Clinical note

FDG PET/CT in monitoring treatment of retroperitoneal fibrosis

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A B S T R A C T

Retroperitoneal fibrosis is an uncommon disease characterized by inflammatory fibrosis typically surrounding abdominal aorta and iliac arteries. The glucose analogue F18-fluorodeoxyglucose can be used to image inflammatory cell activity non-invasively by PET. In this report we investigated the usefulness of the FDG PET/CT in the disease activity and therapy response evaluation of retroperitoneal fibrosis.

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FDG PET/TAC para monitorizar el tratamiento de la fibrosis retroperitoneal

R E S U M E N

La fibrosis retroperitoneal es una enfermedad poco común caracterizada por fibrosis inflamatoria tipicamente en el área de la aorta abdominal y las arterias ilíacas. El análogo de la glucosa F18-fluorodeoxyglucosa puede usarse para obtener imágenes de la actividad celular inflamatoria de manera no invasiva por PET. En este informe, estudiamos la utilidad de FDG PET/TAC en la evaluación de la actividad de la enfermedad y respuesta al tratamiento de la fibrosis retroperitoneal.

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Retroperitoneal fibrosis (RPF) is a rare inflammatory fibrotic process leading to compression of retroperitoneal structures. Although the exact pathogenesis of retroperitoneal fibrosis has not been definitively described, evidence supports that fibrosis often begins around severe atherosclerotic plaques.1 Usually, disease affects people between the ages of 40 and 60 years and half to two-thirds are men.2,3 RPF most commonly causes increasing dull abdominal or back pain and lower extremity swelling and discomfort as early symptoms. Late symptoms are usually result of mass effect including decreasing urine output, uremia, mesenteric ischemia, bowel obstruction, deep venous thrombosis and hypertension.4 Prompt diagnosis of RPF improves chances of preserving renal function.4 The diagnosis is based on radiological findings, primarily CT and magnetic resonance imaging (MRI). CT has the ability to evaluate the extent of fibrosis, associated complications and also possible underlying cause such as an abdominal aortic aneurysm, malignancy or inflammatory process.2 On CT images, RPF usually appears as a bulky, well-defined retroperitoneal mass which is isodense to surrounding muscle. On T1-weighted MRI, retroperitoneal fibrosis demonstrates diffusely low signal intensity, while T2-weighted signal can change depending on the disease stage and underlying inflammation and edema.2 Excretory urography was performed before the widespread use of CT and MRI but it is not routinely used anymore because of its limited sensitivity and specificity. The presence of classic triad of delayed renal contrast excretion and associated hydronephrosis secondary to ureteral involvement, medial deviation of the middle third of both ureters and tapering of the ureteral lumen at the level of L4–L5 vertebra was needed to make a diagnosis of RPF by excretory urography.2 Immunosuppressive therapy and/or hormonotherapy appears to be the treatment of choice.4 Determining disease activity in these patients consist of a crucial point in indicating potential response to medical treatment. Currently, PET/CT imaging has been widely used for inflammatory diseases. Here, we report the value of PET/CT in evaluating disease activity and therapy response in a case of retroperitoneal fibrosis.

Case report

A 48-year-old man with retroperitoneal mass was admitted to urology department. An abdominal CT scan done prior to the PET study showed a mass with soft tissue density around paraaortic and right iliac artery. The patient showed an important reduction in the size of the retroperitoneal thickened mass at 12 months without any treatment. With clinical presentation and imaging findings the patient had the presumed diagnosis of RPF. Then, the patient was referred with a presumed diagnosis of RPF for PET/CT imaging for the metabolic characterization of the retroperitoneal mass. PET/CT images showed heterogeneously increased FDG uptake extending below the renal hilum and tracing along common iliac artery at the right and proximal common iliac artery at the left. There was no other pathological uptake in the body. The patient was treated

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Prompt diagnosis of RPF can provide preserving disease activity, and these parameters are frequently used to monitor the clinical course of RPF. However, these parameters are infrequently shows some residual masses that may represent active disease or may simply be inactive fibrous tissue. The potential role of FDG PET in the assessment of inflammatory disease as in RPF has been reported. PET/CT is a useful adjunct to anatomic imaging and serum inflammatory markers in evaluating the inflammation degree in RPF, and in assessing the response to immunosuppressive therapy.

Acute phase reactants like the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) may be helpful in evaluating disease activity, and these parameters are frequently used to monitor the clinical course of RPF. However, these parameters are nonspecific and not very sensitive. Additionally, despite clinical response and reduction of acute phase reactants, soft tissue abnormalities often present on CT following treatment. Activity of the fibrotic process has a positive correlation with the degree of soft-tissue enhancement on CT. Avid contrast enhancement may be seen in the acute stages. Unfortunately, administration of IV contrast agents cannot be performed in many patients with renal impairment secondary to obstructive uropathy. In such cases, FDG PET may be of benefit in evaluating disease activity. The perivascular inflammation seen on PET/CT suggests the potential for FDG PET to act not only as a diagnostic adjunct to anatomical imaging but also to provide prognostic information regarding the degree of active inflammation and the potential for response to medical treatment. The patients who show FDG avidity could be placed on a trial of immunosuppressive or hormonotherapy and followed to assess the degree of disease regression. If, however, there was no FDG uptake, the patient might be referred for operative management earlier in the disease course and thus spared of potential complications of unnecessary treatment. FDG PET may have an important role in managing RPF by helping to discriminate active from inactive disease.

Fig. 1. Pre-treatment (A) and post-treatment (B) axial PET, axial CT, axial fusion, coronal fusion and MIP PET images. Pre-treatment images showed heterogeneously increased FDG uptake extending below the renal hilum to the aortic bifurcation and tracing along common iliac artery at the right and proximal common iliac artery at the left (lesion SUVmax: 9.9, liver SUVmax: 2.8). No other sites of abnormal FDG metabolism were noted. The patient was treated with tamoxifen and follow-up PET/CT done five months after pharmacotherapy demonstrated FDG avid lesion at the same region (lesion SUVmax: 6.8, liver SUVmax: 3.0). When compared to previous study, partial response to therapy (decrease in SUV is 31.3%) was noted.

Discussion

Typically, RPF typically surrounds the abdominal aorta and iliac arteries extending to neighboring structures, most commonly the ureters and inferior vena cava. Corticosteroids, with addition of immunosuppressants or tamoxifen in resistant cases have been used to treat RPF. In most cases, it can be diagnosed on the basis of radiological findings, especially CT, with identification of a retroperitoneal mass, the absence of other demonstrable renal or ureretic disease or any other pathology that could explain the findings. Prompt diagnosis of RPF can provide preserving renal function. There are additional fibrotic processes outside the retroperitoneum in up to 15% of patients. Medical treatment is often effective for RPF, but follow-up CT after medical treatment frequently shows some residual masses that may represent active disease or may simply be inactive fibrous tissue. The potential role of FDG PET in the assessment of inflammatory disease as in RPF has been reported. PET/CT is a useful adjunct to anatomic imaging and serum inflammatory markers in evaluating the inflammation degree in RPF, and in assessing the response to immunosuppressive therapy. Acute phase reactants like the erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) may be helpful in evaluating disease activity, and these parameters are frequently used to monitor the clinical course of RPF. However, these parameters are nonspecific and not very sensitive. Additionally, despite clinical response and reduction of acute phase reactants, soft tissue abnormalities often present on CT following treatment. Activity of the fibrotic process has a positive correlation with the degree of soft-tissue enhancement on CT. Avid contrast enhancement may be seen in the acute stages. Unfortunately, administration of IV contrast agents cannot be performed in many patients with renal impairment secondary to obstructive uropathy. In such cases, FDG PET may be of benefit in evaluating disease activity. The perivascular inflammation seen on PET/CT suggests the potential for FDG PET to act not only as a diagnostic adjunct to anatomical imaging but also to provide prognostic information regarding the degree of active inflammation and the potential for response to medical treatment. The patients who show FDG avidity could be placed on a trial of immunosuppressive or hormonotherapy and followed to assess the degree of disease regression. If, however, there was no FDG uptake, the patient might be referred for operative management earlier in the disease course and thus spared of potential complications of unnecessary treatment. FDG PET may have an important role in managing RPF by helping to discriminate active from inactive disease.

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