



A great masquerader disease...

Thibault Maillet, Mathilde Funes de La Vega, Céline Duperron, Sylvain Audia

► To cite this version:

Thibault Maillet, Mathilde Funes de La Vega, Céline Duperron, Sylvain Audia. A great masquerader disease.... European Journal of Internal Medicine, 2020, 78, pp.133 - 134. <10.1016/j.ejim.2020.06.034>. <hal-03491445>

HAL Id: hal-03491445

<https://hal.science/hal-03491445v1>

Submitted on 22 Aug 2022

HAL is a multi-disciplinary open access archive for the deposit and dissemination of scientific research documents, whether they are published or not. The documents may come from teaching and research institutions in France or abroad, or from public or private research centers.

L'archive ouverte pluridisciplinaire **HAL**, est destinée au dépôt et à la diffusion de documents scientifiques de niveau recherche, publiés ou non, émanant des établissements d'enseignement et de recherche français ou étrangers, des laboratoires publics ou privés.



Distributed under a Creative Commons CC BY-NC 4.0 - Attribution - Non-commercial use - International License

A great masquerader disease...

Thibault Maillet,¹ Mathilde Funes de la Vega,² Céline Duperron,³ Sylvain Audia.¹

¹Department of Internal Medicine and Clinical Immunology, University Hospital, Dijon, France

²Department of Pathology, University Hospital, Dijon, France

³Department of Urology, University Hospital, Dijon, France

Corresponding author

Sylvain Audia, MD, PhD

Department of Internal Medicine and Clinical Immunology, 14 rue Paul Gaffarel, François Mitterrand
University Hospital, 21000 Dijon, France.

E-mail: sylvain.audia@u-bourgogne.fr

Phone: +333 80 29 34 32

Fax: +333 80 29 39 12

Case description

A 64-year-old-man, with history of hypertension, dyslipidemia, serous chorioretinopathy and past-smoking history (15 pack-years) presented with asthenia and weight loss (6 kg in 6 months). His physical examination was unremarkable and biological tests showed inflammation (CRP 20 mg/L). A retroperitoneal fibrosis was diagnosed on CT-scan with a left pulmonary nodule and mediastinal lymphadenopathy (**Fig 1. A, B**), these lesions being hypermetabolic on PET-scan. A lung nodule biopsy showed no malignant cells, but fibrosis associated with a lymphocytic infiltration. Despite a strong suspicion of cancer, the patient first refused further investigations. Nine months later, a control CT-scan revealed a mass of the left kidney while the pulmonary nodule was stable. As the characteristics of the renal mass argued for a renal cancer on MRI (**Fig 1.C**), the diagnosis of cancer-associated retroperitoneal fibrosis was suspected, and a nephrectomy was performed.

What is the diagnosis?

Pathological examination showed a storiform pattern fibrosis, associated with obliterative phlebitis and a lymphoplasmacytic infiltrate with an IgG4/IgG ratio greater than 50% (**Fig 1. D, E**). The diagnosis of IgG4-Related Disease with pseudotumors of the kidney and the lung, and retroperitoneal fibrosis was retained. The elevated plasma level of IgG4 (255 mg/dL, normal range <135 mg/dL) decreased to 188 mg/dL after nephrectomy. As a progression of retroperitoneal fibrosis would definitively alter renal function, a treatment was indicated. Because of the history of serous chorioretinopathy, steroids were contraindicated, and rituximab was given (1g day 1 and 15) followed by a maintenance therapy (500 mg every 6 months), leading to persistent clinical and metabolic remission at 4-year follow-up. IgG4 concentration decreased to 54 mg/dL. Despite chronic B cell depletion, no infection nor hypogammaglobulinemia were observed.

IgG4-RD is a pleomorphic disease with clinical manifestations including retroperitoneal fibrosis, pancreatitis, salivary gland swelling or ophthalmological involvement (1). Pseudotumors are rare, usually periorbital (1), while pulmonary or renal localizations are exceptional (2). Steroids remain the first line therapy, but rituximab is of interesting as second line therapy and for maintenance therapy (3).

In our case, while the first lung biopsy argued for IgG4-RD, the emergence of a kidney tumor prompt us to revise this diagnosis to suspect a kidney malignancy associated with paraneoplastic retroperitoneal fibrosis, thus motivating nephrectomy. However, clinicians should keep in mind that IgG4-RD can rarely be responsible for kidney tumor and that in such a situation, percutaneous biopsy must be favored to avoid the removal of a non-malignant functional organ.

References

1. Vasaitis L. IgG4-related disease: A relatively new concept for clinicians. *Eur J Intern Med.* 2016; 27:1-9
2. Chougule A, Bal A. IgG4-related inflammatory pseudotumor: A systematic review of histopathological features of reported cases. *Mod Rheumatol.* 2016; 14:1-6
3. Campochiaro C, Della-Torre E, Lanzillotta M. Long-term efficacy of maintenance therapy with Rituximab for IgG4-related disease. *Eur J Intern Med* 2020 Apr; 74:92-98

Figure legend

Figure 1. Radiologic explorations: **A)** CT-scan showing retroperitoneal fibrosis and **B)** lung pseudotumor; **C)** Magnetic Resonance Imaging showing a tumor of the lower part of the left kidney. T1 Fat Sat Gadolinium ponderation. **Histopathology of the kidney:** **D)** Hematoxylin-eosin-saffron staining of the renal tumor, showing an obliterative phlebitis (white arrow) surrounded by a storiform-pattern fibrosis (star) with an inflammatory lymphoplasmacytic infiltration (black arrow) and **E)** the presence of IgG4 plasma cells (Cell Marque, clone EP138, dilution 1/1000).

