Thymoma and immunodeficiency: ^{18}F-FDG-PET/CT imaging of Good syndrome

To the Editor: "Good syndrome" is defined as the combination of thymoma, hypogammaglobulinemia, low numbers of peripheral B cells and variable defects in cell-mediated immunity with a CD4 T lymphopenia and an inverted CD4/CD8+ T-cell ratio [1]. Although thymomas associated with "Good syndrome" are mostly benign, this humoral immunodeficiency may be associated with high risk of metastatic spread thymoma. This report concerns a case of high risk thymoma associated with recurrent lower respiratory tract infections finally revealing "Good syndrome".

A 56 years old man, with no history of myasthenia or respiratory tract infection, was investigated for cough and chest pain. Computed tomography (CT) scan showed a 80x80x30mm mass in the anterior mediastinum suggestive of lymphoma or thymoma. This mass infiltrated the brachiocephalic trunk and the right pleura. An ^{18}F-fluorodeoxyglucose positron emission tomography/CT (^{18}F-FDG PET/CT) has been performed showing high uptake in the thoracic mass (SUVmax 8.6) associated with right and left hypermetabolic hilar lymph nodes suggesting high risk thymoma [2-4] (Fig. 1A). Surgery consisted of an enlarged thymectomy and lymphadenectomy. Histopathological examination showed a type B2 lymphoepithelial thymoma with capsular effraction.

The patient underwent radiotherapy on the mediastinum. In follow-up, the patient presented several lower respiratory tract infections treated with probabilistic antibiotherapy and corticosteroids.

A respiratory tract infection occured six months after the end of radiotherapy. Fluorine-18-FDG PET/CT showed a high uptake in bilateral ground glass opacities of the two lower lobes and hypermetabolic bilateral hilar lymph nodes (Fig. 1B). A new ^{18}F-FDG PET/CT scan was performed three months later attesting the migratory character of the lung opacities and allowed to rule out an unlikely metastatic spread. Complete blood count was normal but lymphocyte immunophenotyping showed significant decrease in B and T CD4 lymphocytes. Total IgG were slightly lower than the normal range (6.92g/L), with low IgG1 (4.68g/L) and IgM levels (0.34g/L), suggesting "Good syndrome". Tests for bacteriological, viral, fungal or parasitic infection performed repeatedly were negative. Antibiotics and gamma globulin injections (administered three years later) were required to treat these recurrent lower respiratory tract infections and to secure prophylaxis against pneumocystis carinii (trimethoprim-sulfamethoxazole). Immunodeficiency worsened whereas no recurrence of thymoma was observed after six years of follow-up.

Although the association of thymoma and immunologic disease is common and refers up to 74% of cases [5, 6], 6% to 11% of patients with a history of thymoma develop "Good syndrome" in the following years [7-9]. These patients are deemed to be at high risk for developing severe sinus or pulmonary infections with encapsulated bacteria, but also viral (in particular cytomegalovirus), parasitic (pneumocystis) or fungal (candida) infections [10]. Chronic diarrhea, of infectious origin or not, is common [11], cytomegalovirus retinitis and refractory gastrointestinal ulcers have also been reported [12, 13].

The diagnosis of thymoma preceded the diagnosis of hypogammaglobulinemia or infection in up to 42.4%, of the patients with an interval of 3 months to 18 years [14]. "Good syndrome" occasionally progresses even after thymectomy and/or corticosteroid treatment [15, 16]. To our knowledge, no imaging report of ^{18}F-FDG PET/CT findings of Good syndrome has been reported.

In conclusion, the association of thymoma, recurrent infectious episodes especially of the respiratory tract and hypogammaglobulinemia is suggestive of "Good syndrome". This diagnosis should be considered when ^{18}F-FDG PET/CT shows hypermetabolic lung opacities in a patient with a history of thymoma.

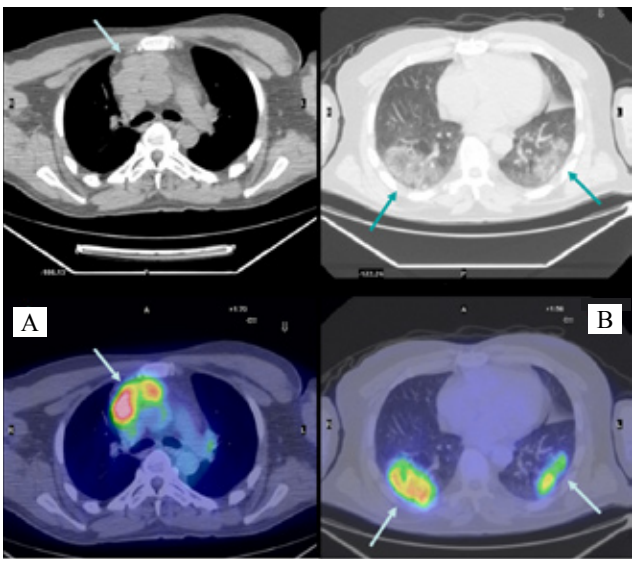

Figure 1. A. Preoperative ^{18}F-FDG-PET/CT. High uptake in the anterior mediastinal tumor. B. ^{18}F-FDG-PET/CT. High uptake in bilateral ground glass opacities of the two lower lobes.

The authors declare that they have no conflicts of interest.

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Antony Kelly[1] MD, Charles Merlin[1] MD, Sébastien Trouillier[2] MD, Florent Cachin[1] MD, PhD, Gaëlle Guettrot-Imbert[3] MD

1. Department of Nuclear Medecine, Jean Perrin Center, 58 rue Montalembert, 63011 Clermont-Ferrand, France.
2. Department of Internal Medicine, Centre Hospitalier Henri Mondor, 50 Avenue de la République, 15002 Aurillac cedex, France.
3. Department of Internal Medicine, Regional Competence Group on Adults' Auto immune and Rare systemic diseases, G. Montpied Hospital, 63003 Clermont-Ferrand Cedex 1, France.

Antony Kelly MD
Department of Nuclear Medecine, Jean Perrin Center, 58 rue Montalembert, 63011 Clermont-Ferrand, France. Tel: +33473278081, Fax: +33473278078, E-mail: antony.kelly@cjp.fr

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