Clinical note

18F-FDG PET-CT usefulness in extra-pancreatic involvement in IgG4 related diseases

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A B S T R A C T

IgG4-related diseases are a group of recently identified entities that include disorders that were previously known by other names, such as Mikulicz disease, Küttners’s tumor, Riedel thyroiditis, among others, as well as some new ones described in the last years.

These pathologies are a challenge for the medical community in terms of diagnosis and characterization due to their wide spectrum of clinical presentation.

Functional imaging can provide a new approach to the comprehension of physiopathology, staging and targeting site of biopsy of IgG4-related diseases.

In this clinical note, we describe five patients who underwent 18F-FDG PET-CT and correlate their findings with previous reports.

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Utilidad de la 18F-FDG PET-TC en las manifestaciones extrapancreáticas de las enfermedades relacionadas con IgG4

R E S U M E N

Las enfermedades relacionadas con IgG4 son un grupo de entidades recientemente identificadas; que engloban a patologías anteriormente conocidas como la enfermedad de Mikulicz, el tumor de Küttners y la tiroiditis de Riedel entre otras, las cuales se creían que eran entidades diferentes, y otras descritas en los últimos años.

Estas patologías plantean un reto diagnóstico para el clínico y el médico nuclear, dado su amplio espectro de presentación clínica.

La imagen funcional puede brindar un nuevo abordaje con respecto a la fisiopatología, estudio de extensión y optimización de los sitios de biopsia, en las enfermedades relacionadas con IgG4.

En esta nota clínica presentamos cinco casos clínicos, valorados mediante 18F-FDG PET-TC y correlacionamos hallazgos con la literatura médica reciente.

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IgG4-related diseases (IgG4-RDs) are a recently established systemic disorder which includes a group of entities that were previously known as other non-related pathologies, such as Mikulicz disease, Küttners’s tumor and Riedel thyroiditis and others, and new ones described in the last years. Kamisawa et al, were the first ones to describe extrapancreatic involvement in patients with autoimmune panreatitis, which has similar histopathologic characteristics (high number of IgG4 positive plasmatic cells, lympho-plasmacytic infiltrate and fibrosis).1,2

IgG4-RDs predominantly affect men (5:1, male:female), between 58 years and 69 years of age, and with antecedents of allergic pathologies (asthma, rhinitis or atopia).3

Multiorganic involvement has been described in meninges, orbital and cerebral tissues, salivary glands, thyroid, lungs, peri-cardium, pancreas, hepatobiliary tract, kidneys, lymph nodes, aorta, retroperitoneum, prostate, breast and skin.3,4

Two Japanese groups established diagnostic criteria for IgG4-RD, as follows5:

1. Diffuse or focal enlargement or mass lesions in one or more organs.
2. Elevated levels of serum IgG4 (more than 135 mg/dL).
3. Histopathological findings:
   (a) Marked lymphocyte and plasmacyte infiltration and fibrosis.
   (b) Infiltration of IgG4+ plasma cells: ratio of IgG4+/IgG+ cells >40% and >10 IgG4+ plasma cells/high power field (HPF).

The presence of three positive criteria confirms diagnosis. A possible case is a patient with 1 and 2 items presents. Due to its recent identification and extensive investigation, probably new diagnosis criteria could be added in the future.

Previous cases of extrapancreatic manifestations of autoimmune pancreatitis and IgG4-RDs had been reported,5 but many of them had synchronous affections. In our series, none of our patients had pancreatic involvement.

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Fig. 1. Sagittal (A) and axial (B) images from CT contrast enhanced, PET and PET-CT are shown, where a mass with increased tracer uptake (SUVmax 4.3) in descendant thoracic aorta is seen (white arrow).

Fig. 2. Axial CT, PET and PET-CT pelvic images in which a high metabolic activity in soft-tissue mass in presacral region (arrow) and bilateral inguinal adenopathies (arrowheads) are shown.

Fig. 3. Coronal reformatted (top row) and axial (bottom row) PET-CT images show a wall thickness in aorta, that extends from the root to aortic arch with a high uptake of radiotracer (SUVmax 11).
Fig. 4. CT, PET and fusion PET-CT axial abdominal images show a soft-tissue density mesenteric mass with an increase in metabolic activity (arrow). No other findings were seen in this study.

Description of cases

Five patients from 57 years to 71 years of age were included. We present each case with its clinical profile and 18F-FDG PET-CT findings:

Patient 1

Aortic thoracic descending enlargement was incidentally seen in a chest X-ray performed in an asymptomatic 57 year-old man. Computed tomography (CT) showed a left paraaortic mass, associated with enlarged mediastinal lymph nodes. Lymphoproliferative syndrome was suspected and 18F-FDG PET-CT was made, revealing an uptake in paraaortic mass and a retrocrural node (Fig. 1).

Fine needle biopsy was performed from paraaortic mass, rendering an inconclusive report. Patient underwent mass resection, and fibrosclerosing lymphadenitis related to IgG4 was confirmed. Subsequently, a corticoid treatment was initiated.

Serum IgG4 level at the time of diagnosis was 246 mg/dL.

Patient 2

A 69 year-old male with alcoholic cirrhosis and diabetes mellitus type 2 was admitted into the emergency room with abdominal pain, fever and sepsis, due to bacterial peritonitis. A retroperitoneal mass and abdominal adenopathies were found in an abdominal CT. Emergency laparotomy was made and a mass biopsy revealed an IgG4 limphadenopathy. IgG4 serum levels reached 5260 mg/dL at that moment.

Findings in 18F-FDG PET-CT were: a retroperitoneal mass and axillaries, external and internal iliac adenopathies, all of them with a slightly increased uptake (Fig. 2).

Patient 3

A 59-year-old man was admitted into the emergency room with suspicion of aortic syndrome, due to a serendipitous aortic wall thickening, that compromised from the root to the middle part of aortic arch, suggesting a hematoma.

The patient underwent emergency surgery, where aortitis signs were observed and IgG4 related periaortitis was shown in the biopsy tissue. During his stay in the hospital, IgG4 serum levels were 295 mg/dL. 18F-FDG PET-CT identified periaortic and mediastinal lymph nodes involvement, with high uptake of the tracer (SUVmax 11) (Fig. 3).

Patient 4

A 71 year-old male with abdominal pain and a mesogastrium mass was referred to our department to perform an 18F-FDG PET-CT (Fig. 4), which showed a mesenteric mass with elevated FDG uptake (SUVmax 7.4).

Exploratory laparotomy was performed and a biopsy was taken to gather intraoperative information.

Fig. 5. Sagittal (top row) and axial (bottom row) CT, PET and PET-CT show a nodular thickening in the root of the mesenterium with an increased FDG activity (SUVmax 4) (arrows).
Positive results for IgG4 related diseases from mesenterium were obtained. In this patient, the IgG4 levels were normal at (82 mg/dL) diagnosis time.

Patient 5

Retroperitoneal masses were found in an abdominal follow-up CT of a 58-year-old male, with antecedents of a resected ileal neuroendocrine tumor and systemic lupus erythematosus. Because of suspicion of recurrent neuroendocrine tumor or new malignancy, an 18F-FDG PET-CT was taken and, subsequently, a biopsy was performed. Histopathological findings showed changes of retractable mesenteritis related to IgG4. Blood levels of IgG4 were 157 mg/dL. Fig. 5 shows 18F-FDG PET-CT findings.

Discussion

Regarding epidemiologic data, all our patients were men with ages between 57 years and 71 years, and four out of five had increased IgG4 serum levels (80%) at diagnosis time. These findings correlate well with other series.5

These patients had different sites of involvement, such as mesenterium, periaortic tissues, retroperitoneum and lymph nodes. There was no pancreatic or hepatobiliary involvement in any of these cases, neither in images nor in clinical examinations. Autoimmune pancreatitis may show extrapancreatic associated involvement up to 80%,2-3,6 At diagnosis time, two patients were asymptomatic and two presented incidental findings in image studies, but the common clinical presentation was the presence of a mass involving thoraco-abdominal organs.4,7

In our series, patients were referred to us for PET-CT, with clinical suspicion of lymphoma, mesenteric neuroendocrine tumor or retroperitoneal fibrosis. IgG4-RD was not initially suspected in any of them. IgG4-related diseases can be difficult to identify in early stages and to discriminate from malignant processes.8 Clinicians and nuclear medicine physicians must keep in mind this differential diagnosis in middle-age male patients with pancreatic autoimmune disease and/or perivascular masses, with a slight uptake on 18F-FDG PET-CT and inconclusive histopathological reports.

18F-FDG PET-CT can play an important role in IgG4-related diseases patients, providing a whole-body overview and allowing the delineation and characterization of the metabolic activity of the disease. Additionally, 18F-FDG PET-CT can evaluate accessible sites for biopsy and, potentially, it could become a good tool to assess treatment response and relapse. Nevertheless, there is no wide evidence or consensus in the timing between scans during the follow-up and no defined criteria have been established to assess therapy response.5,8

Conclusion

IgG4-related diseases are a new entity, which has a wide spectrum of clinical presentations, as well as different sites of involvement. 18F-FDG PET-CT associated to clinical diagnosis criteria, permit diagnose correctly this entity; in our series 18F-FDG PET-CT, allow us to assess the extension and metabolic activity of the disease and target correctly site of biopsy.

References