Case Report

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

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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 IgG4 related disease (Figure 4). Harvested lymph nodes were all benign anthracosis. Postoperative serum levels of IgG and IgG4 were within normal range.

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].

IgG4-Related Disease (IgG4-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 IgG4 positive
plasma cells in the examined tissue, storiform fibrosis (resembling the spokes of a cartwheel), eosinophils in mild to moderate quantities, obliterator phlebitis, and pseudotumoral lesions that tend to form in the affected organs [18]. A wrong diagnosis of IgG₄-RD is commonly encountered because overestimation is given to the role of serum or tissue IgG₄ (up to 30% of the patients may have normal IgG₄ 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 IgG₄ 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 IgG₄-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 IgG₄-related disease [17]. It is worth mentioning that, although there is a decrease in IgG₄ concentration after the start of corticosteroid treatment, serum levels remain high in most patients. Only 30% of the cases with persistent elevated IgG₄ levels had relapses. Also, 10% of the patients with normal IgG₄ levels also experienced recurrences [21]. Now our patient kept follow-ups at the out-patient department. Serum levels of IgG and IgG₄ 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 IgG₄-related disease. Elevated serum level of IgG₄ might be a hint for disease recurrence or progression, or IgG₄-related disease in other organ. Long-term follow-up of the patient is required.

REFERENCE