The Value of FDG PET/CT Imaging in Dermatomyositis As A Paraneoplastic Syndrome in Malignancy Suspicion

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Citation

Abstract
Dermatomyositis (DRM) is a polymyositis accompanied by skin inflammation. It may be seen in various types of cancers as a paraneoplastic syndrome. A 45-year-old male presented with DRM which F-18 flourodeoxyglucose PET/CT (FDG PET/CT) revealed intense hypermetabolic nodule in the right lung and multiple mediastinal LAPs which were previously underestimated in diagnostic CT. The nodule was later biopsied and reported as lung cancer. After chemotherapy treatment the typical skin findings for DRM (Paraneoplastic Syndrome) was disappeared. FDG PET/CT seems to be a usefull method for cancer screening in DRM patients.

INTRODUCTION
Dermatomyositis (DRM) is a clinical syndrome of unknown cause involving the skeletal and myocardial muscles. Five basic diagnostic criteria of DRM are; symmetrical proximal muscle weakness, abnormal muscle biopsy, increased skeletal muscle enzymes, abnormal electromyography and typical skin findings with or without dysphagia and shortness of breath (1). A recent population-based study from Mayo Clinic found the incidence of dermatomyositis to be 13.98 per million in women and 4.68 per million in men (2). DRM may be seen with primary rheumatologic syndromes and in various types of cancers as paraneoplastic syndrome. First, in 1916 Stertz showed the relationship between gastric cancer and DRM (3). It is thought that in patients with malignancy, DRM develops due to a reaction against cancer cells, but the pathophysiology of DRM could not be fully explained yet.

18-F flourodeoxyglucose PET/CT (FDG PET/CT) has been extensively used in tumor imaging recently. Although the role of FDG PET/CT in cancer screening is not well known, there are papers that recommended it as alternative to conventional methods. In this case report with a brief review of literature, we aimed to underline the importance of FDG PET/CT in cancer screening in DRM patients.

CASE REPORT
A 45-year-old male patient with ANA seropositivity and symptoms of DRM as a paraneoplastic syndrome was admitted to the dermatology service. His diagnostic thoracic CT revealed nodular lesion located in the superior-posterior segment of the lower lobe of the right lung without any other pathology. Whole body FDG PET/CT imaging was planned for the metabolic characterization of this nodule. FDG PET/CT demonstrated intense FDG uptake, not only in the suspected pulmornary nodular lesion (Fig.A, arrows), but also in multiple enlarged mediastinal lymph nodes (Fig.B, arrows).
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Additionally symmetric hypermetabolism in the proximal muscles of the upper and the lower extremities was seen in Maximum Intensity Projection (MIP) images (Fig.C, arrows) due to DRM which was later confirmed by muscular biopsy.

Non-small cell lung cancer (NSCLC) was verified by CT guided biopsy. The patient is still alive and after chemotherapy treatment the typical skin findings for DRM (Paraneoplastic Syndrome) was disappeared.

DISCUSSION

Dermatomyositis is a type of connective-tissue disease (CTD) characterized by muscle and skin inflammation. Cancer risk is greater than normal population in DRM patients and the risk is highest in the first year following the diagnosis (3-5). The most commonly reported tumors are ovarian cancer, breast cancer, melanoma and colon cancer (3). Other comorbid cancers commonly associated with DRM are cancers of the lungs/mediastinum, bone/joints and kidney, as well as lymphoma/leukemia and nasopharyngeal cancer (3, 5).

Limaye et al. searched mortality and its predominant causes in inflammatory myositis patients and they found that malignancies were the third most common cause of death in this group of patients (6). Adzic et al. investigated the clinical features of lung cancer in patients with CTD and
they concluded that the majority of CTD patients who developed lung cancer were diagnosed at advanced stage and had poor survival (7). Fardet et al. studied the factors associated with underlying malignancy in DRM patients (8). They found that the independent factors associated with an underlying malignancy in patients with DRM were; an age at diagnosis >52 years, a rapid onset of skin and/or muscular symptoms, the presence of skin necrosis or periungual erythema and a low baseline level of complement factor C4. However, low baseline lymphocyte count (<1500/mm3) was a protective factor of malignancy.

We believe that the evaluation should still begin with a careful history and physical examination and “standard” laboratory evaluation. Any abnormalities found should be thoroughly investigated. The recommended cancer screening in DRM patients includes CT of the thorax/abdomen in all patients, US of the pelvic region and mammography in women, US of testes in men under 50 years and colonoscopy in men and women over 50 (9). If CT-thorax is negative, FDG PET/CT is recommended for screening of the patients with paraneoplastic neurological syndromes (9).

O’Callaghan et al. concluded that the performance of FDG PET/CT, in a single imaging study, for occult malignant disease in patients with paraneoplastic neurological syndromes was comparable to that of broad conventional screening, which includes multiple tests (10).

In our case report, FDG PET/CT demonstrated symmetric hypermetabolism in the proximal muscles of the upper and the lower extremities (Fig. 1C, arrows) due to DRM which was later confirmed by muscular biopsy. Additionally, FDG PET/CT revealed intense hypermetabolic nodule (NSCLC was verified by biopsy) in the right lung and multiple mediastinal LAPs which were previously underestimated in diagnostic CT.

In conclusion; in patients with DRM, one has to be careful for the probability of coexistence of cancer and thus further studies should be performed to detect malignancy. FDG PET/CT seems to be a useful method for both cancer screening and revealing active involvement in this group of patients.

References
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