A case report. Clin Exp Rheumatol 2015; 33(2 Suppl 89): S138-41.