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Imaging Findings in IGG4 Disease with Isolated Thoracic Involvement- A Case Report

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Abstract

Immunoglobulin G4 - related disease (IgG4-RD) is a multisystemic fibro inflammatory disorder that can involve multiple organs and organ systems of the body. While autoimmune pancreatitis and retroperitoneal fibrosis are the most described forms of this entity, a few patients may present with thoracic involvement, either in isolation or more commonly, in association with abdominal and other systemic manifestations. Thoracic presentation of the disease can be in the form of lung parenchymal, interstitial, mediastinal vasculature and pleural involvement. In the mediastinal vasculature, thoracic aorta is the most commonly involved vessel; involvement of the pulmonary artery is less frequently described in literature. We present the imaging findings of an unusual case of IgG4-RD with isolated thoracic involvement in a middle-aged male patient who presented with shortness of breath.Contrast enhanced CT of the chest revealed a poorly marginated mediastinal mass causing encasement and narrowing of the superior vena cava, encasement and cut-off of the right pulmonary artery, with extension of the soft tissue along the right peribronchial interstitium.A solid nodule and mild interstitial changes were also seen in the right lung as part of the disease spectrum.

Keywords: ***IgG4 related disease; Mediastinal fibrosis; Pulmonary artery Stenosis; Peri bronchial soft tissue***

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1. Abbreviations

IgG4-RD: Immunoglobulin G4 related disease; SVC: Superior Vena Cava; ESR: Erythrocyte Sedimentation Rate; EBUS: Endo-bronchial ultrasound; MIP: Maximum Intensity Projection; HRCT: High resolution CT

2. Introduction

IgG4-RD is a set of immune-mediated multisystemic fibro inflammatory disorder having similar cytological findings and pathogenesis [1]. It can affect almost every system in the body, mimicking malignant, infectious, and inflammatory disorders [2]. The most common presentations of IgG4-RD include autoimmune pancreatitis, sclerosing cholangitis, sialadenitis, orbital region disease, retroperitoneal fibrosis, and chronic periaortitis [2]. Thoracic manifestations of IgG4-RD are less common and can present in the form of pulmonary involvement or involvement of mediastinal vasculature [3-5]. While pulmonary lesions can present as lung masses, groundglass opacities, interstitial involvement and peribronchial lesions, mediastinal vascular involvement can be in the form of arteritis or perivascular soft tissue, occasionally leading to vessel stenosis or aneurysm.

3. Clinical Presentation and Investigations

A 46-year-old male presented to the department of pulmonology with complains of mild dyspnoea on exertion of 1 month duration with intermittent low-grade fever. There was no history of cough or weight loss. In view of increased severity of the dyspnoea in the last 3 days, a CT- pulmonary angiogram with HRCT of the chest was performed.

The images revealed a poorly marginated hypodense and mildly enhancing soft tissue in the mediastinum in the right lower paratracheal and hilar region, extending to the subcarinal region, causing encasement and complete cut-off of the right pulmonary artery (FIG. 1 & 2). Collateral vessels from the bronchial artery and intercostal vessels were seen in the soft tissue, reforming the lobar branches of the right pulmonary artery (FIG. 2). The soft tissue was also seen causing encasement and narrowing of the distal SVC (FIG. 2), further extending along the right peri-bronchial interstitium (FIG. 1 & 3) to cause mild narrowing of the right main bronchus. Interstitial septal thickening was seen in the right middle lobe; the right lower lobe showed a 11 mm peripheral nodule with partially spiculated margins (FIG. 3). Mosaic perfusion was noted in bilateral lung parenchyma.

The diagnosis of mediastinal fibrosis was made, and a probable aetiology of tuberculosis was considered. As there were no necrotic mediastinal lymph nodes and no history of cough, possibility of idiopathic fibrosing mediastinitis was also considered in the differential diagnosis. ESR was normal and Montoux test was negative. An attempted biopsy of the mass through EBUS guidance had to be abandoned because of haemorrhage from the multiple collateral arteries within the soft tissue. Serology revealed elevated serum IgG4 levels at 580 mg/dl. In view of the classic radiological findings and serological findings, the diagnosis of possible IgG4-RD was considered based on the Japan guidelines on IgG4-RD [6]. CT scan of the abdomen revealed no focus of involvement either in the pancreas or in the retroperitoneum. The patient was put on steroids and had significant clinical improvement in symptoms. Follow up imaging done after 6 months revealed mild interval reduction in the extent of

mediastinal soft tissue and in the peribronchial involvement (FIG. 2 and 3). There was also reduction in the serum IgG4 levels to 120 mg/dL post treatment.

The patient was on a tapering dose of steroids. During the subsequent follow-up visit, in view of significant clinical improvement, further imaging was not performed.

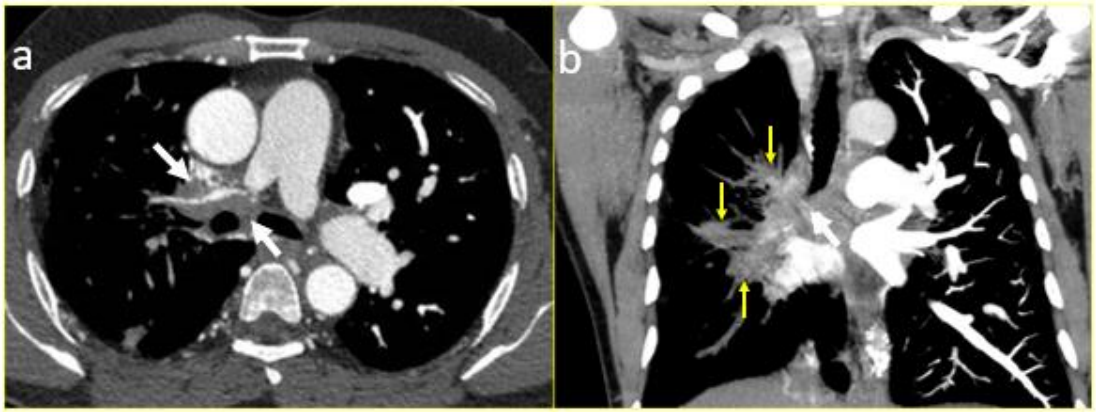

FIG. 1. **Axial image (a) and Coronal MIP image (b) of CT Pulmonary angiogram show encasement and complete cut off of the right pulmonary artery by a soft tissue mass in the right lower paratracheal and hilar region (white arrows in a and b). The soft tissue also extends into the right peri-bronchial region (yellow arrows in b).**

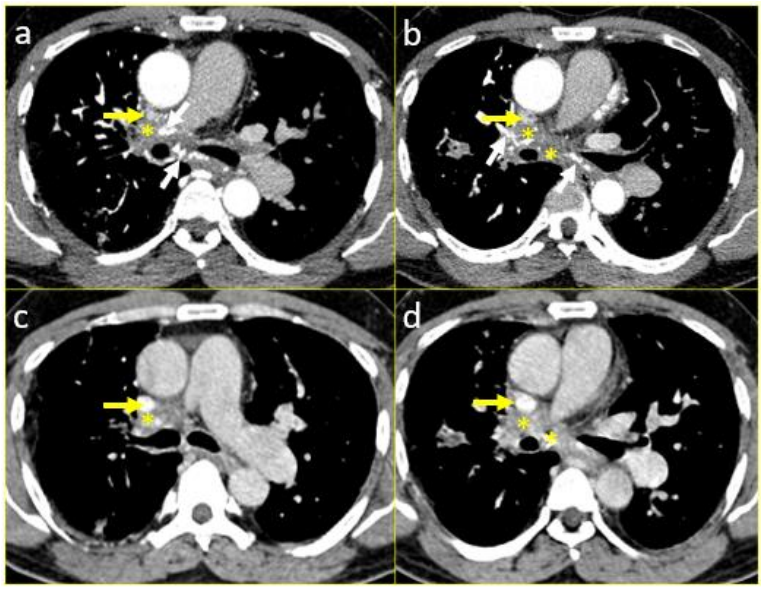

FIG. 2. **Axial arterial phase CT images at diagnosis (a and b) show the soft tissue extending from the right hilar to subcarinal region (yellow Asterix in a and b) causing encasement of the SVC with its narrowing (yellow arrows in a and b). Also seen are collaterals from the bronchial arteries reforming the right pulmonary divisions (white arrow in a and b). Post-treatment images (c and d) show mild reduction in the extent of the soft tissue (yellow Asterix in c and d). There is also reduction in the extent of compression of SVC with improved calibre (yellow arrow in c and d).**

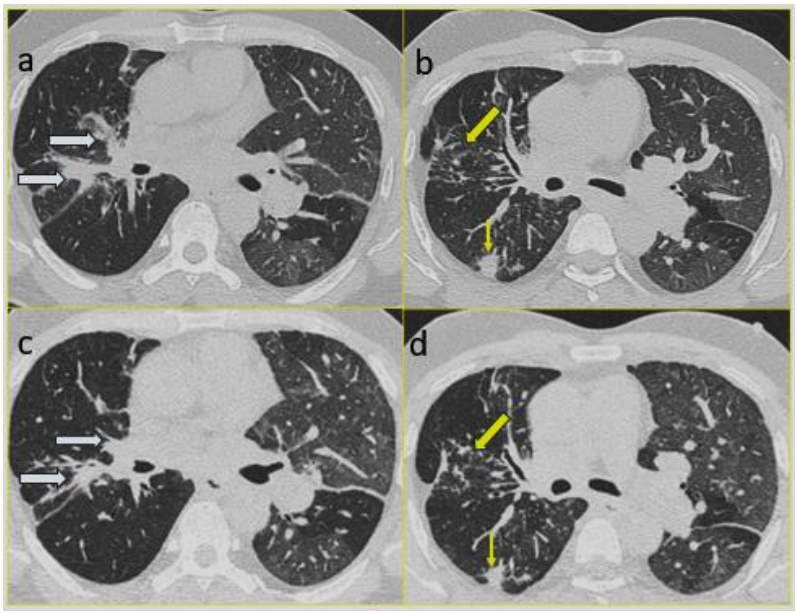

FIG. 3. **Axial images in lung window at 2 different levels (a and b) demonstrate peri-bronchial soft tissue (White arrows in a). Also seen are areas of interstitial thickening in the right middle lobe (large yellow arrow in b) and a peripheral nodule with partially spiculated margins in the right lower lobe (small yellow arrow in b). Post-treatment images at the same levels as Figures 3a and b (c and d) demonstrate mild interval reduction in the extent of peribronchial soft tissue (white arrows in c). The interstitial thickening is persistent (large yellow arrow in d), however, there is mild interval reduction in the size of the peripheral solid nodule in the right lower lobe (small yellow arrow in d).**

4. Discussion

IgG4-RD is a multisystemic fibro inflammatory disorder characterised by inflammation with or without tumefaction and fibrosis, often accompanied by raised serum IgG4 levels. The characteristic histopathological features include a lymphoplasmacytic infiltrate, storiform fibrosis, obliterative phlebitis and dramatic IgG4 plasma cell infiltrates [6]. Though histology is desirable for diagnosis, in a subset of patients with classical clinical, serological or radiological findings, diagnosis can be made in the absence of biopsy [6,7].

Lung involvement in IgG4 related disease presents in four different forms: Peribronchial thickening, which is the most common presentation; the others being the solid nodule type, round-shaped groundglass opacity and alveolar interstitial type presenting radiologically as interstitial pneumonia [1,8]. Three of these classic manifestations i.e., peri bronchial thickening, interstitial involvement and solid nodule were present in this patient. Though none of these findings in isolation are specific for IgG4 related disease, the presence of two or more findings, especially in the presence of multisystem disease, should be considered as favourable for diagnosis of IgG4 related disease [1]. Patients with thoracic involvement may have no symptoms or may present with mild symptoms of shortness of breath.

Pulmonary vascular manifestations of this disease are rare and may present on imaging as arteritis with or without aneurysm and pulmonary artery stenosis [2]. The imaging findings in contrast enhanced CT are vessel wall thickening and enhancement in arteritis and in the form of perivascular soft tissue and mediastinal fibrosis causing none to severe luminal narrowing. The

presence of pulmonary artery stenosis may lead to pulmonary hypertension [4,5,9]. Retro-mediastinal fibrosis, paravertebral soft tissue, peri aortitis and coronary arteritis are the other described extrapulmonary thoracic manifestations [1,10]. Thoracic manifestations of IgG4 related disease are seldom seen in isolation; they are most often seen together with some form of extrathoracic manifestation such as retroperitoneal or pancreatic involvement [11]. Our case was an unusual presentation of isolated thoracic involvement with no clinical or imaging signs of either pancreatic or retroperitoneal involvement on imaging.

5. Conclusion

IgG4 related disease can occasionally present with isolated thoracic manifestations. Awareness of the classic pulmonary, mediastinal and vascular imaging findings of this entity are important to guide the clinician towards diagnosis and treatment.

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