CASE REPORT

Splenic lesion of IgG4-related disease in FDG-PET/CT

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Received: May 9, 2014 **Accepted:** May 21, 2014 **Online Published:** May 27, 2014
DOI: 10.5430/crim.v1n2p150 **URL:** http://dx.doi.org/10.5430/crim.v1n2p150

Abstract

Recent several reports showed that 18F-fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) scans accurately, and safely identifies the multiple organs' involvements in IgG4-related diseases (IgG4-RD) patients. Disease activity in two patients, aged 74 and 73 years (serum IgG4, 807 and 1630 mg/dl, respectively), with IgG4-RD were monitored using FDG-PET/CT. Scans before treatment showed splenomegaly with high FDG uptake. Inflammatory aneurysm, submandibular gland swelling, splenomegaly and tubulointerstitial nephritis resolved after treatment with prednisolone. Follow-up FDG-PET/CT at 4 months, with no symptoms, showed a significant decrease in FDG accumulation and improvement of splenomegaly in splenic lesion of IgG4-RD. Thus, splenic lesions detected by FDG-PET/CT might be novel involvements of IgG4-RD.

Keywords

IgG4-related disease, Spleen, Splenomegaly, FDG-PET/CT

1 Introduction

IgG4-related diseases (IgG4-RD) include autoimmune pancreatitis, retroperitoneal fibrosis, cholangitis, tubulointerstitial nephritis, breast lesions, prostatitis, lung lesions, Mikulitz's disease (lacrimal gland or salivary gland) and Kuttner's tumor (salivary gland) [1]. Moreover, the diagnosis criteria for IgG4-RD is consisted of only three items: 1) organ enlargement, mass or nodular lesions, or organ dysfunction, 2) a serum IgG4 concentration >135 mg/dl, and 3) histopathological findings of >10 IgG4+ cells/HPF and an IgG4+/IgG+ cell ratio >40% [1]. New diagnostic imaging methods for IgG4-related disease (IgG4-RD) have been sought. Recent several reports showed that 18F-fluorodeoxyglucose-positron emission tomography/computed tomography (FDG-PET/CT) scans accurately, and safely identify the multiple organs' involvements in IgG4-RD patients [2, 3]. We demonstrated herein two patients with IgG4-RD, whose splenic lesions as novel involvement of IgG4-RD could be detected using FDG-PET/CT scans.

2 Case presentation

Disease activity in two patients, aged 74 and 73 years (serum IgG4, 807 and 1630 mg/dl, respectively), with IgG4-RD (Patient 1 and 2, respectively) were monitored using FDG-PET/CT. These patients clinically presented submandibular swelling, acute kidney injury, left hypochondralgia and splenomegaly, and were diagnosed according to the findings that

biopsies of the submandibular gland and kidney revealed abundant IgG4-positive plasma cells. Scans before the steroid treatment showed splenomegaly with high 18F-fluorodeoxyglucose (FDG) uptake (see Figure 1A, B, C and D, arrows). High IgG4 and IgE levels, and hypocomplementemia improved, and inflammatory aneurysm, submandibular gland swelling, splenomegaly and tubulointerstitial nephritis resolved after treatment with prednisolone. Follow-up FDG-PET/CT at 4 months, with no symptoms, showed a significant decrease in FDG accumulation and splenomegaly in splenic lesion of IgG4-RD (see Figure 1A', B', C' and D', arrows).

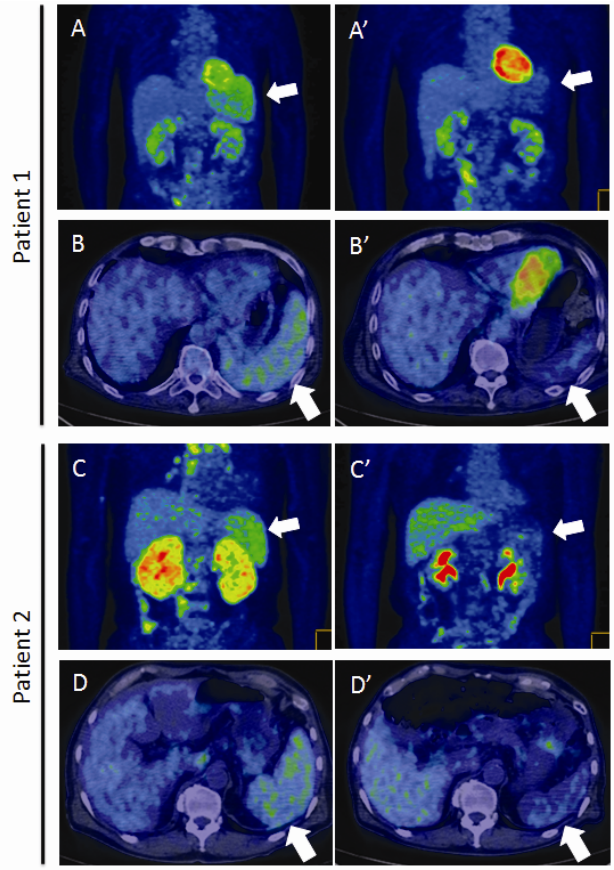

Figure 1. Evaluation of splenic involvement by FDG-PET/CT before and after treatment

3 Discussion

To our knowledge, there were several reports about the possibility of splenic involvement in IgG4-RD [4-7]. These reports indicated that sclerosing angiomatoid nodular transformation (SANT), as inflammatory pseudotumor, with increased numbers of IgG4+ plasma cells in spleen might be cases of IgG4-related sclerosing lesions of the spleen [4-6]. One report showed that inflammatory pseudotumor-like follicular dendritic cell sarcoma of the spleen with increased IgG4-positive plasma cells might be one of possible IgG4-RD [7]. In the present study, sarcoidosis and lymphoma were excluded by both laboratory (normal angiotensin converting enzyme and lysozyme levels) and pathological examinations. Furthermore, although pseudotumor was not revealed in spleen in both cases, splenic lesions detected by FDG-PET/CT in the present cases might be active involvements of IgG4-RD, considering that splenic lesions, splenomegaly and high FDG uptake in spleen, clinically improved by the treatment as same as other lesions of IgG4-RD. In fact, recent studies suggested that evaluation before and after treatment could show in most IgG4-RD cases a good correlation of FDG-PET/CT results with treatment response and disease activity [8]. Treatment effects could be assessed by the disappearance of FDG uptake [9]. Thus, splenomegaly and high FDG uptake in spleen detected by FDG-PET/CT as the present cases might be a possible prodromal lesion of SANT or inflammatory pseudotumor-like follicular dendritic cell sarcoma. We need to investigate

further cases with splenic lesion in IgG4-RD in order to elucidate the relationship between IgG4-RD and splenic involvement detected by FDG-PET/CT.

Acknowledgement

This study was supported by a grant from the Ministry of Education, Science, Culture and Sports of Japan government (to Yoshinori Taniguchi).

Disclosure statement

The authors declare no conflict of interest.

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