Case Report

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Retroperitoneal fibrosis associated with orbital pseudotumor without evidence of IgG4: A case report with review of literature

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Summary **Retroperitoneal fibrosis (RPF) is a rare disease characterized by chronic inflammation and periaortic fibrosis that affects retroperitoneal structures and often entraps the ureters. The idiopathic form has an incidence of 0.1-1.3/100,000 person-years. A substantial percentage of patients with idiopathic retroperitoneal fibrosis (IRF), as well as patients with orbital pseudotumor, is associated with IgG4-related disease (IgG4-RD). It is not clear what percentage of IRF is related to the spectrum of the IgG4-RD or if both represent different stages of the same disease (especially in those cases with extra-retroperitoneal involvement). Histopathological features such as storiform fibrosis, obliterative phlebitis and tissue infiltration of IgG4-positive plasma cells (ratio $IgG4^+$/IgG higher than 0.4) are essential to identify this association. Extra-retroperitoneal manifestations are often presented among patients with IgG4-related RPF. About 90% of cases of IRF have a good prognosis, with adequate response to treatment. We report a case of a 59-year-old woman with history of past occupational asbestos exposure and smoking habit. She was diagnosed with RPF, periaortitis and orbital pseudotumor, without histopathologic or serologic features of IgG4-related disease. This could be related to the fact that the biopsy was done in a place with scarce inflammatory activity but high fibrosis. We want to emphasize the usual need to perform several biopsies or to be guided by positron emission tomography (PET-CT) in order to achieve a histopathological confirmation. Our case differs from the main IgG4 international cohorts in the involvement of the retroperitoneum, aorta and eye, whereas the usual involvement includes liver, pancreas, lymph nodes and salivary glands. Our patient had lower IgG4 serum levels than those described in the international cohorts. However, they were similar to those of the Spanish population.**

Keywords: Idiopatic retroperitoneal fibrosis, IgG4-related disease, orbital pseudotumor, periaortitis, asbestos

1. Introduction

Idiopathic retroperitoneal fibrosis (IRF) is a rare immune-mediated condition with systemic involvement that affects retroperitoneal structures; it is characterized by chronic inflammation and periaortic fibrosis. The incidence rate is 0.1-1.3/100,000 person-years with a median age at diagnosis of 40-60 years and affects males more frequently (*1-3*). The majority of cases (70%) of retroperitoneal fibrosis (RPF) are idiopathic, whereas one third of cases are secondary to other causes such as drugs, cancer or infections (*2,4*). Its diagnosis can be challenging due to the wide range of associated diseases, and the need to eliminate secondary causes. Although it has a benign course in the majority of cases, it can also lead to severe complications. Therefore, it is important to treat it at an early stage and monitor it very closely (*4*).

Although it has been recently included in the spectrum of IgG4-related disease (IgG4-RD), this association has not been proved in all patients. IgG4-RD is a systemic fibroinflammatory condition

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characterized by the infiltration of IgG4-positive plasma cells in the affected tissues (mostly pancreas, salivary glands and lymph nodes, although it can affect a large number of tissues) (*3,5,6*).

We present the case of a 59-year-old woman diagnosed with retroperitoneal fibrosis. In this particular patient, orbital pseudotumor presented as an extra-retroperitoneal lesion, which would point to an IgG4-related disease, but the histo-pathological and laboratory findings suggested otherwise.

2. Case Report

A 59-year-old woman with a history of occupational asbestos exposure and a smoking habit came to the Emergency department with a 1-month history of abdominal pain, vomiting, hyperthermia, weakness and 5 kg weight loss. She had progressively developed bilateral proptosis over the last year. Upon arrival at the hospital, physical examination showed fever (37.8ºC), tachycardia, bilateral restriction on upgaze, mild ascites and a palpable mesogastric abdominal mass. Laboratory tests revealed the following alterations: leukocytes 17,200/µL (neutrophils 10,900/µL, monocytes 5,100/µL, lymphocytes 1,100/µL), Hemoglobin 10.7 g/dL, platelets 404,000/µL, elevated C reactive protein (CRP) and erythrocyte sedimentation rate (ESR) (12.7 mg/dL and 44 mm respectively), albumin levels 3.1 g/dL and ferritin 392 µg/L. Other biochemistry blood parameters including glucose, liver enzymes, renal function, serum angiotensin-converting enzyme, electrolytes and 24-hour urine analysis were normal. Peripheral blood smear confirmed monocytosis. Blood cultures were sterile. Urine infection caused by Escherichia coli was detected in the urine culture. Despite appropriate antibiotic therapy, fever persisted. Concentrations of B, T-CD8^{+} and NK lymphocytes were low. Low levels of serum IgG (516 mg/dL) and elevated values of IgE (192 mg/dL) were observed. IgG4 (0.18 g/dL), IgA and IgM were within normal range.

A computerized tomography (CT) showed a soft tissue mass around the aortic arch, the descending aorta and iliac arteries that infiltrated the retroperitoneum, the mesenteric vessels and the inferior vena cava and caused bilateral obstructive uropathy. An inflammatory infiltration of orbital soft tissues suggestive of orbital inflammatory pseudotumor was also described (Figure 1). There was no evidence of lymph node enlargement. Positron emission tomography (PET-CT) revealed inflammatory activity of the infiltrative retroperitoneal, periaortic and orbital masses as well as in pericardium, right atrium, pleura and perinephric space (Figure 2). Further studies including transthoracic echocardiogram, large bone radiography, upper endoscopy and colonoscopy, were normal. Tumoral biomarkers and autoantibodies were negative, with the exception of elevated CA-125 values (114 U/mL). Core needle biopsy of the retroperitoneal mass demonstrated a fibroblastic proliferation and IgG plasma cells without atypia on a collagenous and adipose tissue stroma. Immunohistochemical (IHC) staining did not reveal IgG4 or other cell line markers. These findings were compatible with the diagnosis of idiopathic retroperitoneal fibrosis (Figure 3).

The patient needed parenteral nutrition due to oral feeding intolerance. She also developed obstructive uropathy and it was necessary to implant an ureteral stent. Treatment with high doses of intravenous methylprednisolone (1 g/day) for 3 days was started with a significant clinical improvement with resolution of proptosis, fever, vomiting, ascites and abdominal pain as well as with normalization of the acute phase reactants (APR). The initial corticosteroids bolus were followed by lower doses of oral prednisone. She was discharged 2 weeks after the beginning of the immunosuppressive treatment to be seen on an

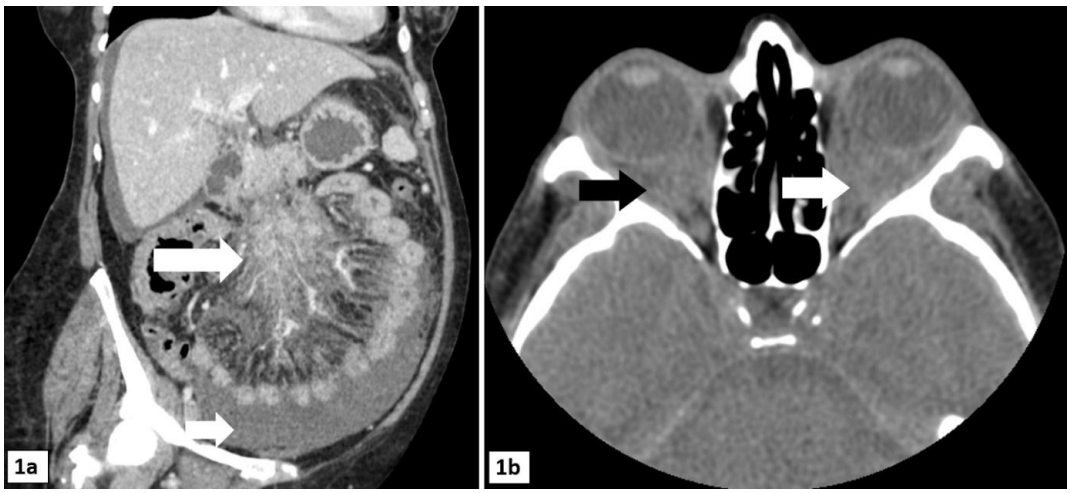

Figure 1. (1a), Abdominal CT. Mesenteric and retroperitoneal infiltrative mass surrounding mesenteric vessels and the inferior vena cava (big arrow). Free intraperitoneal fluid (small arrow). **(1b),** Cranial TC. orbital inflammatory pseudotumor (arrows).

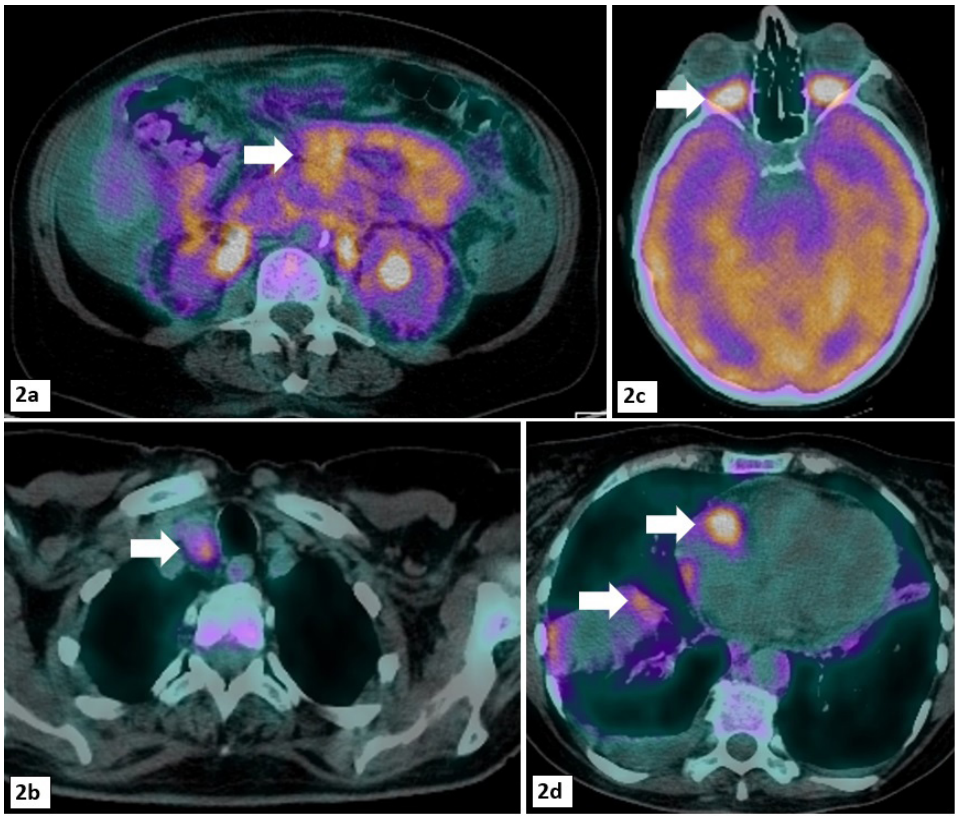

Figure 2. PET-TC. Inflammatory activity (arrows) of the infiltrative retroperitoneal **(2a),** periaortic **(2b)** and orbital masses **(2c)** as well as pericardium, right atrium, pleura **(2d)** and perirenal space.

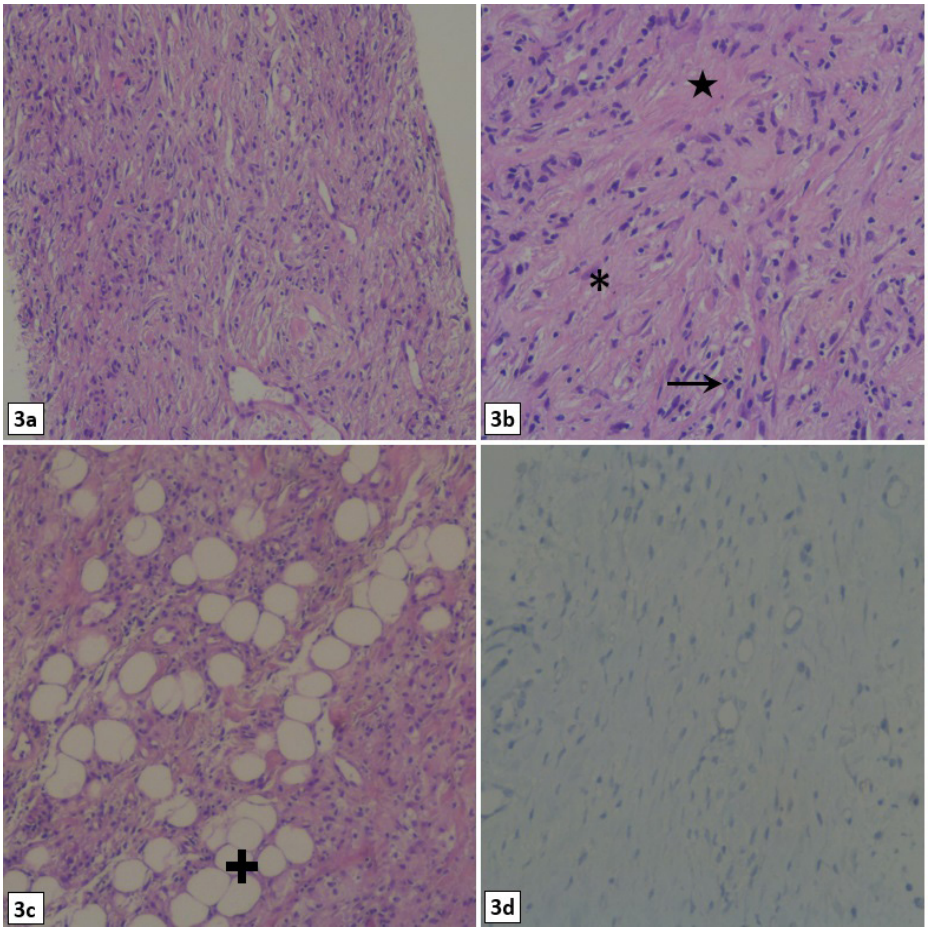

Figure 3. (3a, 3b and 3c), fibroblastic proliferation (*) and plasma cells (arrow) without atypia on a collagenous (star) and adipose tissue stroma (cross). **(3d),** Negative immunohistochemical staining for Ig G4.

outpatient basis. 3 days after that the patient had clinical deterioration and was admitted to intensive care unit with an abdominal sepsis due to urinary tract infection and pseudomembranous colitis. Treatment with broad-spectrum antibiotics and vasoactive drugs was initiated. Unfortunately, the patient did not respond to the treatment and evolved with multiple organ failure (hepatic, renal and encephalopathy) and was deceased after one week.

3. Discussion

RPF, also named Ormond's disease, is a rare process characterized by chronic inflammation and periaortic fibrosis that affects retroperitoneal structures. The incidence rate is 0.1-1.3/100,000 person-years with a median age at diagnosis of 40-60 years and affects males more frequently (*1*). The pathophysiological mechanisms of this disease remain unknown. Some studies suggest that an immune-mediated mechanism might be involved, based on the frequent association of RPF with autoimmune disorders (*2,3*). The majority of cases (70%) of RPF are idiopathic whereas one third of cases are secondary to other causes such as drugs (derivatives of ergot alkaloids, methyldopa, beta-blockers and biological agents); metastatic neoplasm with a desmoplastic response (*e.g*, carcinoma of the prostate, breast, lung or genitourinary tract among others); carcinoid tumors; infections such as tuberculosis, histoplasmosis and actinomycosis or primary retroperitoneal malignancies such as sarcomas or lymphomas (*2,4*). Other processes such as secondary amyloidosis, surgical interventions, abdominal trauma, barium enemas, radiotherapy, mesenteric panniculitis and particular forms of histiocytosis – specially Erdheim-Chester disease (ECD) – have also been described (*2,7-9*). Some studies have concluded that asbestos or tobacco exposures are strong risk factors for the development of idiopathic retroperitoneal fibrosis (IRF) (*10*).

IgG4-RD is a relatively new clinical entity, which can be described as a systemic fibroinflammatory condition with variable organ involvement characterized by the infiltration of IgG4-positive plasma cells in the affected tissues (*3*). Up to 50% of patients with IgG4-RD usually have a history of chronic allergic conditions. It can affect virtually any territory and the territories most frequently affected are pancreas, salivary glands and lymph nodes (*5*). The diagnosis of this entity is based on clinical, serologic and pathological studies. It is widely thought that IRF belongs to the spectrum of IgG4-RD (*6*). Nevertheless, there are some major points that continue to be a cause for debate. The differences in clinical manifestations, response to treatment and prognosis between IgG4-related and -unrelated IRF are not known very well, and therefore they need to be investigated with larger prospective studies. As we do not know what their differences and similarities are, we can not know what proportion of patients can be "IgG4-related". Therefore, most authors use anatomopathological and IHC criteria to classify them (storiform fibrosis, obliterative phlebitis and tissue infiltration of IgG4-positive plasma cells). The comparison between the two groups seems to associate more frequently with the presence of extra-retroperitoneal manifestations to IgG4-related IRF (*3*).

Clinical manifestations are similar in the different forms of RPF and often initially nonspecific. The definitive diagnosis is usually delayed until there is significant organ damage. At the time of diagnosis most cases frequently present with obstructive uropathy and altered renal function. The most frequent symptom is abdominal pain. Systemic symptoms are frequent and include malaise, fatigue, nausea, vomiting, anorexia, myalgia and loss of weight. Lower extremity edema, deep vein thrombosis, mesenteric ischemia, claudication of upper and lower extremities, scrotal swelling, varicocele, hydrocoele or secondary arterial hypertension are complications that appear as a result of the compression of lymphatic and blood vessels (*2,3,11*). Clinical features and laboratory findings are nonspecific, therefore IRF is a diagnosis of exclusion (3).

Most patients have increased levels of CRP and ESR and its monitoring can be useful to evaluate the clinical course of the disease, although it has limited prognostic value. Other laboratory findings are chronic inflammatory anemia, leukocytosis, eosinophilia and decreased glomerular filtration rate. The urinary sediment is most often normal (*2,4*). Evaluation of antinuclear antibodies (ANAs), anti-neutrophil cytoplasmic antibodies, anti-thyroid microsomal antibodies, and antithyroglobulin antibodies is necessary. ANAs are positive in 60% of cases and autoimmune thyroiditis is the autoimmune disease most frequently associated (*12*). Serum IgG4 concentrations are elevated in approximately 60-70% of patients with IgG4-related IRF (*10*).

Imaging studies play a key role in the diagnosis. While ultrasonography is useful for the identification of hydronephrosis or aneurysmatic aortic dilatation, CT is considered the gold standard for the diagnosis. It can show the presence of homogeneous tissue isodense to muscle, surrounding the lower abdominal aorta and the iliac arteries, and often enveloping other intra-abdominal structures. It also allows to rule out secondary causes (*13*). MRI usually provides better definition of IRF (*14*). PET-CT is recommended to evaluate the extension, activity and evolution of this entity. PET-CT demonstrates higher uptake in active phases of the evolution, whereas advanced phases are characterized by marked fibrosis and therefore the uptake is lower. It guides the best management option allowing to decide between immunosuppressive treatment and decompression surgery (*15*).

Table 1. Characteristics of IgG4-RD cohorts from China, US, Italy, Spain and Japan

Items	Hong Kong, China (*n* = 20) (*20*)	Massachusetts, US (*n* = 125) (*21*)	Milan, Italy (*n* = 41) (*22*)	Spain (*n* = 55) (*23*)	Hokuriku region, Japan (*n* = 235) (*24*)	Beijing, China (*n* = 118) (*25*)
Male	42 (76.4%)	76 (60.8%)	26 (63.4%)	38 (69.1%)	189 (80.4%)	82 (69.5%)
Age (years)	62 (27-86)	55 (24-83)	62 (55-67)	53 (41-64)	67 (35-86)	53 (19-80)
Organ involvement						
Retroperitoneum	7 (12.7%)	23 (18.4%)	8 (19.5%)	15 (27.27%)	9 (4%)	31 (26%): retroperitoneal fibrosis and periaortitis
Hepatobiliary and pancreatic system	26 (47.3%)	24 (19.2%)	Pancreas: 17 (41.5%); biliary tree: 4 (9.8%)	Pancreas: 9 (16.36%)	Pancreas: 142 (60%); biliary tree: 31 (13%)	Pancreas: 45 (38.1%); sclerosing cholangitis: 21 (17.8%)
Aorta	NA	14 (11.2%)	4 (9.75%)	4 (7.27%)	28 (20%)	NA
Salivary gland	24 (43.6%)	Submandibular: 35 (28%); parotid: 21 (16.8%)	8 (19.5%)	9 (16.36%)	81 (34%)	76 (64.4%)
Lymph node	8 (14.5%)	34 (27.2%)	5 (12.2%)	2 (1.81%)	34 (14%)	77 (65.3%)
Eye	8 (14.5%)	28 (22.4%)	Orbit: 3 (7.3%); lacrimal glands: 2 (4.9%)	Orbital pseudotumor: 12 (21.82%); lacrimal glands: 8 (14.55%)	Orbit: 9 (4%); l acrimal glands: 53 (23%)	Orbital pseudotumor: 10 (8.5%); lacrimal glands: 60 (50.8%)
Lung	7 (12.7%)	22 (17.6%)	1 (2.4%)	4 (7.27%)	31 (13%)	32 (27.1%)
Renal system	2 (3.6%)	15 (12%)	1 (2.4%)	4 (7.27%)	54 (23%)	29 (24.6%)
Central nervous system	1 (1.8%)	Meninges: 3 (2.4%)	Meninges: 3 (7.3%)	Meninges: 2 (3.64%)	NA	NA
Skin/soft tissue	1 (1.8%)	2 (1.6%)	NA	NA	NA	5 (4.2%)
No. of involved organ systems	1.7 (1-5)	2.3 (1-7)	1.51 (1-3)	NA	NA	NA
Multiorgan involvement	NA	41 (39.78%) *n* = 103	17 (41.46%)	36 (47.3%)	136 (58%)	93 (78.8%)
Serum IgG4 (mg/dL)	660.5 (116-2100) *n* = 48	216.56 (28-817) *n* = 105	284 (132-545)	163.90 (30.8-1145-2)	470 (22-4150) *n* = 229	1521.8
Total IgG (mg/dL)	2202.1 (1080-5900) *n* = 43	1339.58 (868-2114) *n* = 93	NA	NA	NA	2300
IgG4/IgG ratio	0.29 (0.04-0.65) *n* = 43	NA	NA	104.29	NA	0.38
Histopathological confirmation	40 (72.7%)	125 (100%)	30 (73.2%)	55 (100%)	150 (64%)	64 (54.2%)
Treatment						
Glucocorticoids	37 (67.3%)	64 (51.2%): 86% improved; 77% non-remission	36 (87.8%): 70.73% remission; 29.27% non-remission	47 (85.5%): 43.6% complete response; 43.7% partial response (< 50% of regression)	167 (71%): 24% non remsion; 10% mortality	114 (96.6%)
Surgery	19 (34.5%)	50 (40%)	4 (9.75%)	16 (29.1%)	21 (9%)	NA
Other	12 (21.81%): other immunosuppressants	16 (13%): urethral and hepatobiliary stents	10 (24.39%): hepatobiliary stent	19 (34.5%): other immunosuppressants	NA	71 (60.2%): glucorticoids and other immunosuppressants

IgG4-RD: IgG4-related disease; NA = not available.

Anatomopathological study of biopsy samples is necessary to rule out secondary causes and to establish a definitive diagnosis, especially when imaging techniques do not show characteristic IRF findings. Biopsy samples reveal both a fibrous tissue (type I collagen, fibroblasts and myofibroblasts) and a variably inflammatory infiltrate (lymphocytes, macrophages, plasma cells, and more rarely eosinophils) organized into perivascular and diffuse patterns. In the perivascular pattern, aggregated lymphocytes surround the small retroperitoneal vessels and usually have a central core of B cells and a periphery of $CD4^+$ and $CD8^+$ T cells. In cases of IRF secondary to malignant conditions it is usually necessary to obtain multiple biopsies, because neoplastic cells are frequently scattered into abundant fibrous tissue (*2*). The IgG4-RD more commonly shows obliterative phlebitis, mild-to-moderate eosinophil infiltrate and fibrosis with a storiform pattern and positive IHC for IgG4 (with a ratio $IgG4^+$/IgG higher than 40%). It is usually necessary to perform several biopsies or to be guided by PET-CT in order to get an histopathological confirmation from a zone with high inflammatory activity (*3,10*). ECD is characterized by infiltration of typically lipid-laden histiocytes with admixed or surrounding fibrosis. On IHC staining, ECD histiocytes are positive for CD68, CD163, and Factor XIIIa, and negative for CD1a and Langerin (*9*).

Treatment should be started as soon as possible. In case of renal failure due to hydronephrosis, ureteral decompression (ureteral stents or nephrostomies) should rapidly be accomplished in order to avoid permanent renal damage (*16*). In cases of secondary RPF we need to treat the underlying condition. Medical treatment of IRF consists in early administration of prednisone (0.5-1 mg/kg/day during the first month). After clinical re-evaluation, if remission is achieved the initial dose should be progressively decreased. Most studies recommend to maintain steroid treatment for at least 9 months (*2,3*). If there is a contraindication to prednisone therapy, some studies suggest the use of tamoxifen (*2,17*). In case of refractory forms (patients who fail to achieve clinical or radiologic improvement within 4 to 6 months) mycophenolate mofetil or methotrexato associated with a low dose of prednisone have been proposed as good alternatives. Recent studies with rituximab, infliximab and tocilizumab have demonstrated good results although further studies are needed to draw definitive recommendations (*2,18,19*).

Regarding follow-up, patients should be monitored clinically, and imaging techniques and laboratory tests should be used. It includes inflammatory markers and renal function status. Ultrasonography is a useful and harmless technique which allows surveillance of obstructive uropathy. PET-CT enables the evaluation of disease activity and its extension.

IRF tends to have a good prognosis and evolution with treatment. Approximately 90% of cases have an adequate response to treatment with a mortality rate lower than 10%. The relapse frequency is estimated to be around 10%, especially after the withdrawal of steroid treatment (*4*).

We performed a systematic review in PubMed using the terms "IgG4-related disease" and "case series" in English and 71 articles were found. We excluded case reports, review articles and case series that addressed only a specific organ involvement of IgG4-related disease. We also reviewed the references of those articles. We found ten articles of IgG4-RD case series and we finally included six of them in Table 1 (*20-25*). Unlike the principal IgG4-RD cohorts, in which the most frequently affected organs are liver, pancreas, lymph nodes and salivary glands, our case shows involvement of retroperitoneum, aorta and eye. Despite being considered a multisystemic illness, most of the described cases only had apparent infiltration in one organ. Our patient had lower IgG4 serum levels than those described in the international cohorts. However, they were similar to those of the Spanish population. As it happens on many occasions, our case did not show histopathologic features of IgG4-related disease, probably because only one biopsy was performed and this disease usually shows a patchy effect on tissues. The patient evolved with multiple organ failure and was deceased three weeks after the immunosuppresive treatment was initiated, despite that both IRF and IgG4-RD tend to have a good prognosis with low frequency of complications and favorable evolution with treatment (*20-25*).

In conclusion, we presented a case of a 59-year-old woman with idiopathic retroperitoneal fibrosis, periaortitis and orbital pseudotumor, without histopathologic or serologic features of IgG4-related disease. This could be related to the fact that the biopsy was done in a place with scarce inflammatory activity but high fibrosis. We want to emphasize the usual need to perform several biopsies or to be guided by PET-CT in order to achieve a histopathological confirmation. Although knowledge about IRF has significantly improved, it still remains an ambiguous condition. A lot of questions still need to be answered, especially about the pathogenesis. Further research on the relationship between IRF and IgG4-RD is required.

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