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Possible IgG4-related respiratory disease (IgG4-RRD) - a rare presentation as a Pancoast tumour-like

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We declare that the patient approved the case report publishing by signing a written informed consent form. The case report followed the ethical guidelines established by the Declaration of Helsinki.

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Abstract

IgG4-related respiratory disease (IgG4-RRD) is a recently recognized disease that can have multiple presentations. In many cases, the exclusion of malignancy is essential. We present a case of a male patient with right omalgia and supraclavicular swelling. Radiologic exams showed a lung mass suggestive of a Pancoast tumour in the right upper lobe. Patient underwent transthoracic biopsies that revealed fibromuscular tissue with fibrosis, lymphoplasmacytic infiltrate and a significant increase in IgG4 cells with 15% IgG4/IgG ratio. He started on prednisolone with clinical and radiologic improvement. As it is a heterogeneous disease it is important that clinicians are aware of the possible presentations.

Keywords: Immunoglobulin G4-Related Disease; Pancoast; Lung mass.

Introduction

IgG4-RRD is a recently recognized immune-mediated and usually multiorgan disease that presents many times as a tumorous lesion [1-3]. It may affect the lungs in different forms but can emerge as a pulmonary pseudotumour [2, 4].

The exclusion of a neoplastic process is fundamental to the correct diagnosis of a IgG4-RRD [2]. To the best knowledge of the authors, the initial presentation similar to a Pancoast tumour has not yet been described. The purpose of this case report is to improve scientific knowledge about the possible presentations of this rare disease.

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Case report

A fifty-two years old, non-smoker and not exposed to second hand tobacco smoke, male patient with a personal history of gastritis medicated with Omeprazole, presented right omalgia and supraclavicular swelling.

An ultrasound of the right shoulder was performed, which revealed cuff tendinosis and a heterogeneous hypoechoic area posterior to the pectoralis major muscle.

He performed a chest Computed Tomography (CT) for further clarification which revealed a tumour mass suggestive of a Pancoast tumour in the right upper lobe measuring 46x34mm. In Figure 1 we can see the lung mass on a CT scan performed just before the first transthoracic biopsy.

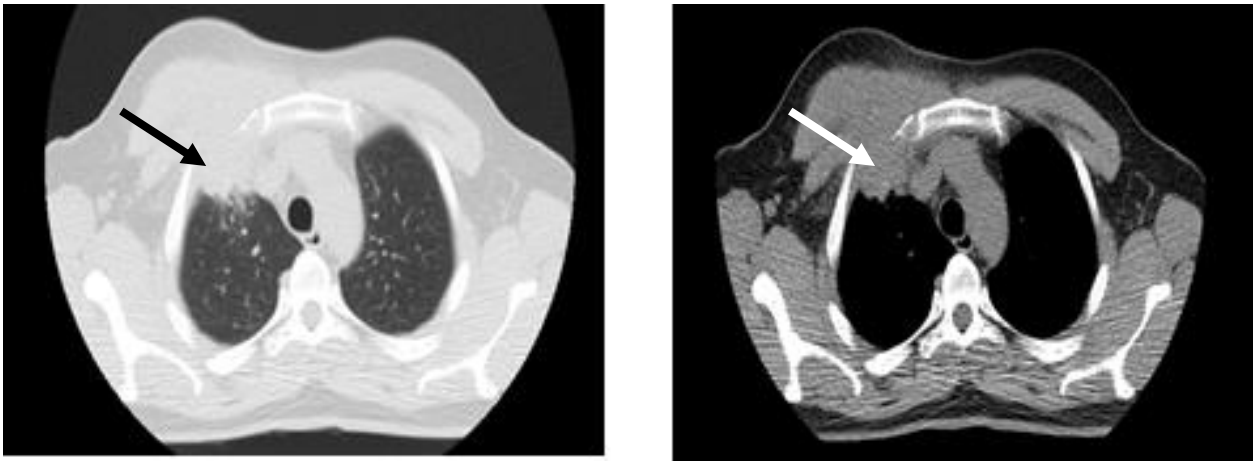

Figure 1. CT image (performed just before the first transthoracic biopsy) of lung mass suggestive of Pancoast tumour (arrow) in pulmonary and mediastinal window.

An 18-fluorodeoxyglucose positron emission tomography (18-FDG PET) was performed, showing high 18-FDG uptake (maximum SUV 16.6) in the identified mass extending to the soft tissues of the chest wall and to the right hilar and right paratracheal ganglia.

Due to high suspicion of lung cancer presenting as a Pancoast tumour, it was performed two transthoracic biopsies at different times that revealed fibromuscular tissue with fibrosis, lymphoplasmacytic infiltrate and a 15% IgG4/IgG ratio in the first biopsy and a significant increase in IgG4 cells (63 cells/HPF) in the second one. These findings were suggestive of IgG4-RRD.

He also performed a chest Magnetic Resonance Imaging (MRI) that revealed an ovoid lesion in the right upper lobe measuring 40x30 mm with signs of invasion of the anterior chest wall. Abdominopelvic CT showed no signs of other organs involvement.

Immunological study revealed increased sedimentation velocity (62mm/h), slightly increased Immunoglobulin G (1490mg/dl) with increased Immunoglobulin G4 (247mg/dl) which established an IgG4/IgG serum ratio of 16.6%.

Patient was referred to Rheumatology, and initiated prednisolone 40 mg/day (0.5 mg/kg/day) with clinical improvement, decrease in the size of the lesion (Figure 2) and decrease in IgG4 serologic values.

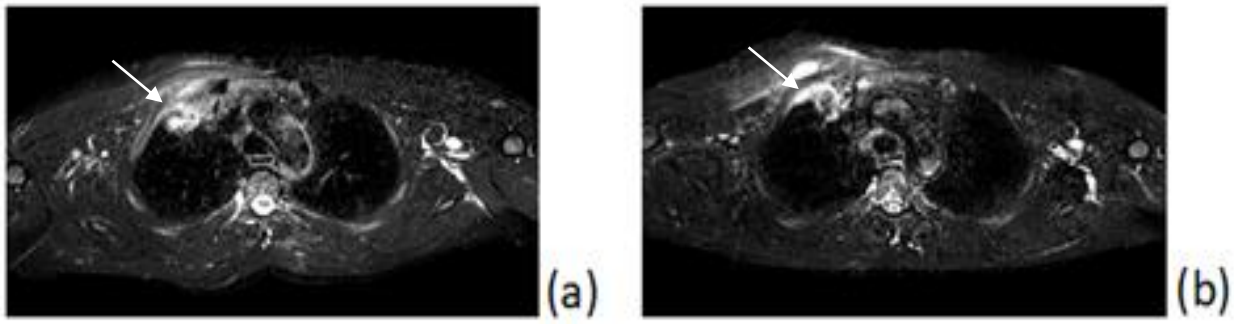

Figure 2. MRI identifying lung mass (arrow) before (a) and after (b) treatment

There was a relapse with worsening of the pain and swelling with the tapering of the Prednisolone dose. After relapse it was performed a third biopsy to confirm the diagnosis that revealed muscular tissue with sclerosing areas and inflammatory infiltrate with > 10 IgG4 cells/HPF, which was also suggestive of IgG4-RRD. Corticosteroid therapy was increased.

After that, patient was hospitalized because of a bacteraemia due to *S. aureus* methicillin sensitive, probably in the context of systemic corticosteroid therapy and biopsy puncture. This hospitalization was complicated with a Stevens-Johnson syndrome, which resolved after receiving immunoglobulins and corticosteroids. After hospitalization, patient clinically improved, allowing prednisolone tapering.

Currently patient maintains stable, with no recurrence of pain. MRI shows stability of the size of the thoracic lesion (30 mm) and patient remains under surveillance, medicated with 20 mg prednisolone/day.

Discussion and Conclusion

The clinical presentation of IgG4-RRD is highly variable [2-3]. When this disease presents as a pulmonary nodule or mass, the exclusion of lung cancer is essential [2-3]. It is not possible to differentiate IgG4-RRD and lung cancer only by CT, MRI or 18-FDG PET [3]. Thus, the diagnosis usually requires lung biopsy [3].

The increase in serum IgG4 is not diagnostic but is suggestive of this disease, usually with IgG4 > 135 mg/dl [4-5] and serum IgG4/IgG ratio >10 [2], which was found in our patient. The histopathological features of IgG4-RRD are a lymphoplasmacytic infiltrate, fibrosis and obliterative phlebitis [1-2, 4]. Most authors defend the presence of >10 IgG4 /HPF and an IgG4/IgG ratio > 40% in tissue sample to make the diagnosis [2, 4, 5].

In our case, the patient had >10 IgG4/HPF but not a ratio >40%. However, the clinical presentation suggestive of IgG4-RRD, the increase in serum IgG4, the absence of another diagnosis to explain the changes found, and the clinical and imaging initial response to corticosteroid therapy allowed us to make a presumptive diagnosis of this pathology, despite not meeting all the criteria.

The treatment of IgG4-RRD is based on corticosteroid therapy with good initial response but with a relapse of 40-76% of patients, usually during tapering of the corticosteroid dose [2-3]. The use of glucocorticoid sparing therapies is still being studied [2-3]. Due to lack of long-term studies of these patients, long-term prognosis cannot be established. However, it is known that, without treatment, this disease can be progressive and cause considerable morbidity and mortality [2].

In conclusion, IgG4-RRD is a recently identified disease whose diagnosis is often difficult with a highly variable clinical presentation. When this disease presents as a pulmonary nodule or mass, the exclusion of cancer is fundamental [2-3]. Improving knowledge about its possible presentations is essential for clinicians to be alert and suspicious of this condition.

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