Case Report

Contrast-enhanced ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography in immunoglobulin G4-related retroperitoneal fibrosis

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ABSTRACT

Immunoglobulin G4 (IgG4)-related disease encompasses a wide variety of immune disorders previously thought be distinct. IgG4-related retroperitoneal fibrosis is one such entity. Metabolic imaging with ^{18}F-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography/computed tomography (PET/CT) can be useful in the management of IgG4-related retroperitoneal fibrosis. We here discuss the case of 63-year-old male with IgG4-related retroperitoneal fibrosis and the role, ^{18}F-FDG PET/CT played in his management.

Keywords: ^{18}F-fluorodeoxyglucose, immunoglobulin G4-related disease, positron emission tomography/computed tomography, retroperitoneal fibrosis

INTRODUCTION

Immunoglobulin G4-related disease (IgG4-RD) has been recognized as a wide spectrum immune-mediated inflammatory disease in past few years.[1] It can involve multiple organs and tissues such as the pancreas (autoimmune pancreatitis), biliary tract, lacrimal gland, salivary gland (sclerosing sialadenitis - Mikulicz disease), thyroid gland (Riedel's thyroiditis), lung, kidney, mediastinum (mediastinal fibrosis), and the retroperitoneum (retroperitoneal fibrosis/lymphoplasmacytic aortitis),[2] all characterized by a typical histopathological finding of infiltration by IgG4 rich plasma cells and storiform fibrosis.[3] IgG4-related retroperitoneal fibrosis can occur as part of multisystem disease or can present in isolation. Metabolic imaging with ^{18}F-fluorodeoxyglucose (^{18}F-FDG) positron emission tomography/computed tomography (PET/CT) can play an important role in diagnosis of IgG4-related retroperitoneal fibrosis.[4] We here present a case of IgG4-related retroperitoneal fibrosis and discuss the potential implications of ^{18}F-FDG PET/CT in such patients.

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CASE REPORT

A 63-year-old male presented with progressive pain abdomen, weight loss, and loss of appetite of 4 months duration. He was nondiabetic and nonhypertensive. He also had a history of cutaneous psoriasis managed with topical treatment. On examination, there was mild fullness in the left renal region. Hemogram was within normal limits. His erythrocyte sedimentation rate was elevated (60 mm in 1 h). His renal and liver function tests were normal. Ultrasound of the abdomen revealed dilated left pelvicalyceal system and left upper ureter. Left kidney was otherwise normal. Also noted were multiple matted preaortic lymph nodes with mass formation, compressing the middle part of the left ureter, causing obstructive left hydronephrosis. He underwent cystoscopy with bilateral retrograde pyelography and left DJ stenting. Suspecting a malignant etiology, the patient underwent contrast-enhanced ^{18}F-FDG PET/CT. PET/CT findings revealed a heterogeneously enhancing retroperitoneal soft tissue mass encasing the left ureter, aorta, left renal vessels, and left common iliac vessels, with heterogeneous ^{18}F-FDG uptake [maximum standardized uptake value (SUVmax)-3.7] [Figures 1a-c and 2a-i]. No other hypermetabolic lesion was seen in the rest of the body. Based on PET/CT findings, diagnosis of active retroperitoneal fibrosis was given. To get a definitive tissue sample, the patient underwent exploratory laparotomy and incisional biopsy. Operative findings revealed diffuse plaque like retroperitoneal mass engulfing the left ureter and adherent to aorta and iliac vessels. Biopsy was taken from the most metabolically active part of the mass near aortic bifurcation. Frozen section was negative for malignancy and

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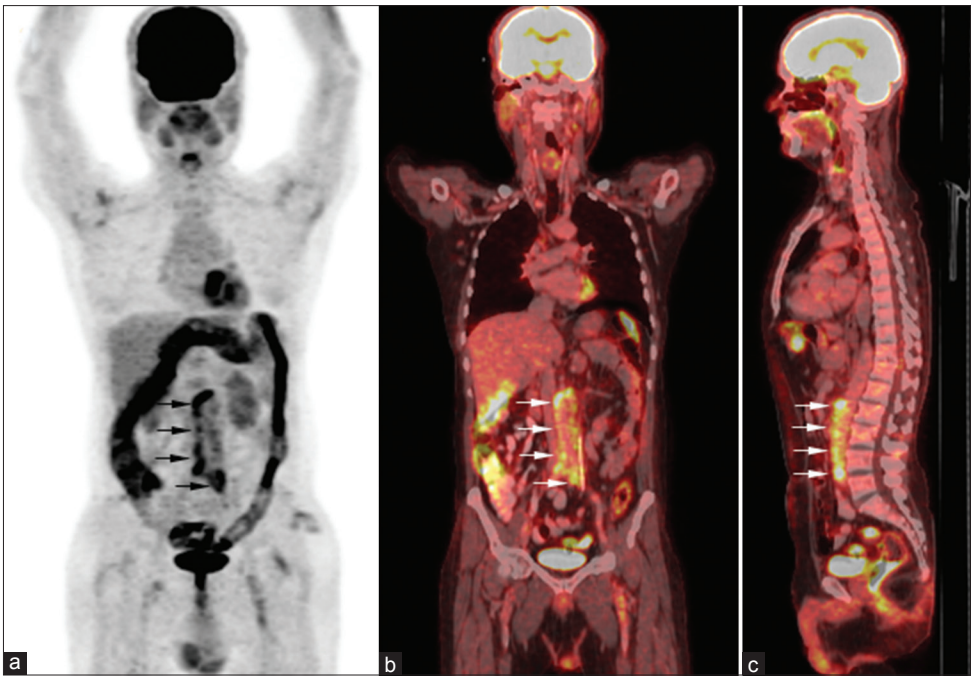

Figure 1: Maximum intensity projection positron emission tomography (PET) image (a) showing the patchy retroperitoneal lesion with moderate to intense ^{18}F-fluorodeoxyglucose (^{18}F-FDG) uptake (arrows, maximum standardized uptake value-3.7). No other abnormal hypermetabolic focus is seen in the rest of the body. Coronal (b) and sagittal (c) PET/computed tomography images demonstrating ^{18}F-FDG avid retroperitoneal soft tissue mass encasing the abdominal aorta (arrows)

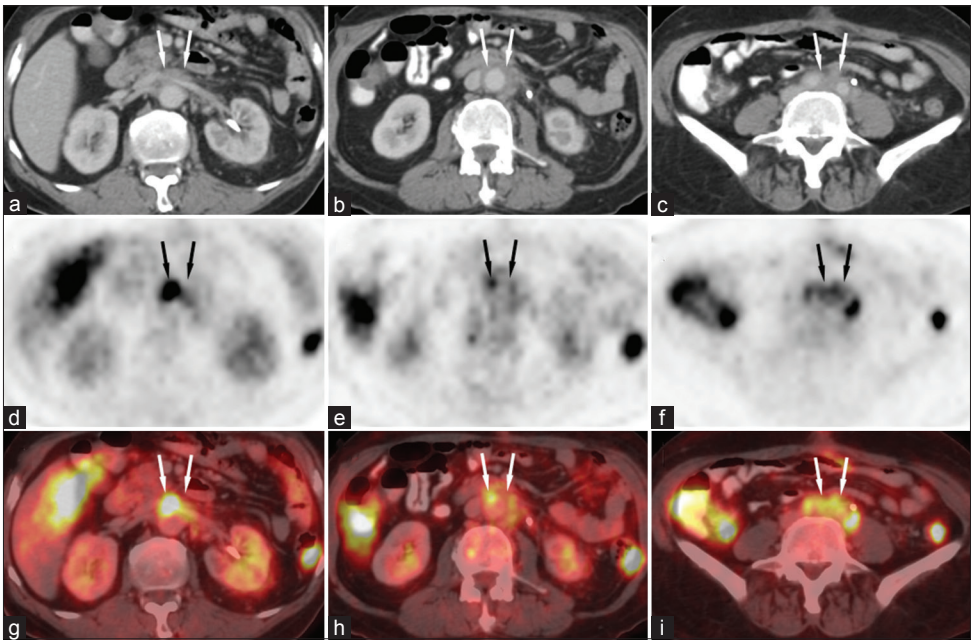

Figure 2: Transaxial contrast-enhanced computed tomography (CT) images of the abdomen (a-c) showing heterogeneously enhancing retroperitoneal soft tissue mass extending from the level of superior mesenteric artery to left iliac region and encasing abdominal aorta, left renal vessels, left iliac vessels, and left ureter (arrows). On transaxial positron emission tomography (PET) (d-f) and PET/CT (g-i) images, heterogeneous ^{18}F-fluorodeoxyglucose uptake is seen in the retroperitoneal mass (arrows)

suggestive of retroperitoneal fibrosis/pseudotumor. Histopathology revealed intense storiform fibrosis with dense lymphoplasmacytic infiltrate in collagenized connective tissue stroma and many reactive lymphoid follicles. No granuloma was seen. There was no evidence of lymphoma. Immunohistochemistry revealed substantial increase in IgG4 expressing plasma cells. These findings confirmed IgG4-related retroperitoneal fibrosis. Serum IgG4 levels were also elevated (IgG4-167 mg/dl). The patient was put on oral steroids with marked symptomatic improvement.

DISCUSSION

Immunoglobulin G4-related disease can mimic malignancy, clinically as well as on imaging, but can be treated easily with

corticosteroids.[5] Making a diagnosis of IgG4-RD is a challenge due to the wide spectrum of clinical presentations. Recently, two diagnostic criteria have been proposed for IgG4-RD: (1) Serum IgG4 concentration >135 mg/dl and (2) >40% of IgG positive plasma cells being IgG4 positive and >10 cells/high powered field of biopsy sample.[3] In the present case, the serum IgG4 levels were elevated (167 mg/dl) and there was a preponderance of IgG4 positive plasma cells in the tissue biopsy. However, in many instances the serum IgG4 levels might not be elevated. In addition, it is not ethical or feasible to biopsy all sites of suspected disease involvement or biopsy may not be adequate for diagnosis.[6] These difficulties can be obviated with the use of ^{18}F-FDG PET/CT.

^{18}F-fluorodeoxyglucose PET/CT is commonly employed in oncological imaging. In patients with IgG4-RD, PET/CT is generally employed to differentiate it from malignancy.[7] In the present case too, PET/CT was performed to exclude lymphoma. While the pattern of disease on ^{18}F-FDG PET/CT was strongly suspicious for IgG4-related retroperitoneal fibrosis in this patient as per the criteria (patchy retroperitoneal lesion with moderate to intense ^{18}F-FDG uptake) described by Zhang *et al*.,[8] histopathology is required to confirm the diagnosis. Due to the metabolic nature of the study, ^{18}F-FDG PET/CT can also be employed for selecting the appropriate site of biopsy.[9] In the present case, biopsy was performed from the most metabolically active portion of the mass, near the aortic bifurcation. Another major contribution of ^{18}F-FDG PET/CT is whole body mapping of disease extent in IgG4-RD.[10,11] In the present case apart from retroperitoneal mass, no other hypermetabolic lesion was seen in the rest of the body. Another potential application of ^{18}F-FDG PET/CT can be for the assessment of therapeutic response in IgG4-RD.[12] Unfortunately, in this present, patient post therapy PET/CT was not available.

CONCLUSION

^{18}F-fluorodeoxyglucose PET/CT can play an important role in the management of IgG4-related retroperitoneal fibrosis by supporting the diagnosis, demonstrating the disease extent, helping the selection of appropriate biopsy site and possibly for monitoring therapeutic response.

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