Clinical note

Pulmonary intravascular lymphoma detected by FDG PET-CT: A case report


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ABSTRACT

Intravascular lymphoma is a rare subtype of extranodal Non-Hodgkin’s lymphoma. Its prognosis is poor in a high percentage of cases due to its insidious appearance and low clinical suspicion. Its diagnosis is usually only reached after an autopsy. It may affect different organs as a whole or only one organ. It is extremely rare that the lung is the only damaged organ. Its diagnosis depends on the clinician’s suspicion and proper evaluation with imaging studies as well as correct selection of the organ to be biopsied. When detected on time, the treatment of choice is a combination of a series of chemotherapy associated to a monoclonal antibody (anti-CD20). We present the case of a male patient who underwent a positron emission tomography-computed tomography with 2-[F-18]-fluoro-2 deoxy-D-glucose (FDG) due to symptoms suggestive of a lymphoproliferative disease with no clear structural abnormalities. The images led to a diagnosis of pulmonary intravascular large B cell lymphoma.

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Pulmonar intravascular linfoma detectado mediante FDG PET-TC: a propósito de un caso

RESUMEN

El linfoma intravascular de células B grandes es un subtipo raro de linfoma no Hodgkin extranodal. Por su presentación insidiosa y bajo índice de sospecha reviste un mal pronóstico en una gran proporción de casos, siendo diagnosticado durante la autopsia. Puede afectar a diversos órganos en su conjunto o de forma aislada, siendo el compromiso pulmonar único una forma de presentación extremadamente rara. Su diagnóstico depende de la sospecha del médico clínico y de una adecuada valoración mediante estudios de imágenes y la correcta selección del órgano a biopsiar. Si se detecta a tiempo, es tratado con una combinación de quimioterapia asociada a un anticuerpo monoclonal (anti-CD20). En esta nota clínica, exponemos el caso de un paciente que presentaba un síndrome de anemia y pérdida de peso, con una tomografía por emisión de positrones con 2-[F-18]-fluoro-2 deoxy-D-glucosa (FDG) para sospecha de enfermedad linfoproliferativa con estudios anatómicos sin alteraciones evidentes, se le diagnostica un linfoma intravascular pulmonar de células B grandes.

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INTRODUCTION

Large B-cell intravascular lymphoma is a rare subtype of extranodal Non-Hodgkin lymphoma (NHL) characterized by the malignant proliferation of lymphoid cells in small arteries, veins and capillaries and representing less than 1% of all lymphomas. The rapid progression of this disease is translated into a bad prognosis.

Intravascular lymphoma is not predominant in either sex and is usually presented at an average age of 70 years. The Symptomatology is poor and unspecific and may only present with fever of unknown origin or more floridly with B symptoms (fever, nocturnal sweating and weight loss), anemia and an increase in LDH. Diagnosis is achieved by the finding of atypical lymphoid cells, CD20+, within the arterioles, small veins and capillaries of the organs affected. Treatment consists of combined multiple drug chemotherapy (CHOP) associated with anti-CD20 monoclonal antibodies (Rituximab®).

The organs most commonly affected are the skin and the central nervous system, followed in order of frequency by organs such as the lung, kidneys, adrenal glands and the prostate. Solitary and primary involvement of the lung is rare, with the diagnosis often being obtained post mortem due to the difficulties in its detection. Imaging methods may demonstrate different patterns of involvement of the affected organs but, on occasions, do not demonstrate any disease, particularly with the methods which only provide anatomical analysis (computed tomography [CT], ultrasound, etc.). In these latter cases, functional imaging (scintigraphy) or hybrid (PET-CT) techniques in nuclear medicine may be very useful for demonstrating organ involvement, despite no appearance of morphological changes. Simultaneously, these methods provide a more complete and sensitive scan of the whole body allowing better staging.
We describe the diagnosis of a large B-cell intravascular lymphoma at a pulmonary level by PET-CT with FDG in a patient with B symptoms, thrombocytopenia, raised serum LDH and scarce unspecific findings in conventional imaging studies.

Clinical case

A 71-year-old male was attended for weight loss (9 kg) of 3 months evolution with hyporexia, nocturnal sweating and dry cough without fever.

Physical examination was anodyne. Biochemical studies showed: hemoglobin: 12.2 g/dl, hematocrit: 37.2%, white blood cells: 4100 cells/mm$^3$ (50% neutrophils), platelets: 80,000 platelets/mm$^3$, reticulocytes: 1.9%, total bilirubin: 1.35 mg/dl, direct bilirubin: 0.37 mg/dl, alkaline phosphatase: 334 IU/l, aspartate aminotransferase: 44 IU/l, alanine transferase: 41 IU/l, lactate dehydrogenase: 2670 IU/l and normal protein values.

Bone marrow biopsy was performed, showing a cellularity of 30%, a myeloerythroid relationship of 3:1, dyserythropoietic changes in red cells, myeloid series with a slight deviation to the left and 1% CD34+ cellularity. Negative fibrosis. No phenotypic abnormalities compatible with the lymphoproliferative syndrome were observed on flow cytometry.

A CT of the chest, abdomen and pelvis was requested, with oral and endovenous contrast, showing homogeneous splenomegaly as the only finding.

On persistence of the symptomatology a PET-CT with FDG was requested to localize the disease. The patient presented with 8 h of fasting and glycemia of 92 mg/dl prior to the study. A dose of 377.77 MBq (10.21 mCi) was injected one hour prior to the study. For opacification of the intestinal loops the patient ingested one liter of water dissolved with 30 ml of iodine, and during the tomography 70 ml of non-ionic contrast were injected. The study showed an increase in diffuse metabolic activity in both pulmonary parenchyma accompanied by a subtle increase of bilateral parenchymous attenuation (diffuse) (Fig. 1A). Homogenous splenomegaly and discrete diffuse uptake of the radiotracer was also observed in the bone marrow (Fig. 1B). The SUVmax of the lung (left base), the spleen and bone marrow (L2) was of 6.0, 3.4 and 3.6, respectively.

In view of these findings a bronchoalveolar lavage was performed which was normal. A slight increase in the number of small lymphocytes was of note. Finally, surgical biopsy was performed demonstrating pulmonary tissue with preserved structure which showed thickened septa in some sectors by the presence of atypical cells with large positive nuclei with lymphoid markers and CD20 in the interior of the capillary vessels. Some medium sized vessels were also affected (Fig. 2). The anatomopathologic result was a large B-cell CD20+ intravascular lymphoma.

Discussion

In our search of the literature this is the second clinical case in which intense uptake of the radiotracer in the lung during the performance of a PET-CT allowed correct diagnosis of a large B-cell intravascular lymphoma.6

This disease is difficult to diagnose because of the unspecific nature of the symptoms. This difficulty to suspect the diagnosis facilitates disease progression and explains its high mortality,
although if detected early, adequate chemotherapy treatment may be implemented to improve the survival, and, in some cases, even achieve complete disease remission.\textsuperscript{3}

Single or predominant involvement of the lung is extremely rare within this subtype of lymphomas,\textsuperscript{1} making the diagnosis even more difficult due to the low index of clinical suspicion (considering diseases of low prevalence). In our case, the detection of the same was due mainly to the use of PET-CT as a diagnostic tool in a patient with symptoms suggestive of a lymphoproliferative process. The utility of this imaging method in the staging and restaging of this disease in other localizations has already been established and reported by different authors.\textsuperscript{3,7}

The definitive diagnosis of the disease is anatomopathological. Nonetheless, in some cases, when the clinical deterioration becomes urgent, some authors propose the initiation of chemotherapy for diagnostic suspicion, even before having the definitive anatomopathological diagnosis.\textsuperscript{8} This did not occur in our case because of the good clinical condition of the patient, despite the persistence of the symptoms.

Treatment is based on a combined chemotherapy schedule (CHOP) in addition to a monoclonal antibody (Rituximab\textsuperscript{8}), which has shown better results.\textsuperscript{8}

The patient is currently under chemotherapy regime according to the recommendations in the literature awaiting a new control on completion of the treatment.

References