

Lymphomatoid granulomatosis

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Section: Chest imaging

Area of Interest: Adrenals Lung Thoracic wall Kidney

Thorax Neuroradiology brain

Procedure: Diagnostic procedure

Procedure: Comparative studies

Imaging Technique: CT

Imaging Technique: PET-CT

Imaging Technique: MR

Imaging Technique: MR-Diffusion/Perfusion

Special Focus: Neoplasia Case Type: Clinical Cases

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Patient: 71 years, male

Clinical History:

A 71-year-old male patient presented with a 6-month history of upper abdominal pain radiating to the back, fatigue and 10 kg weight loss.

Imaging Findings:

A thoracoabdominal CT (Fig 1) revealed bilateral heterogeneous adrenal masses, multiple bilateral pulmonary nodules and a soft-tissue lesion next to 10th dorsal vertebrae, without bony injury.

The body PET-CT (Fig 2) performed for screening of primary neoplasm showed increased uptake of FDF in both adrenal masses, thoracic soft-tissue lesion and diffusely in pulmonary fields. A focal increased uptake was seen in right renal cortex.

Follow up PET-CT (Fig 3) performed 3 months later showed enlargement of pulmonary nodules and right adrenal mass. The hypermetabolic focus of right renal cortex increased significantly, corresponding to a focal cortical lesion visible on CT. Several foci of increased uptake were seen in brain parenchyma.

Cranial MRI (Fig 4) showed two large right occipital and left temporo-occipital lesions with large surrounding oedema without significant mass effect, ring enhancement, intralesional peripheral haemorrhage, low ADC values and low perfusion.

Left laparoscopic adrenalectomy finally revealed the diagnosis of lymphomatoid granulomatosis.

Discussion:

Lymphomatoid granulomatosis (LYG) is a very rare B-cell extranodal lymphoproliferative disorder. There is an angiocentric and angiodestructive accumulation of atypical B-cell lymphocytes infected by Epstein-Barr virus (EBV) and reactive T-cell lymphocytes. Lymphoid cells directly accumulate within affected tissues in the form of infiltrative nodular lesions [1, 3].

LYG is closely related to host's underlying immune system. Although most patients do not have a preexisting

diagnosis of an immunodeficiency, evidence of immune dysregulation can be found in almost all. Patients may have a history of recurrent infections, autoimmune illnesses, other lymphoproliferative disorders or be under immunosuppressive treatment [1]. LYG generally presents in men between the third and fifth decades of life, although patients can be affected at any age [4].

Constitutional symptoms such as weight loss, fever and fatigue are present in most patients.

Lungs are virtually always involved in LYG, characteristically in the form of multiple bilateral pulmonary nodules of variable size in a bronchovascular distribution, mainly in mid and lower lung fields. In 30% cavitation caused by necrosis is seen. Marked accumulations of FDG are seen on 18F-FDG-PET/CT. Since LYG is an extranodal lymphoproliferative disorder, there is no hilar lymphadenopathy [1, 3].

CNS involvement occurs in 25-50% of cases, showing multiple focal asymptomatic lesions involving the white matter, deep gray matter or brainstem, characterised by punctate linear enhancement. Larger lesions may be solid or demonstrate ring enhancement. They have variable surrounding oedema and may be associated with leptomeningeal enhancement [1, 2].

Involvement of the kidneys (40-50%), focal nodular lesions without organ dysfunction, and skin (25%-50%), subcutaneous-dermal nodules and purplish eruptions that can ulcerate, is also frequently seen [1].

Histopathologic grading of LYG is performed depending on the number of EBV-positive atypical B-cells: grade I <5 lymphocytes in a highpower field, grade II, 6-19, grade III >20.

LYG has a poor prognosis, with mortality over 60% after 5 years. Management depends principally on the histopathologic grade. Patients taking immunosuppressive treatments should stop them. In low-grade cases a brief period of observation may be justified, as some will undergo spontaneous remission. High-grade LYG requires immediate therapy, generally with combined chemotherapy [1, 4].

The rareness of LYG and its nonspecific presentation contributes to delays in diagnosis. Considering that pulmonary manifestations are characteristic and almost always present, LYG should be included in the differential diagnosis of pulmonary processes that share those manifestations, so EBV-encoded RNA in situ hybridisation (ISH) can be ordered on biopsy specimens.

Differential Diagnosis List: Lymphomatoid granulomatosis, Granulomatosis with polyangiitis, Eosinophilic granulomatosis with polyangiitis, Tuberculosis, Sarcoidosis, Non-hodgkin lymphoma, Lung metastases

Final Diagnosis: Lymphomatoid granulomatosis

References:

- Roschewski M, Wilson WH (2012) Lymphomatoid Granulomatosis. *Cancer J* 18(5):469-474 (PMID:[23006954](#))
- Kano Y, Kodaira M, Ushiki A et al (2017) The Complete Remission of Acquired Immunodeficiency Syndrome-associated Isolated Central Nervous System Lymphomatoid Granulomatosis: A Case Report and Review of the Literature. *Intern Med* 56:2497-2501 (PMID: [28824078](#))
- Yamanouchi T, Kawanami S, Kamitani T et al (2016) Lymphomatoid Granulomatosis Two Different Phenotypes of Computed Tomography Findings. *J Thorac Imaging* 31:80-82 (PMID: [27768635](#))
- King TE (2017) Pulmonary lymphomatoid granulomatosis. In: UpToDate , Post, TW (Ed), UpToDate, Waltham, MA, Accessed on December 2017

Figure 1

a



Description: Bilateral heterogeneous adrenal masses, suspicious of malignancy. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

b



Description: MIP reconstruction of the lung showing pulmonary nodules. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

C



Description: MIP reconstruction of the lung showing pulmonary nodules. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

d



Description: Pulmonary nodules. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

e



Description: Pulmonary nodules. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

f



Description: Soft-tissue lesion next to 10^o dorsal vertebrae. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

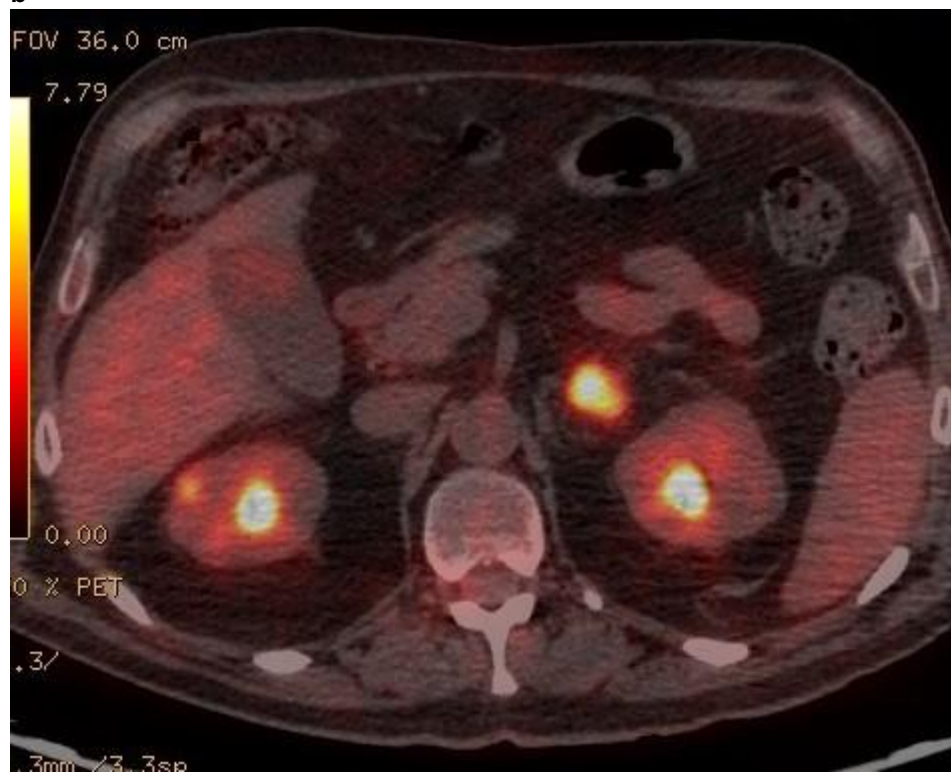
Figure 2

a



Description: Increased uptake of FDF in both adrenal masses. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain.

b



Description: Focal increased uptake in right renal cortex. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain.

c



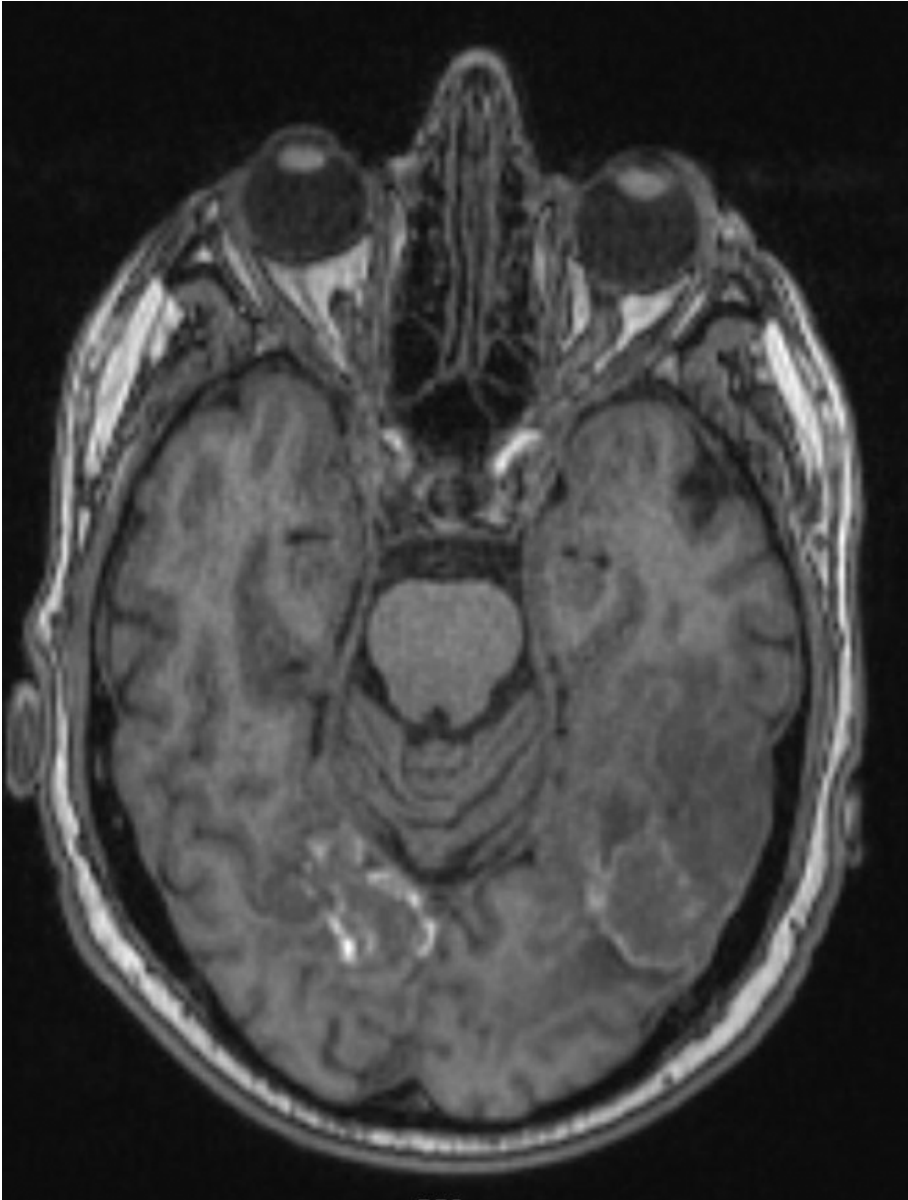
Description: Increased uptake of FDF in thoracic soft-tissue lesion. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain.



Description: Diffusely increased uptake of FDF in pulmonary fields. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain.

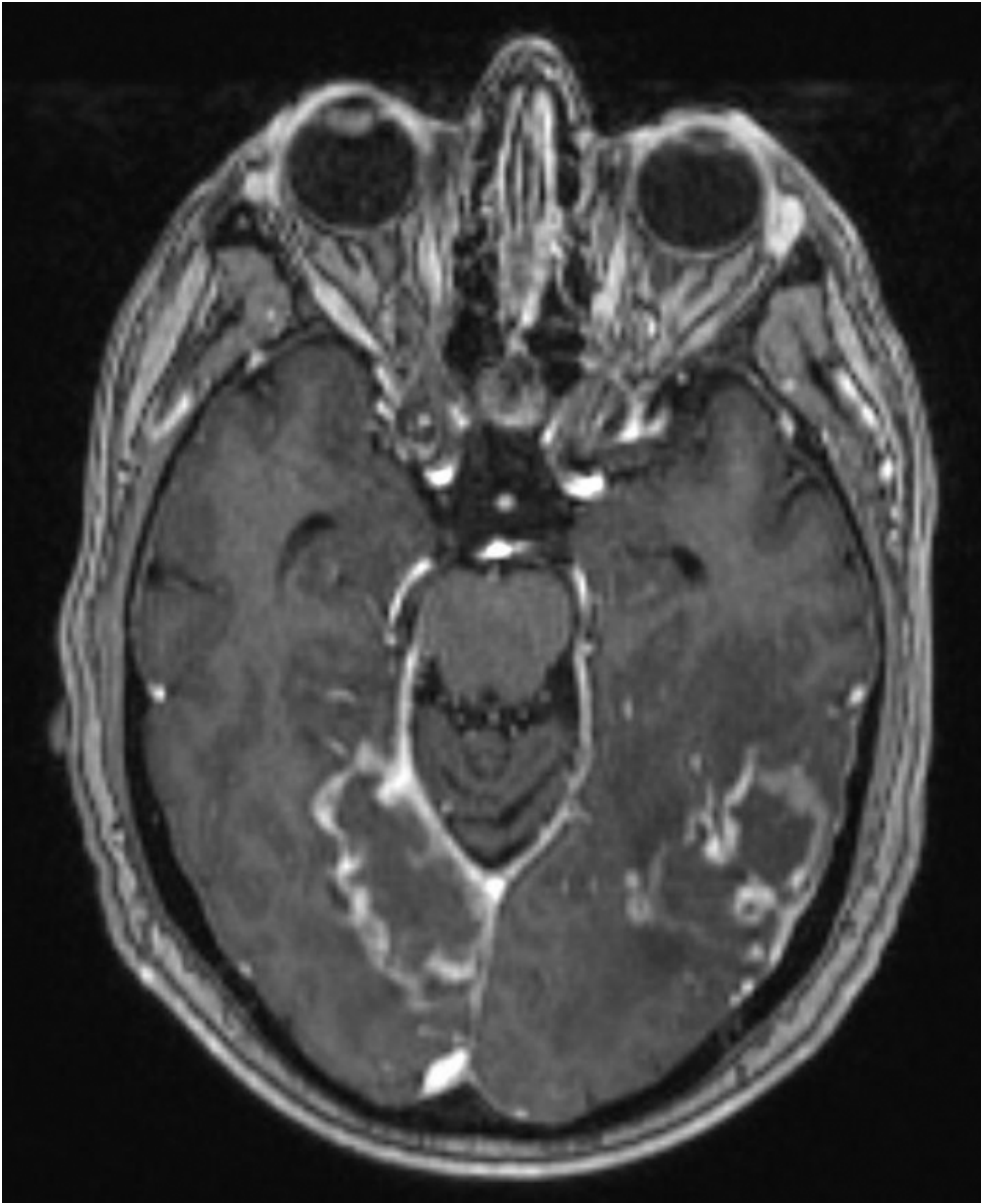
Figure 3

a



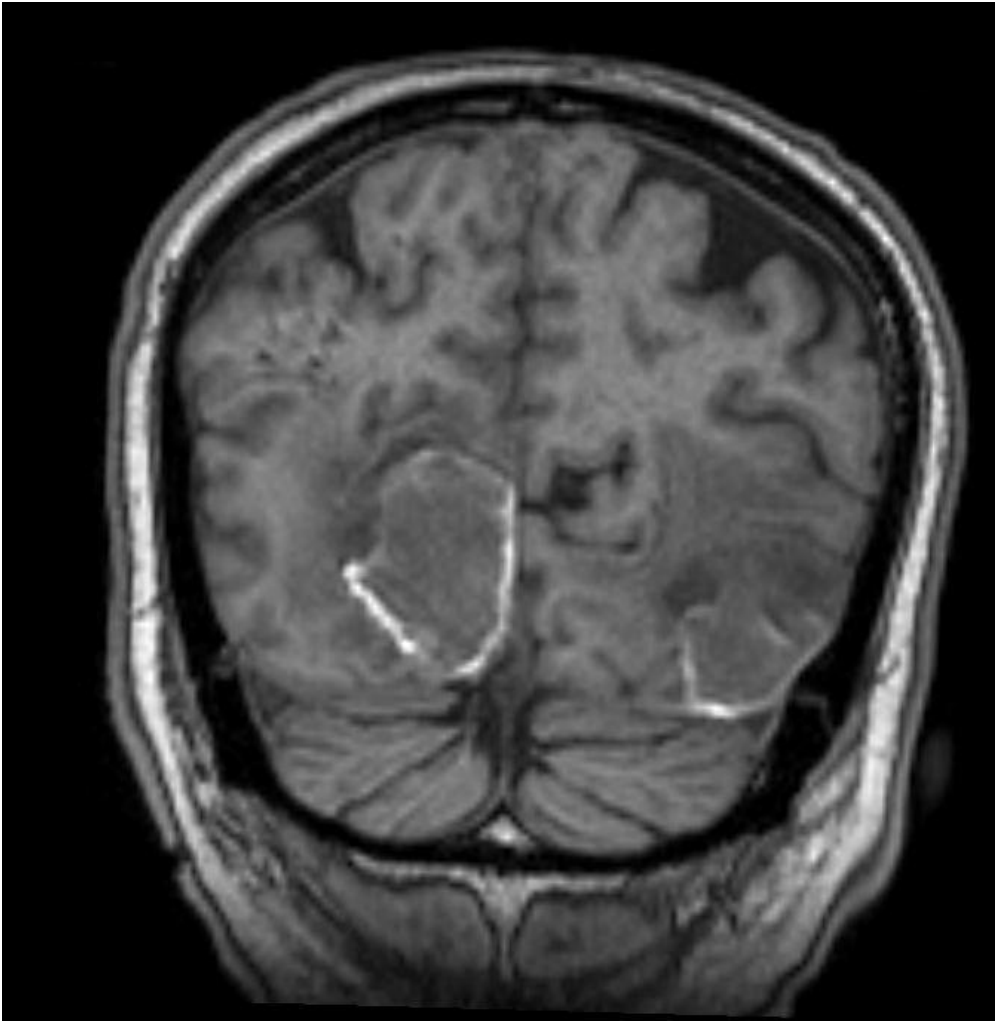
Description: T1. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

b



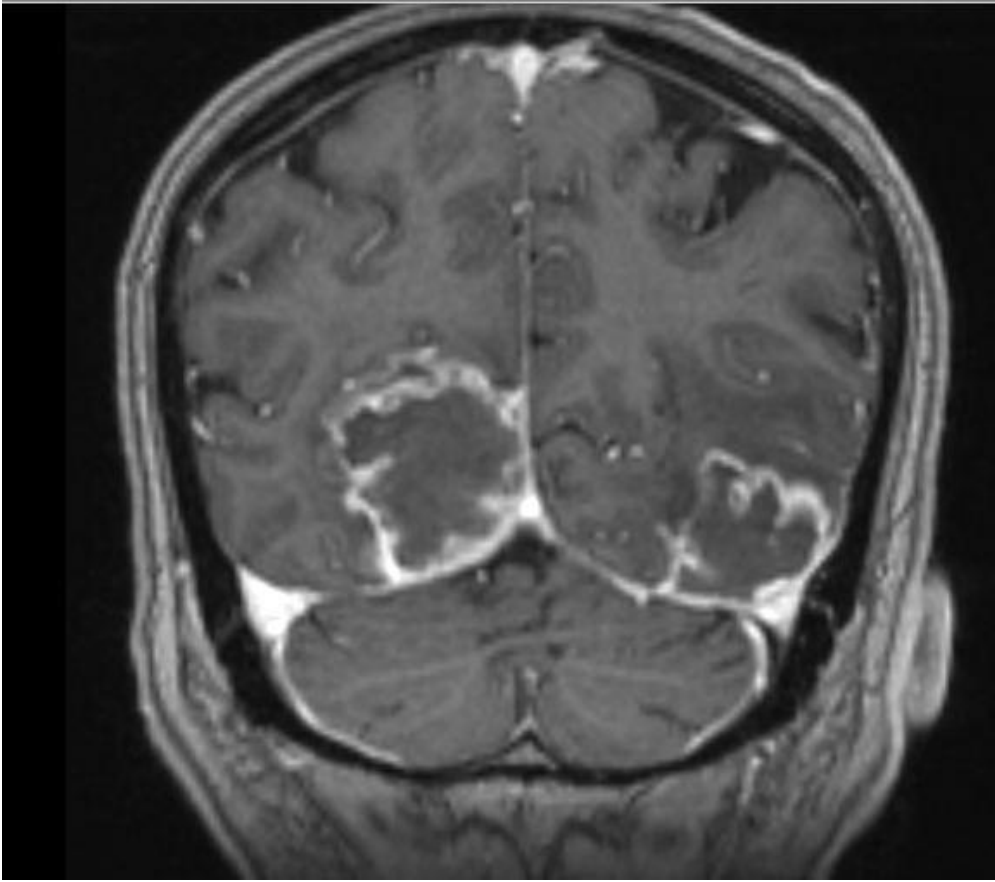
Description: T1 gadolinium. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

c



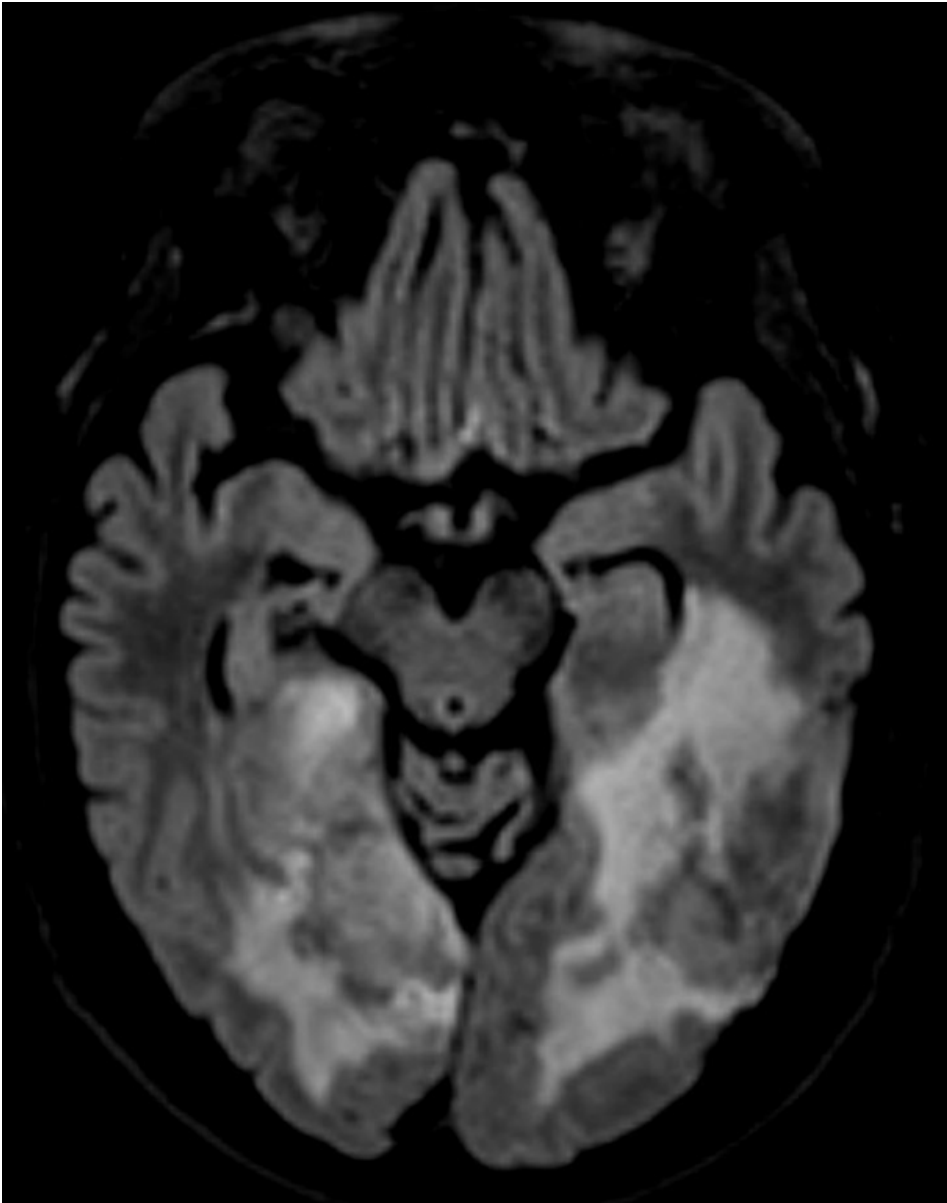
Description: T1 coronal. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

d



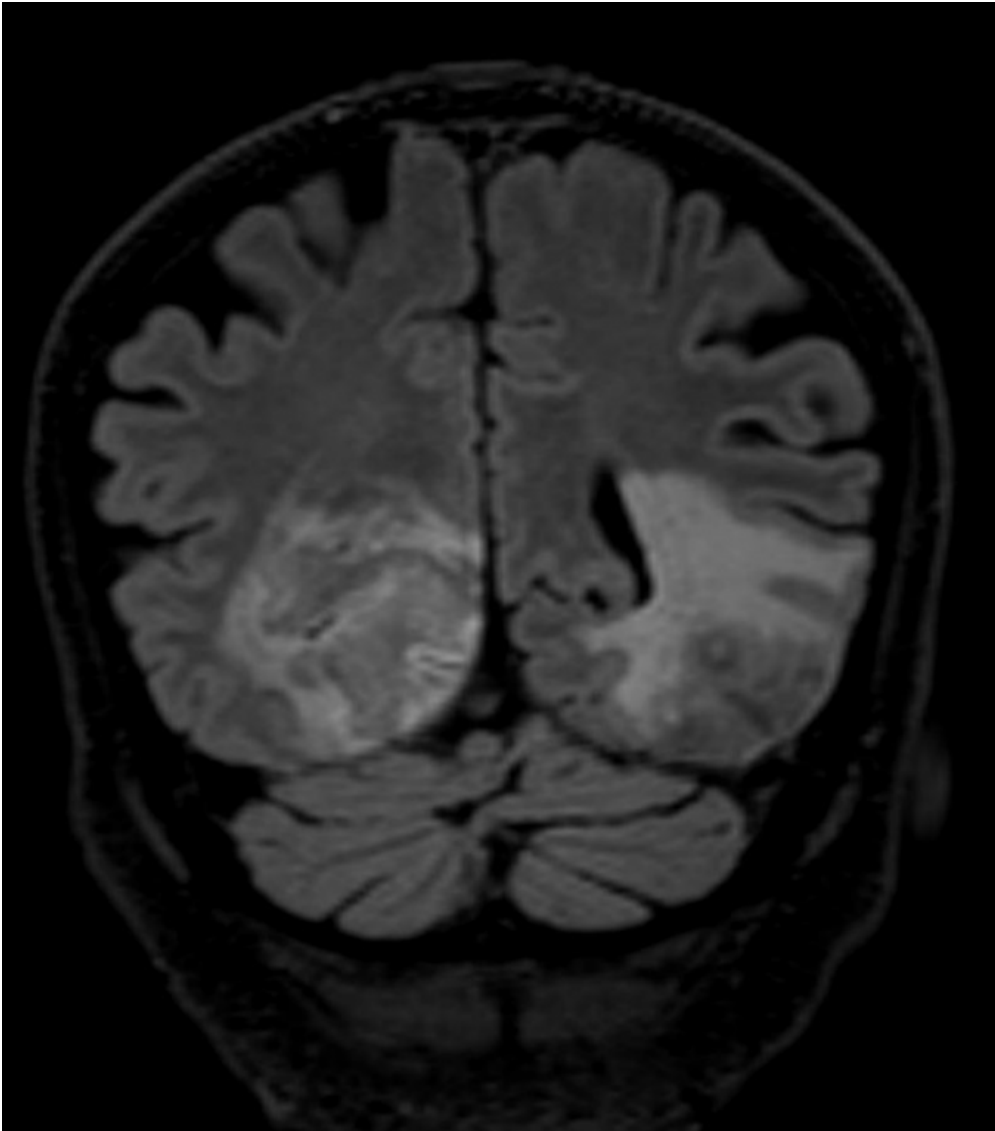
Description: T1 gadolinium coronal. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

e



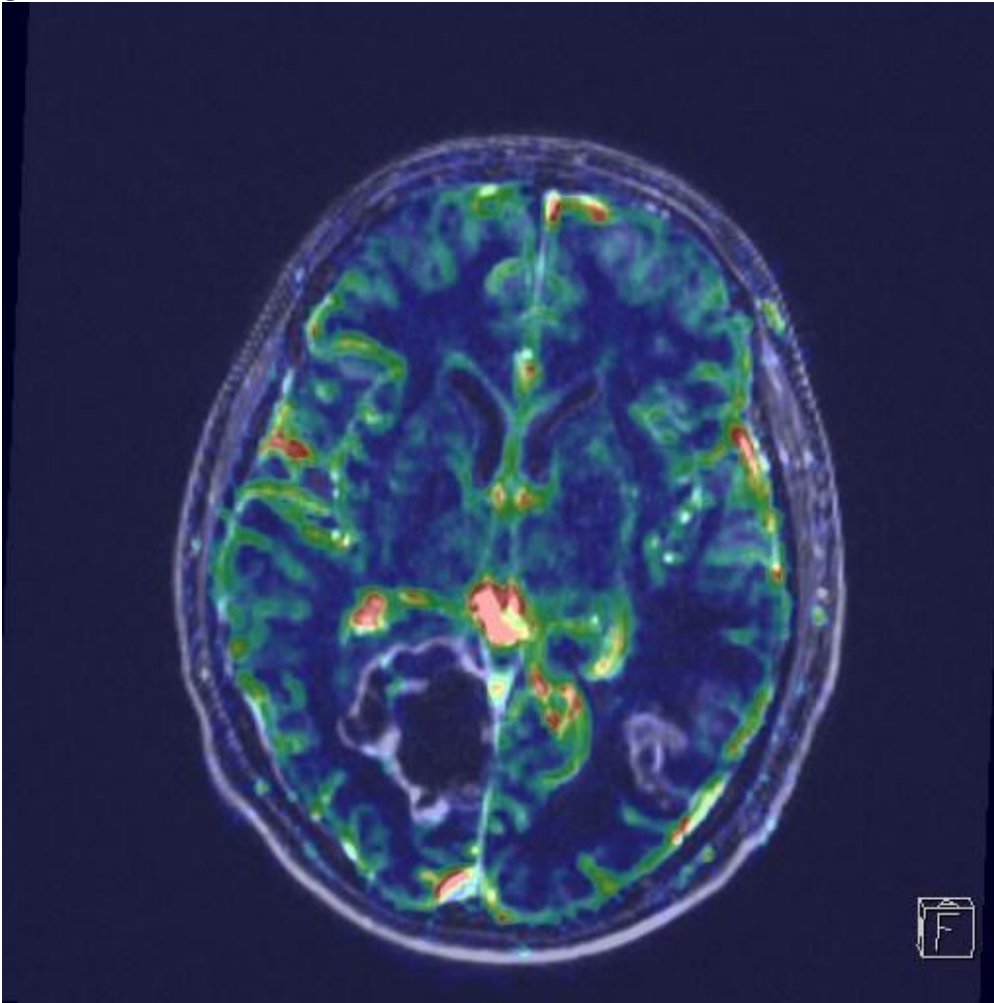
Description: FLAIR. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

f



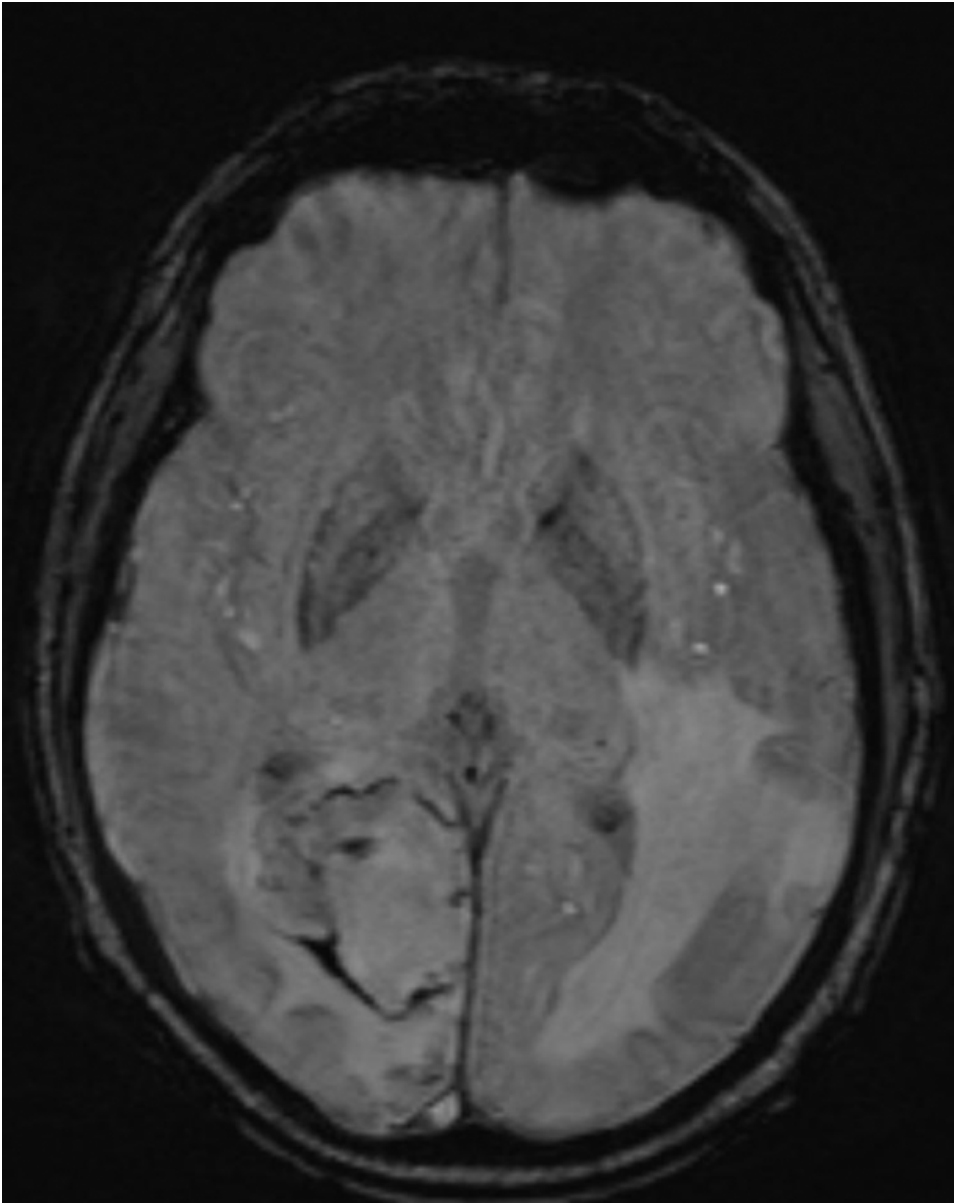
Description: FLAIR coronal. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

g



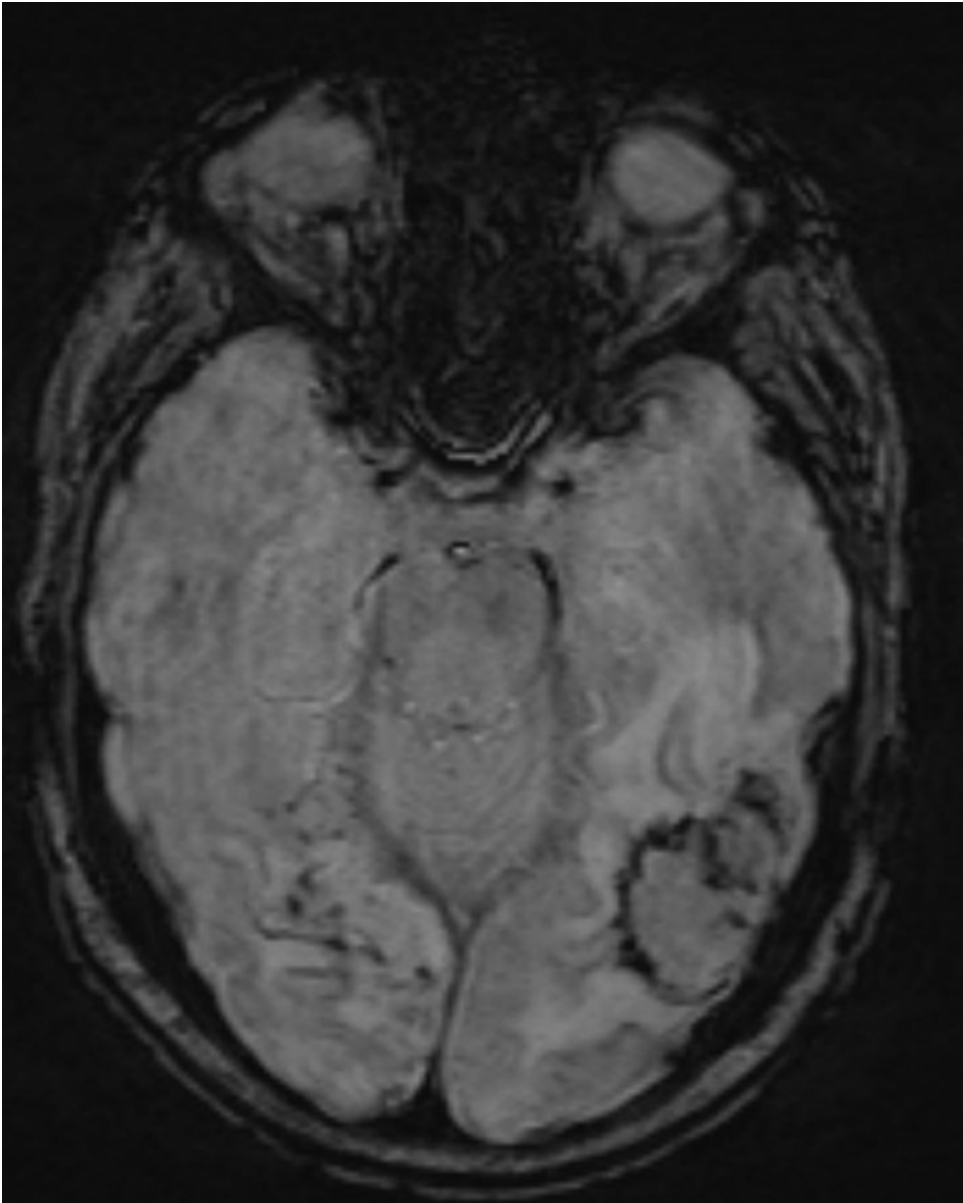
Description: Perfusion. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

h



Description: SWI. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

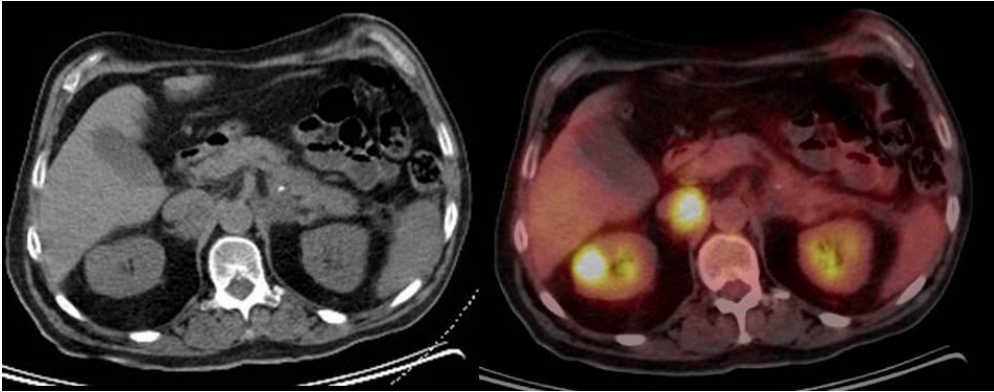
i



Description: SWI. **Origin:** Department of Radiology, Hospital of Basurto, Bilbao, Spain

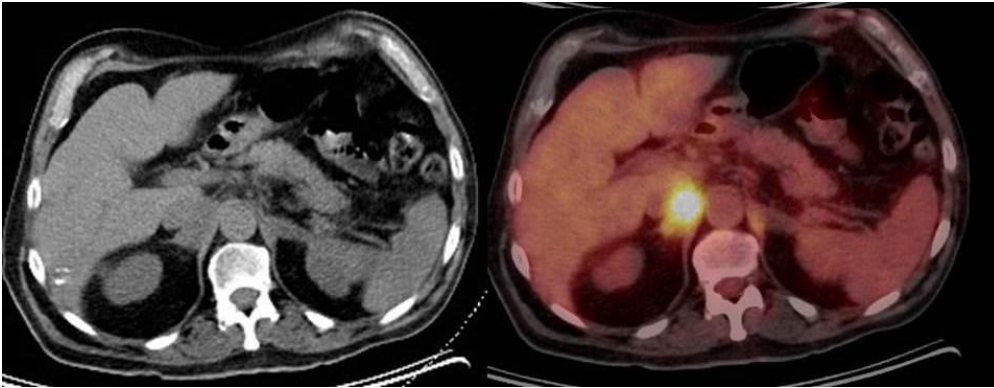
Figure 4

a



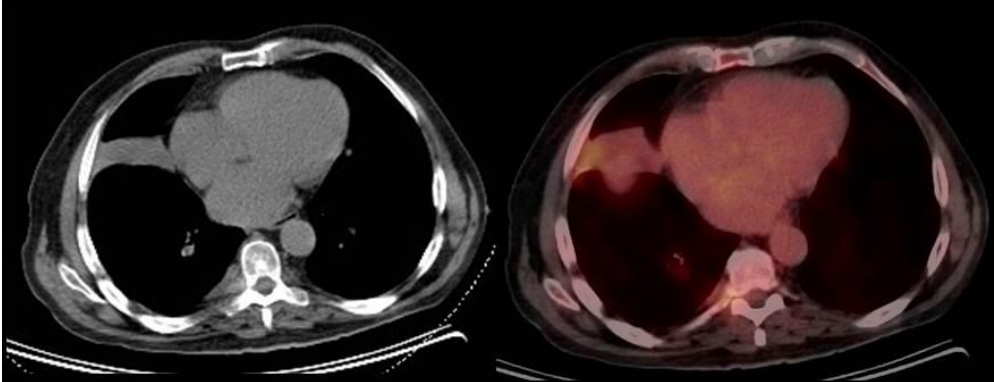
Description: Right renal cortex lesion. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain

b



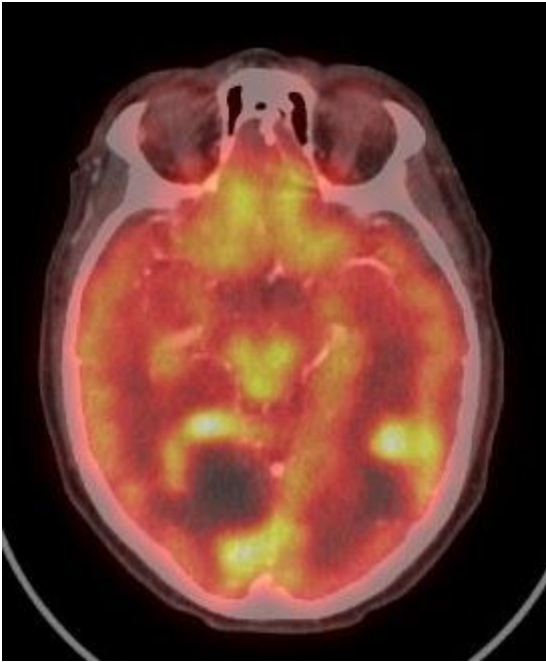
Description: Right adrenal mass. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain

c



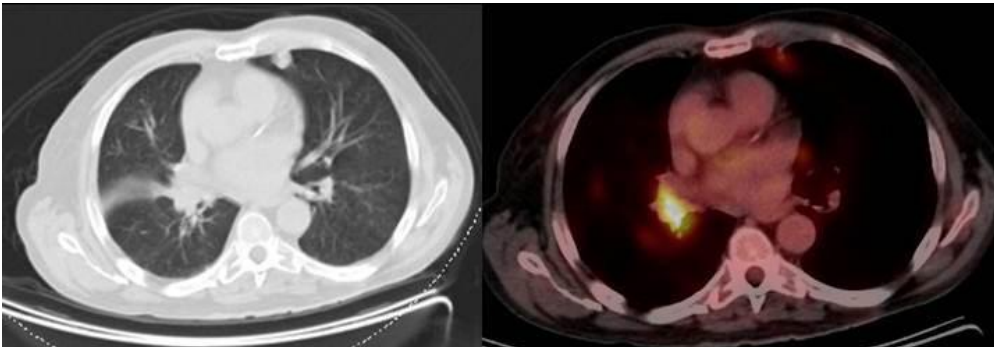
Description: Soft-tissue lesion. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain

d



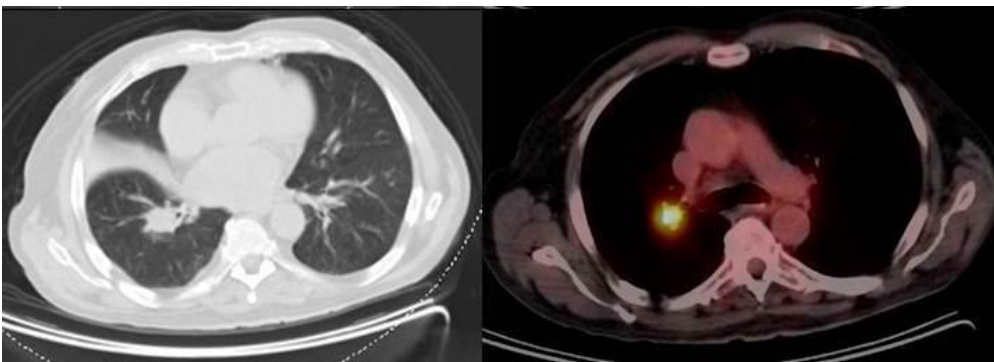
Description: Cerebral foci of FDG increased uptake. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain

e



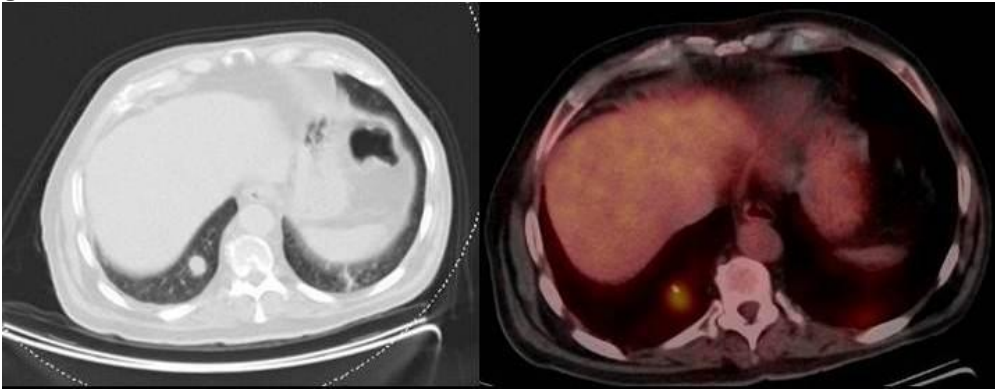
Description: Pulmonary nodules. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain

f



Description: Pulmonary nodules. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain

g



Description: Pulmonary nodules. **Origin:** Department of Nuclear Medicine, Hospital of Cruces, Spain