

# Mediastinal Mass in a Patient with IgG4-Related Disease

Kesato Iguchi<sup>1</sup>, Osamu Ishibashi<sup>1</sup>, Norio Takayashiki<sup>2</sup>, Hiroaki Satoh<sup>3</sup>

<sup>1</sup>Division of Surgery, <sup>2</sup>Division of Pathology and <sup>3</sup>Division of Respiratory Medicine, Mito Medical Center, University of Tsukuba

## Article Information

Received: 09 Nov 2015

Accepted: 01 Dec 2015

Plagiarism software: Turnitin

## Keywords:

Mediastinal mass,  
IgG4-related disease,  
Mediastinal tumor



Hiroaki Satoh

## ABSTRACT

**Introduction:** IgG4-related disease is now considered to result from an imbalance in the regulatory immune system, and it is diagnosed on the basis of clinical and histopathologic evidence such as the presence of lymphoplasmacytic infiltrates, especially increased IgG4-positive plasma cells and fibrotic sclerosing tissue.

**Case report:** We report herein a 45-year-old male patient with mediastinal mass with IgG4-related disease. IgG4-related disease can involve any organ but the most frequently involved sites include pancreato-hepatobiliary tract, salivary glands, and kidneys, generally manifesting as a mass in one or more sites mimicking a neoplasia. In the thoracic region, IgG4-related disease manifests as interstitial lung disease and fibrosing mediastinitis. Mediastinal lesion in our patient was quite resemble to mediastinal tumor derived from anterior mediastinum. This is the first case with mediastinal mass in a patient with this disease.

**Conclusion:** Although very rare, we should rule out this disease as differential diagnosis in patients with anterior homogeneous density mediastinal mass simulating mediastinal tumor.

## INTRODUCTION

IgG4-related disease can involve multiple organs, particularly exocrine organs such as pancreas, salivary glands, and biliary tract.<sup>1,2</sup> When it affects the thorax, it commonly manifests interstitial lung disease and fibrosing mediastinitis.<sup>3-11</sup> We present herein a case of IgG4-related disease mimicking as mediastinal tumor in the anterior mediastinum.

## CASE REPORT

A 45-year-old-man was admitted to our hospital due to incidentally detected mediastinal mass, which was not found in chest radiograph a year before. The patient had 5-year history of bronchial asthma. His serum

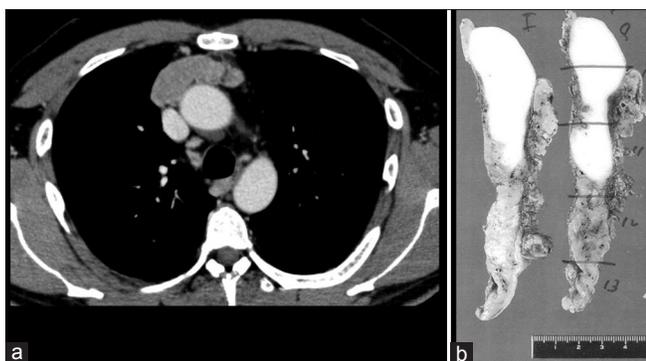
immunoglobulin level, including igG4, was slightly elevated (114 mg/dL); normal range, 4.8-105 mg/d). The results of other biochemical tests were normal. Chest CT scan revealed a well-defined homogenous anterior mediastinal mass (Figure 1a). As the mass was located at anterior mediastinum, we suspected thymoma, germ cell tumor, or lymphoma. Although the patient had no symptom, video-assisted thoracic surgery (VATS) was planned extended thymectomy with combined resection of the surrounding involved structures. After carrying out thoracotomy, we found a hen's egg-sized mass with smooth surface in anterior mediastinum. Adhesion of the mass to the surrounding tissue was not observed, but left internal thoracic artery and vessel were involved. The tumor was completely excised (Figure 1b). Pathological specimen obtained from VATS showed storiform pattern of fibrosis and immunohistochemical staining for IgG4 positive cells (Figure 2a and b). Pathological diagnosis was IgG4-related disease. Postoperative course was not remarkable. Thereafter, oral prednisolone was prescribed for 18 months (tapered off from an initial dose of 40mg/day). The patient was followed up for 2 years and had no mass lesion including mediastinum.

## Access this article online

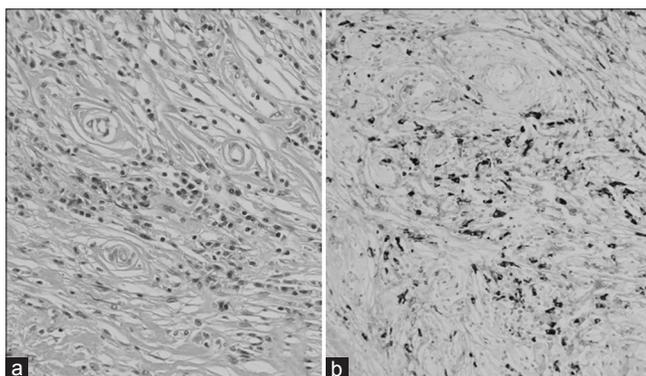
Website:	Quick Response code
www.actamedicainternational.com	
DOI: 10.5530ami.2016.1.44	

## Corresponding Author:

Hiroaki Satoh, MD, PhD, Division of Respiratory Medicine, Mito Medical Center, University of Tsukuba, Miya-machi 3-2-7, Mito, Ibaraki, 310-0015, Japan. Tel: +81-29-231-2371. E-mail: hirosato@md.tsukuba.ac.jp



**Figure 1:** Chest CT scan revealed a well-defined homogenous anterior mediastinal mass (a), Macroscopic view of completely resected a hen's egg-sized mass with smooth surface in anterior mediastinum (b)



**Figure 2:** Pathological specimen obtained from video-assisted thoracic surgery showed storiform pattern of fibrosis (a) and immunohistochemical staining for IgG4 positive cells (b)

## DISCUSSION

Although the precise mechanisms are still unclear, IgG4-related disease is now considered to result from an imbalance in the regulatory immune system.<sup>12,13</sup> IgG4-related disease develops predominantly in males.<sup>2,9</sup> In majority of patients, IgG4-related disease is diagnosed on the basis of clinical and histopathologic evidence such as the presence of lymphoplasmacytic infiltrates, especially increased IgG4-positive plasma cells and fibrotic sclerosing tissue.<sup>2,9</sup> As for treatment, IgG4-related disease has been shown to respond favorably to corticosteroid.<sup>14</sup> Azathioprine or rituximab is now useful in cases of relapse or failure to wean off corticosteroid.<sup>2,9</sup>

IgG4-related disease can involve virtually any organ but the most frequently involved sites include the pancreas, bile ducts and gallbladder, liver, salivary glands, and kidneys generally manifesting as a mass in one or more sites mimicking a neoplasia.<sup>2,9</sup> In the thoracic region, IgG4-related disease manifests as interstitial lung disease and fibrosing mediastinitis.<sup>9-11</sup> There have been three large prospective cohort of patients with IgG4-related disease. In a Japanese study of 114 patients with IgG4-related disease, 26 (22.8%) of

them had pulmonary involvement and only 1 (0.9%) of them had mediastinal fibrosis.<sup>10</sup> In a French study of 25 patients with IgG4-related disease, 3 (12%) of them had pulmonary involvement and none of them had mediastinal fibrosis.<sup>11</sup> In a recent Chinese study of 118 patients with IgG4-related disease, 32 (2%) of them had pulmonary involvement and only 4 (3.4%) of them had mediastinal fibrosis.<sup>9</sup> To our best knowledge, however, mediastinal mass associated with this disease at mediastinum has not been previously reported. Therefore, this is the first case with mediastinal mass in patient with this disease.

Mediastinal lesion in our patient was quite resemble to mediastinal tumor derived from anterior mediastinum. Differential diagnosis of anterior mediastinal mass includes thymoma, germ cell tumor, and lymphoma.<sup>15,16</sup> Neurogenic tumors are atypical with regard to location. Thymoma appears as homogeneous mass and germ cell tumor usually appears as heterogenous mass with calcification in enhanced CT scan. Taking the location and density of the mass into consideration, we preoperatively evaluated it was thymoma derived from anterior mediastinum.

We present herein a case of IgG4-related disease mimicking as mediastinal tumor in the anterior mediastinum. Although very rare, we should rule out this disease as differential diagnosis in patients with anterior homogenous density mediastinal mass, mimicking mediastinal tumor.

## REFERENCES

1. Brito-Zeron P, Ramos-Casals M, Bosch X, Stone JH. The clinical spectrum of IgG4-related disease. *Autoimmun Rev* 2014; 13: 1203-10.
2. Islam AD, Selmi C, Datta-Mitra A, et al. The changing faces of IgG4-related disease: Clinical manifestations and pathogenesis. *Autoimmun Rev* 2015 Jun 23. pii: S1568-9972(15)00134-2.
3. Shigemitsu H, Koss MN. IgG4-related interstitial lung disease: a new and evolving concept. *Curr Opin Pulm Med* 2009; 15: 513-6.
4. Miyashita T, Yoshioka K, Nakamura T, et al. A case of lymphomatoid granulomatosis-like lung lesions with abundant infiltrating IgG4-positive plasma cells whose serum IgG4 levels became high following the start of corticosteroid therapy. *Intern Med* 2010; 49: 2007-11.
5. Dias OM, Kawassaki Ade M, Haga H, Cukier A, Carvalho CR. Immunoglobulin G4-related systemic sclerosing disease in a patient with sclerosing cholangitis, inflammatory pseudotumors of the lung and multiple radiological patterns: a case report. *Clinics (Sao Paulo)* 2011; 66: 1983-6.
6. Yi ES, Sekiguchi H, Peikert T, Ryu JH, Colby TV. Pathologic manifestations of immunoglobulin (Ig)G4-related lung disease. *Semin Diagn Pathol* 2012; 29: 219-25.
7. Yamashita H, Takahashi Y, Ishiura H, Kano T, Kaneko H, Mimori A. Hypertrophic pachymeningitis and tracheobronchial stenosis in IgG4-related disease: case presentation and literature review. *Intern Med* 2012; 51: 935-41.
8. Kitada M, Matuda Y, Hayashi S, et al. IgG4-related lung disease showing high standardized uptake values on FDG-PET: report of two cases. *J Cardiothorac Surg* 2013; 8: 160.

9. Lin W, Lu S, Chen H, et al. Clinical characteristics of immunoglobulin G4-related disease: a prospective study of 118 Chinese patients. *Rheumatology (Oxford)* 2015 Jun 22. pii: kev203
10. Zen Y, Nakanuma Y. IgG4-related disease: a cross-sectional study of 114 cases. *Am J Surg Pathol* 2010; 34: 1812-9.
11. Ebbo M, Daniel L, Pavic M, et al. IgG4-related systemic disease: features and treatment response in a French cohort: results of a multicenter registry. *Medicine* 2012; 91: 49-56.
12. Umehara H, Nakajima A, Nakamura T, et al. IgG4-related disease and its pathogenesis-cross-talk between innate and acquired immunity. *Int Immunol* 2014; 26: 585-95.
13. Smit W, Barnes E. The emerging mysteries of IgG4-related disease. *Clin Med* 2014; 14 Suppl 6: s56-60.
14. Campbell SN, Rubio E, Loschner AL. Clinical review of pulmonary manifestations of IgG4-related disease. *Ann Am Thorac Soc* 2014; 11: 1466-75.
15. Carter BW, Okumura M, Detterbeck FC, Marom EM. Approaching the patient with an anterior mediastinal mass: a guide for radiologists. *J Thorac Oncol* 2014; 9(9 Suppl 2): S110-18.
16. Motus IY, Bazhenov AV, Massard G. Surgery for huge mediastinal tumors. *Asian Cardiovasc Thorac Ann* 2015; 23: 846-50.

**How to cite this article:** Iguchi K, Ishibashi O, Takayashiki N, Satoh H. Mediastinal Mass in a Patient with IgG4-Related Disease. *Acta Medica International*. 2016;3 (1):216-218.

**Source of Support:** Nil, **Conflict of Interest:** None declared.