International Journal of
Case Reports & Short Reviews

Case Report

Pulmonary Rosai-Dorfman Disease with Feature of IgG4 Related Disease: a Case Report in Taipei Veterans General Hospital

Chia Tsung Hung[1]*, Yu Chung Wu[1] and Yi Chen Yeh[2]

[1]Division of Thoracic Surgery, Department of Surgery, Taiwan
[2]Department of Pathology and Laboratory Medicine, Taipei Veterans General Hospital, Taipei, Taiwan

***Address for Correspondence:** Chia Tsung Hung, Division of Thoracic Surgery, Department of Surgery, Taiwan, Tel: +886-932-064-257; ORCiD: https://orcid.org/0000-0002-2490-4472; E-mail: john770614@gmail.com

Submitted: 17 June 2019; **Approved:** 26 June 2019; **Published:** 27 June 2019

Cite this article: Hung CT, Wu YC, Yeh YC. Pulmonary Rosai-Dorfman Disease with Feature of IgG4 Related Disease: a Case Report in Taipei Veterans General Hospital. Int J Case Rep Short Rev. 2019;5(6): 041-043.

ISSN: 2638-9355

BACKGROUND

Rosai-Dorfman Disease (RDD), also known as Sinus Histiocytosis with Massive Lymphadenopathy (SHML), is a rare nonmalignant histioproliferative disorder [1,2], first described by Rosai and Dorfman in 1969 [3]. It affects not only lymph nodes but also other organs. It can mimic lung carcinoma and shows increased activity on the whole-body PET scan [4].

CASE PRESENTATION

A 53-year-old nonsmoking female patient had no underlying disease. She had received regular health check-ups since 2016, and annual chest radiographs reported one enlarging lesion over right lower lung field (Figure 1). She had no specific symptom except occasional cough. She visited our out-patient department. Chest Computed Tomography (CT) scan was arranged, and one soft-tissue nodule was found over right lower lobe of lung (Figure 2). Another protruding mass with overlying irregular mucosa from the RB8 orifice leading to 80% occlusion was noted by bronchoscopy (Figure 3). Bronchoscopic biopsy of the endobronchial lesion was done by interventional pulmonologist, and pathology report showed chronic inflammation. CT-guided biopsy of the lung nodule was not feasible because the tumor was too close to blood vessels. Due to rapid progression of the lesions and the possibility of malignancy that could not be ruled out, surgical biopsy was arranged. Owing to the central location of the lesions, Video-Assisted Thoracoscopic Surgery (VATS) for RLL lobectomy with radical lymph node dissection was done in 2018. Intraoperative specimen revealed one intraparenchymal tumor and the other endobronchial tumor, causing obstructive pneumonitis. Surgical pathology disclosed Rosai-Dorfman disease with features of IgG_4 related disease (Figure 4). Harvested lymph nodes were all benign anthracosis. Postoperative serum levels of IgG and IgG_4 were within normal range.

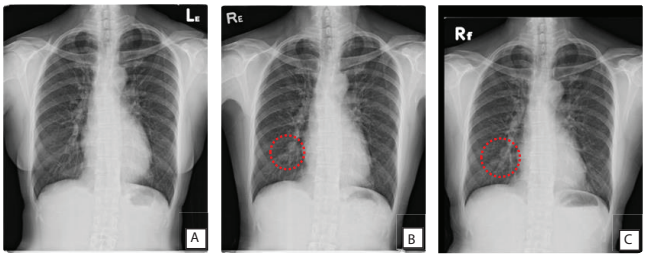

Figure 1: The 3 chest radiographs were taken in 2016, 2017, and 2018 respectively. (A) Clean right lung field;
(B) Pulmonary nodules appeared in the right lower lung field; (C) Enlarged pulmonary nodules

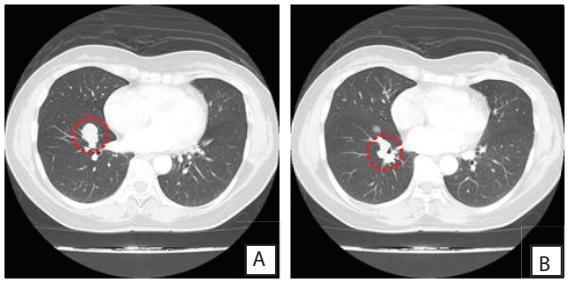

Figure 2: Chest CT scan: (A) RLL intraparenchymal tumor; (B) Endobronchial tumor

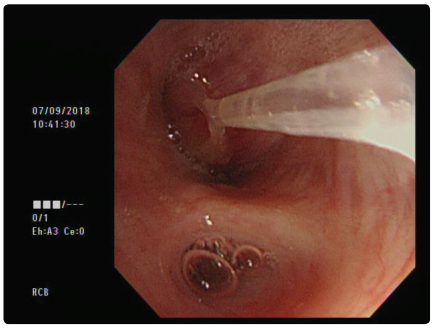

Figure 3: Bronchoscopy showed one protruding mass with overlying irregular mucosa from the RB8 orifice leading to 80% occlusion. Biopsy was done

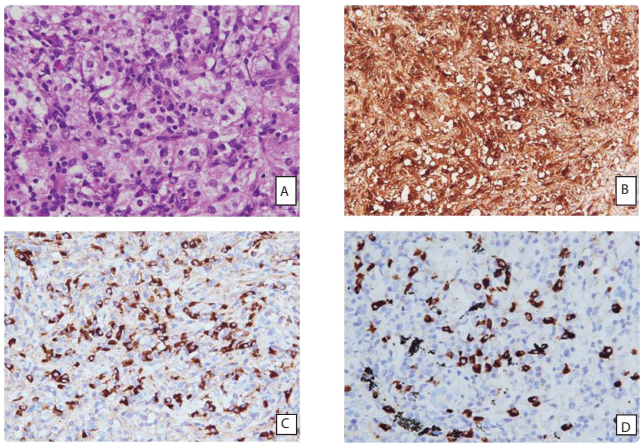

Figure 4: Sections of the both tumors show intensive infiltration of mixed population of lymphocytes and many plasma cells with lymphoid follicle formation, and marked fibrosis in storiform pattern. (A) Large histiocytes with abundant pale eosinophilic cytoplasm and emperipolesis are seen; (B) These large histiocytes are positive for S100 stain; (C) Many plasma cells are positive for IgG stain; (D) Many plasma cells are positive for IgG4 stain

DISCUSSION

Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, typically manifests as painless, massive lymphadenopathy that frequently affects cervical lymph nodes and is accompanied by fever, leukocytosis, increased erythrocyte sedimentation rate, and hypergammaglobulinemia [5]. Extranodal involvement has been reported in up to 43% of cases including skin, soft tissue, nasal cavity, bone, orbit, intrathoracic structures, central nervous system, breast, urogenital tract, and gastrointestinal tract [4,6-14], and 23% of cases had extranodal involvement only [1]. Pulmonary involvement is rare and occurs approximately in only 3% of cases with extranodal disease [1].

IgG_4-Related Disease (IgG_4-RD) is a pathological entity recently recognized by the medical world that can affect any organ or system [15], including the pancreas, bile ducts, eyes, salivary glands, lungs, heart, kidneys, skin, aorta, ganglia, meninges, prostate, breast, thyroid, retroperitoneal tissue, etc [16]. The basics of this condition have begun to be constructed since 2003, when patients with autoimmune pancreatitis have also been observed to have extrapancreatic manifestations [15]. The histopathological examination is considered the gold standard in obtaining a diagnosis [17]. The histological criteria are: diffuse lymphoplasmacytic infiltrate, numerous IgG_4 positive

plasma cells in the examined tissue, storiform fibrosis (resembling the spokes of a cartwheel), eosinophils in mild to moderate quantities, obliterative phlebitis, and pseudotumoral lesions that tend to form in the affected organs [18]. A wrong diagnosis of IgG_4-RD is commonly encountered because overestimation is given to the role of serum or tissue IgG_4 (up to 30% of the patients may have normal IgG_4 values) [19].

El-Kersh K, et al. [20] reported a 76-year-old African-American male patient presenting with interstitial lung involvement without lymphadenopathy. The patient underwent video-assisted thoracoscopy with right upper lobe wedge biopsy, with the diagnosis of RDD. Notably, a significant proportion of plasma cells were IgG_4 positive. Within 1 year the patient developed generalized lymphadenopathy. Despite trials of corticosteroid therapy, the patient's symptoms progressed within a few months. The patient elected hospice care where he passed away.

Our patient did not have lymphadenopathy as well. We originally planned to do wedge resection in order to obtain specimens for frozen control for tissue proof. However, the tumor was centrally located, making it difficult for wedge resection only. As a result, lobectomy was performed. Surgical pathology showed Rosai-Dorfman disease with features of IgG_4-related disease. Though lobectomy might be considered overtreatment, the obstructive pneumonitis observed in the specimens made lobectomy much more reasonable. It was reported that corticosteroid could be used to treat both Rosai-Dorfman disease [20] and IgG_4-related disease [17]. It is worth mentioning that, although there is a decrease in IgG_4 concentration after the start of corticosteroid treatment, serum levels remain high in most patients. Only 30% of the cases with persistent elevated IgG_4 levels had relapses. Also, 10% of the patients with normal IgG_4 levels also experienced recurrences [21]. Now our patient kept follow-ups at the out-patient department. Serum levels of IgG and IgG_4 mildly elevated, but there was no specific symptom or sign, and no medication is needed.

Rosai-Dorfman disease is a rare disease. It is scarcer when combined with the feature of IgG_4-related disease. Elevated serum level of IgG_4 might be a hint for disease recurrence or progression, or IgG_4-related disease in other organ. Long-term follow-up of the patient is required.

REFERENCE

1. El Kersh K, Perez RL, Guardiola J. Pulmonary IgG4+ Rosai-Dorfman disease. BMJ Case Rep. 2013. https://bit.ly/2Fvma96
2. Apperley ST, Hyjek EM, Musani R, Thenganatt J. Intrathoracic Rosai-Dorfman disease with focal aggregates of IgG4-bearing plasma cells. Case report and literature. Ann Am Thorac Soc. 2016; 13: 666-670. https://bit.ly/2Yc19aN
3. Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. Arch Pathol. 1969; 87: 63-70. https://bit.ly/2RygfF1
4. Ali A, Mackay D. Rosai-Dorfman disease of the lung. Thorax. 2009; 64: 908-909. https://bit.ly/2JkyES7
5. Dalia S, Sagatys E, Sokol L, Kubal T. Rosai-Dorfman disease: tumor biology, clinical features, pathology, and treatment, Cancer Control. 2014; 21: 322-327. https://bit.ly/2J8GC0j
6. Anders RA, Keith JN, Hart J. Rosai-Dorfman disease presenting in the gastrointestinal tract. Arch Pathol Lab Med. 2003; 127: 74-75. https://bit.ly/2X8Jpk9
7. Hida AI, Yagi SI, Obase Y, Nishimura H, Akiyama T, Irei I, et al. Rosai-Dorfman disease presenting as a solitary mediastinal mass. Pathol Int.2009; 59: 265-268. https://bit.ly/31RdyTE
8. La Barge DV 3rd, Salzman KL, Harnsberger HR, Ginsberg LE, Hamilton BE, Wiggins RH 3rd, et al. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): imaging manifestations in the head and neck. AJR Am J Roentgenol. 2008; 191: 299-306. https://bit.ly/2Loaybo
9. Lao IW, Dong Y, Wang J. Rosai-Dorfman disease of the pericardium: a case report and review of literature. Int J Clin Exp Pathol. 2014; 7: 3408-3412. https://bit.ly/2X5qfqi
10. Ma H, Zheng Y, Zhu G, Wu J, Lu C, Lai W. Rosai-Dorfman disease with massive cutaneous nodule on the shoulder and back. Ann Dermatol. 2015; 27: 71-75. https://bit.ly/2KC8PQn
11. O'Gallagher K, Dancy L, Sinha A, Sado D. Rosai-Dorfman disease and the heart. Intractable Rare Dis Res. 2016; 5: 1-5. https://bit.ly/2Ne8LIR
12. Paryani NN, Daugherty LC, O'Connor MI, Jiang L. Extranodal Rosai-Dorfman disease of the bone treated with surgery and radiotherapy. Rare Tumors. 2014; 6: 132-134. https://bit.ly/2FyezXf
13. Pham CB, Abruzzo LV, Cook E, Whitman GJ, Stephens TW. Rosai-Dorfman disease of the breast. American Journal of Roentgenology. 2005; 185: 971-972.
14. Walters DM, Dunnington GH, Dustin SM, Frierson HF, Peeler BB, Kozower BD et al. Rosai-Dorfman disease presenting as a pulmonary artery mass. Ann Thorac Surg. 2010; 89: 300-302. https://bit.ly/2LiKJto
15. Stone JH, Zen Y, Deshpande V. IgG4-related disease. N Engl J Med. 2012; 366: 539-51. https://bit.ly/2NcTUOI
16. Umehara H, Okazaki K, Masaki Y, Kawano M, Yamamoto M, Saeki T et al. A novel clinical entity, IgG4-related disease (IgG4RD): general concept and details. Mod Rheumatol. 2012; 22: 1-14. https://bit.ly/2YfT3Oa
17. Opriţa R, Opriţa B, Berceanu D, Diaconescu IB. Overview of IgG4 - Related Disease. J Med Life. 2017; 10: 203-207. https://bit.ly/2RzQVhT
18. Deshpande V, Zen Y, Chan JK, Yi EE, Sato Y, Yoshino T, et al. Consensus statement on the pathology of IgG4-related disease. Mod Pathol. 2012; 25: 1181-1192. https://bit.ly/2IMsTgT
19. Sah RP, Chari ST. Serologic issues in IgG4-related systemic disease and autoimmune pancreatitis. Curr Opin Rheumatol. 2011; 23: 108-113. https://bit.ly/2J7SM9I
20. AL Umairi R, Blunt D, Hana W, Cheung M, Oikonomou A. Rosai-Dorfman Disease: Rare Pulmonary Involvement Mimicking Pulmonary Langerhans Cell Histiocytosis and Review of the Literature. Case Rep Radiol. 2018; 2018: 2952084. https://bit.ly/2WZNiTy
21. Kamisawa T, Shimosegawa T, Okazaki K, Nishino T, Watanabe H, Kanno A, et al. Standard steroid treatment for autoimmune pancreatitis. Gut. 2009; 58: 1504-1507. https://bit.ly/2IN4U13