18F-FDG-PET/CT imaging features at initial diagnosis and in the context of early treatment monitoring in IgG4 related Pulmonary Inflammatory Pseudotumor

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Abstract

The $^{18}$F-FDG-PET/CT imaging features in pulmonary inflammatory pseudotumor are illustrated, in a proven case both at initial diagnosis and in the context of early monitoring of corticosteroid therapy in this “great mimicker of malignancy”. Complete surgical resection had to be finally undertaken in view of relative non-response to corticosteroid therapy advocated first line, though the patient had shown some symptomatic response and modest reduction of SUVmax. $^{18}$F-FDG avidity in untreated cases of the pulmonary inflammatory pseudotumor has been quite characteristic and may be potentially utilized to evaluate early response to administered non-surgical treatment and also detect residual disease/recurrence following therapeutic intervention of this IgG4 related disease.
Introduction:

Pulmonary inflammatory pseudotumor is a rare related disease entity characterised by non-neoplastic proliferation of inflammatory cells. It is a close mimic of lung malignancy (1) and can at times demonstrate excellent response to corticosteroideal therapy (2), the usually employed first-line therapeutic agent for induction of remission for the IgG4-related diseases. The application of Fluorodeoxyglucose ($^{18}$F-FDG)-positron emission tomography PET-CT has been described in literature only in the recent years (1).

Case Report

The patient, a 12 year old boy, presented with symptoms of dry cough and right sided chest pain of 8 months duration. Chest radiograph (Figure 1) showed right mid zone mass lesion and contrast enhanced computed tomography (CT) of chest (Figure 2) showed heterogeneously enhancing mass with calcification in right middle lobe with obstruction of right middle lobe bronchus (Figure 3). CT guided biopsy of the mass showed immunoglobulin (IgG4) expressing plasma cell infiltrate consistent with inflammatory pseudotumor. For further confirmation serum IgG4 levels were ordered which were significantly elevated 12.1 gm/l (normal range: 0.049-1.985g/l). $^{18}$F-FDG-PET/CT (Figure 4a) showed metabolically active soft tissue lesion with intense $^{18}$F-FDG uptake (SUVmax 7.7) with coarse calcification in the right middle lobe. Patient was started on steroid therapy and was reassessed at 6 weeks. He showed clinical response in form of subsidence of symptoms however there was no radiological resolution. Repeat PET-CT (Fig 4b) showed some reduction in $^{18}$F-FDG uptake (SUVmax 4.4) and the patient continued on
steroid but demonstrated no further improvement. The patient underwent surgical excision at 6 months following the last scan: a firm to hard tumour was identified measuring 6 X 5 X 4.5 cm abutting the overlying pleura. On immunohistochemistry, the ratio of IgG to IgG4 was almost 1:1. The findings were consistent with IgG4 related disease involving lung. The case presented highlights that IgG4 related IPT can be intensely ¹⁸F-FDG avid akin to lung malignancy and reduction in ¹⁸F-FDG uptake following steroid therapy can be observed early in the disease course.

**Discussion:**

Inflammatory pseudotumours, though commonly described in lungs, can involve other organs also such as liver, spleen, kidney, heart, brain, lymph nodes, salivary glands, breast, soft tissues and skin, mediastinum, mesentery, trachea and bronchii, orbit, sinonasal cavity and other organs of genitourinary and gastrointestinal tract (urinary bladder, epididymis, stomach, small and large intestine and esophagus) (1,3,4,5,6). Involvement of multiple organs has also been rarely reported (5). These tumor-like reactive masses of unknown etiology are now classified under the umbrella of immunoglobulin G4-related diseases (1). Pulmonary inflammatory pseudotumours primarily occur in young patients, with more than half occurring below 40 years (3,5). Immunoglobulin G4 (IgG4) -related diseases are a group of immune-mediated disorders characterized by mass forming fibro-inflammatory lesions involving multiple organs or locations (3); the characteristic histopathological features include dense
lymphoplasmacytic infiltrate, storiform pattern of fibrosis and obliterative phlebitis (3). The consensus suggests advocating histopathology as the primary characteristic and tissue IgG4 counts and IgG4:IgG ratios as the secondary characteristics for establishing the diagnosis (3). Non-operative treatment options e.g. corticosteroids and radiotherapy are sometimes employed, though surgical resection has been described as the best treatment with excellent long term prognosis with complete resection of the mass.

In a previous case report by Huellner et al, the lesion also demonstrated intense \(^{18}\text{F-FDG}\) uptake in the pulmonary lesion at the initial diagnosis and the patient had undergone surgical excision at the first instance in view of inconclusive diagnosis on bronchoscopic biopsy (7). Thus, the present report could be considered as an initial endeavour to assess treatment response to corticosteroid therapy with \(^{18}\text{F-FDG PET/CT}\).

**Conclusion:**

As described in the present communication, \(^{18}\text{F-FDG avidity}\) in untreated cases of the PIPT has been quite characteristic and uniformly observed and may be potentially utilized to evaluate early response to administered non-surgical treatment and also detect residual disease/recurrence following therapeutic intervention.
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Figure 1: Chest radiograph showing right midzone mass lesion.
Figure 2: Contrast enhanced computed tomography of chest showing heterogeneously enhancing mass with calcification in right middle lobe.
Figure 3: Histopathology with immunohistochemistry of CT guided biopsy showing IgG4 expressing plasma cell infiltrate.
**Figure 4a**: Baseline PET-CT showing metabolically active lesion with calcification in right middle lobe.

**Figure 4b**: Follow up PET-CT at 6 weeks following initiation of corticosteroid therapy showing decreased uptake in previously active areas.
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