Right Atrial Mass with Inflow Obstruction in a patient with IgG4-related Disease

Shek Joyce
Department of Medicine and Geriatrics, Princess Margaret Hospital, HKSAR, shekjoycesht@gmail.com

Wu Lok Yi
Department of Medicine and Geriatrics, Princess Margaret Hospital, HKSAR

Wong Chun Kit
Department of Medicine and Geriatrics, Princess Margaret Hospital, HKSAR

Lu Jianlin
Department of Medicine and Geriatrics, Princess Margaret Hospital, HKSAR

Lam Wai Lung William
Department of Medicine and Geriatrics, Princess Margaret Hospital, HKSAR

See next page for additional authors

Follow this and additional works at: https://www.jhkcc.com.hk/journal

Part of the Cardiology Commons, Cardiovascular Diseases Commons, and the Medical Education Commons

Recommended Citation

This Case Report is brought to you for free and open access by Journal of the Hong Kong College of Cardiology. It has been accepted for inclusion in Journal of the Hong Kong College of Cardiology by an authorized editor of Journal of the Hong Kong College of Cardiology.
Right Atrial Mass with Inflow Obstruction in a patient with IgG4-related Disease

Authors
Shek Joyce, Wu Lok Yi, Wong Chun Kit, Lu Jianlin, Lam Wai Lung William, Mak Wai Man Vivien, and Chan Ngai Yin

This case report is available in Journal of the Hong Kong College of Cardiology: https://www.jhkcc.com.hk/journal/vol29/iss2/7
CASE REPORT

Right Atrial Mass with Inflow Obstruction in a Patient with IgG4-Related Disease


Department of Medicine and Geriatrics, Princess Margaret Hospital, Hong Kong, China

Abstract

Cardiac tumour is a rare cause of heart failure. We report a case of right atrial mass found in a patient with known immunoglobulinG4-related disease (IgG4-RD), who presented with right heart failure, renal impairment and thrombocytopenia. Multimodal investigations were performed to reach the diagnosis of diffuse large B-cell lymphoma with cardiac involvement. Chemotherapy was initiated in an attempt for disease control. We highlight the potential link between IgG4-related disease and lymphoma.

Keywords: Cardiac tumor, IgG4 related disease, Cardiac lymphoma, Diffuse large B cell lymphoma, Heart failure, Endomyocardial biopsy

Case report

An 80-year old Chinese male was admitted for acute heart failure. He had history of stable chronic obstructive pulmonary disease, Kimura’s disease, IgG4-related disease with chronic lymphadenopathy and IgA-lambda paraproteinemia.

He first presented with groin lymphadenopathy 19 years ago in 2003. Excisional biopsy showed reactive changes only. Subsequently, he developed a left lacrimal gland tumour and right submandibular mass in 2005. Surgical excision of both masses revealed the diagnosis of Kimura’s disease. In 2013, he was noted to have rising globulin level, from a baseline of 45 g/L to 54 g/L. Further investigation revealed polyclonal hyper gammaglobulinemia and the presence of an IgA-lambda paraprotein measuring 7 g/L, not associated with systemic manifestations of multiple myeloma. CT thorax in 2015 showed enlarged right hilar and mediastinal lymph nodes but biopsy showed no malignancy. He was followed up for interval monitoring of symptoms and paraprotein level.

He developed further enlargement of neck masses in 2016. Excisional biopsy of the left submandibular lymph node showed probable IgG4-related lymphadenopathy, supported by the histological features of a plasma-cell-rich chronic inflammatory infiltrate associated with fibrosis, increased number of IgG4+ plasma cells up to 100/hpf, and increased IgG4/IgG ratio >40% by immunostaining. Together with an elevated serum IgG4 level at 17 g/L and clinical presentation, the diagnosis of IgG4-related disease was made. His cervical lymphadenopathy became progressively symptomatic and he was started on steroid therapy since 2017. He responded well clinically and serologically with IgG4 dropped to 2.8 g/L. He was maintained on low dose prednisolone 7.5 mg daily since 2017 with satisfactory symptom control.

At the age of 80, he presented with repeated admissions within 2 months for recurrent heart failure.
symptoms, including exertional dyspnoea, orthopnoea and lower limb oedema, suboptimally relieved by diuretics. Lymphadenopathy was not apparent upon this presentation. Echocardiogram showed a large heterogeneous mass in the right atrium (RA), which measured 5 cm across, occupying most of the dilated right atrium with extension to the inferior vena cava (IVC) (Figure 1). There was congested IVC with flow acceleration seen across the mass, causing a right ventricular inflow obstruction with a mean gradient of 4 mmHg (Figure 2). There was no pericardial effusion. Computed tomography of the thorax, abdomen and pelvis showed an intracardiac mass up to 7.5 cm over the right atrium, as well as prominent mediastinal, hilar and perigastric lymph nodes up to 2.4 cm. Biochemically, he developed new-onset progressive renal impairment, with baseline creatinine level of 100 umol/L rising to 190 umol/L, as well as deteriorating thrombocytopenia from a baseline platelet count of 127 × 10^9/L to 45 × 10^9/L. Of note, the levels of both IgA-lambda paraprotein and IgG4 remained stable at 6 g/L and 5.823 g/L, respectively. Contrast injection in the IVC revealed large filling defect in RA (Figure 3).

Decision-making

The differential diagnoses at this stage included multiple myeloma with extramedullary plasmacytoma involving the heart, cardiac involvement by IgG4-RD, or primary cardiac tumours like lymphoma, atrial myxoma, or cardiac angiosarcoma. Multi-disciplinary approach was adopted in the decision-making process, involving input from cardiologists, haematologists and radiologists. Bone marrow examination showed a normocellular marrow and mild plasmacytosis without light chain restriction, excluding multiple myeloma as the cause of renal impairment and thrombocytopenia. In view of the rare involvement of heart by IgG4-RD and severity of the presentation, histologic evaluation of the mass was deemed necessary for a diagnosis that could guide definitive treatment. We subsequently performed biopsy to the RA mass under intracardiac echocardiography (ICE) guidance. Histology came back to be diffuse large B cell lymphoma. Reduced intensity chemotherapy was started in view of the patient’s impaired cardiac and renal function as well as advanced age. Unfortunately, he could not tolerate chemotherapy and succumbed during the first cycle.

Discussion

IgG4-related disease has only been identified within this century [1], and our understanding and knowledge of this disease continue to evolve. Given its frequent multi-organ involvement and common laboratory findings of eosinophilia and polyclonal hypergammaglobulinemia, it has many mimickers...
like multicentric Castleman disease, lymphoma, plasma cell neoplasms, and hypereosinophilic syndromes, which may represent a diagnostic challenge both for the physician and the pathologist [2]. The development of haematologic complications in the setting of IgG4-RD requires a high index of suspicion, as illustrated in this case.

We report a patient who developed diffuse large B-cell lymphoma 6 years after the diagnosis of well-controlled IgG4-related disease. To date, the number of similar published cases in both Asian and Western series, of IgG4-RD patients who subsequently developed haematological malignancies, is less than 50 [3,4]. The most commonly-reported lymphoma that develop in the setting of IgG4-RD were diffuse large B-cell lymphoma as well as extranodal marginal zone lymphoma, yet follicular lymphoma and peripheral T-cell lymphoma have also been reported [3]. It has also been postulated that diffuse large B-cell lymphoma could represent clonal transformation from an underlying low-grade B-cell lymphoma co-existing with but undiagnosed during the time of IgG4-related disease [3].

The possible aetiologic mechanism underlying the lymphomagenesis of IgG4-related disease has not been well-delineated, but it is known that chronic inflammation predisposes to lymphoma [5]. A retrospective study also reported that patients with IgG4-RD have an increased incidence of non-Hodgkin lymphoma (standardized incidence ratio of 400 [95% CI 109-1024]) [6]. More epidemiological studies are needed to demonstrate this association.

In summary, we report a rare case of cardiac lymphoma presenting with heart failure in a patient with IgG4-related disease. Physicians should be alerted to this potential but rare possibility during the evaluation of a patient with unexplained acute heart failure with underlying IgG4-related disease.
Ethics information
Not applicable.

Acknowledgements and funding
None.

Conflict of interest
None declared.

References


