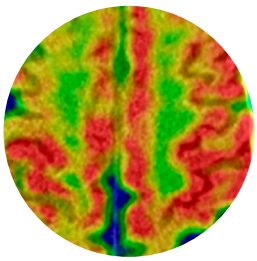

Contribution of the 18F-FDG PET/CT IN sporadic Creutzfeldt-Jakob Disease

Aporte de la 18F-FDG PET/TC en la enfermedad de Creutzfeldt-Jakob esporádica

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Summary

Sporadic Creutzfeldt-Jakob disease (sCJD) is an extremely rare transmissible neurodegenerative disorder characterized by rapidly progressive dementia. 18F-fluoro-2-deoxy-D-glucose positron emission tomography (^{18}F-FDG-PET/CT) in these patients has described bilateral parietal, frontal and occipital cortical hypometabolism, without alterations in the cerebellum or basal ganglia, which could contribute to the differential diagnosis of rapidly progressive dementia. We present the case of a 75-year-old man with a history of prostate cancer and bipolar affective disorder, with a two-week picture of behavioral and mood changes, cognitive deficit, visual and auditory hallucinations and spatial disorientation with rapid progression. Subsequently, the patient presented slow gait, tremor in lower limbs and right Babinski. Brain magnetic resonance imaging (MRI) showed diffusion restriction in the bilateral frontal and temporal cortex and cingulate gyrus, with diagnostic suspicion of paraneoplastic syndrome versus prion disease. ^{18}F-FDG-PET/CT showed hypometabolism in the bilateral frontal cortex and right temporal and parietal lobe. Measurement of 14-3-3 protein, T-Tau protein and real-time shake-induced prion protein conversion (RT-QUIC) in cerebrospinal fluid confirmed the diagnosis of prion disease.

Resumen

La enfermedad de Creutzfeldt-Jakob esporádica (ECJe) es un trastorno neurodegenerativo transmisible, extremadamente raro, caracterizado por demencia rápidamente progresiva. En la tomografía por emisión de positrones con 18F-fluoro-2-desoxi-D-glucosa (^{18}F-FDG-PET/TC) de estos pacientes se ha descrito hipometabolismo cortical bilateral parietal, frontal y occipital, sin alteraciones en el cerebelo ni en los ganglios basales, lo que podría contribuir con el diagnóstico diferencial de demencia rápidamente progresiva. Se presenta el caso de un hombre de 75 años de edad, con antecedente de cáncer de próstata y trastorno afectivo bipolar, con cuadro de dos semanas de cambios comportamentales y anímicos, déficit cognitivo, alucinaciones visuales y auditivas y desorientación espacial con rápida progresión. Posteriormente, el paciente presenta marcha lenta, temblor en miembros inferiores y Babinski derecho. La resonancia magnética (RM) cerebral mostró restricción a la difusión en la corteza frontal y temporal bilateral y giro del cíngulo, con sospecha diagnóstica de síndrome paraneoplásico versus enfermedad por priones. La ^{18}F-FDG-PET/TC demostró hipometabolismo en la corteza frontal bilateral y lóbulo temporal y parietal derechos. La medición de la proteína 14-3-3, proteína T-Tau y conversión de proteína priónica inducida por agitación en tiempo real (RT-QUIC) en líquido cefalorraquídeo confirmó el diagnóstico de enfermedad por priones.

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Introduction

Creutzfeldt-Jakob disease (CJD) is a prion encephalopathy characterized by rapidly progressive dementia that invariably leads to death (1, 2). The worldwide incidence rate is one to two cases per million people per year (3). Few cases have been reported in Colombia since its first description in 1973 (4). We present the case of a patient with Creutzfeldt-Jakob disease, with characteristic findings in MRI and 18F-FDG-PET/CT; the latter, although it has not yet been included in the diagnostic criteria, offers a great contribution in the differential diagnosis.

Clinical Case

75-year-old man, with pathological antecedents of arterial hypertension, prostate cancer and bipolar affective disorder under treatment. Family history of Alzheimer's disease in the mother.

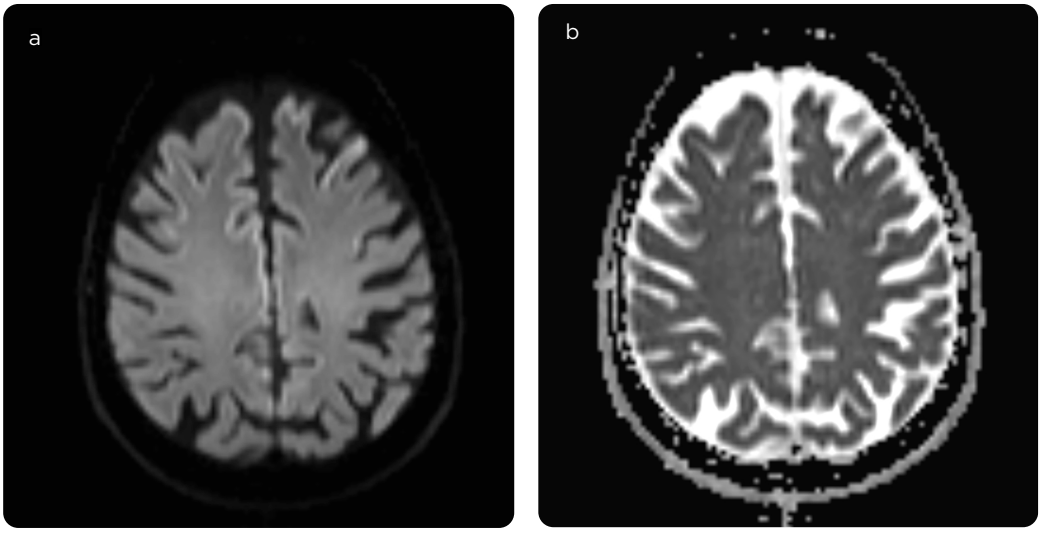

Figure 1. Brain MRI in diffusion sequences (b1000 coefficient) and ADC, axial. a) Areas of diffusion restriction visualized as high signal in the bilateral frontal cortex of left predominance, and in the cortex of the cingulate gyrus and b) corresponding to areas of low signal in the ADC map.

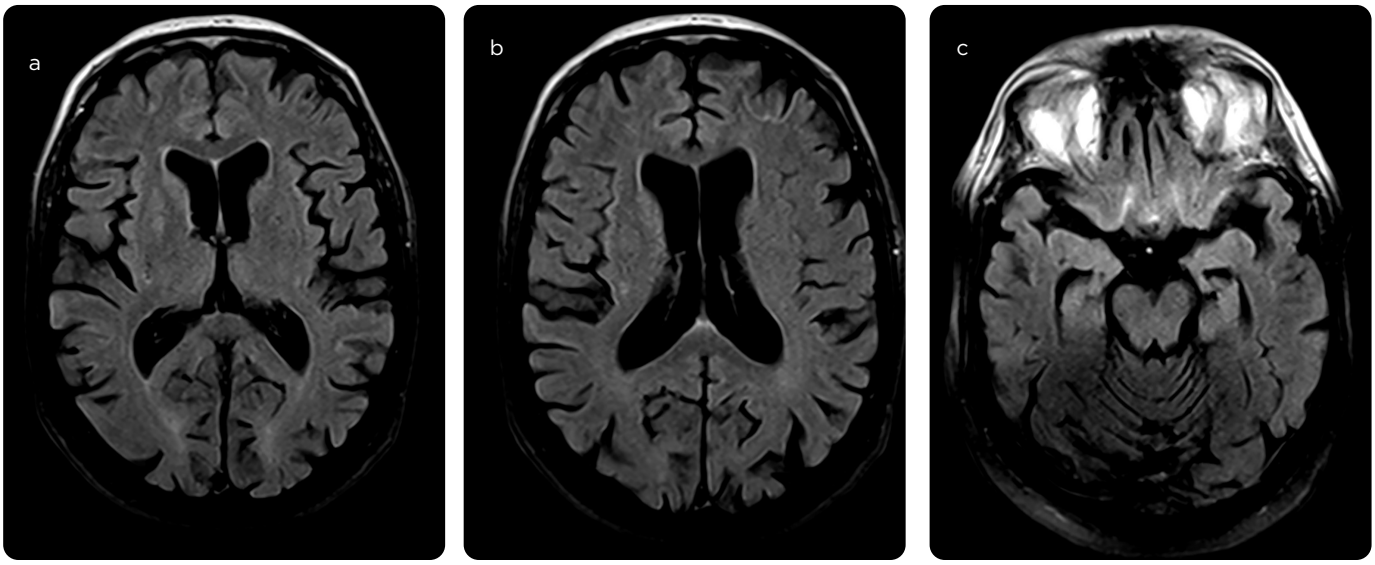

Figure 2. Brain MRI FLAIR sequence, axial. a, b) Areas of discrete high signal in the bilateral frontal cortex, in the cingulate gyrus cortex and c) in the bilateral temporal lobe cortex.

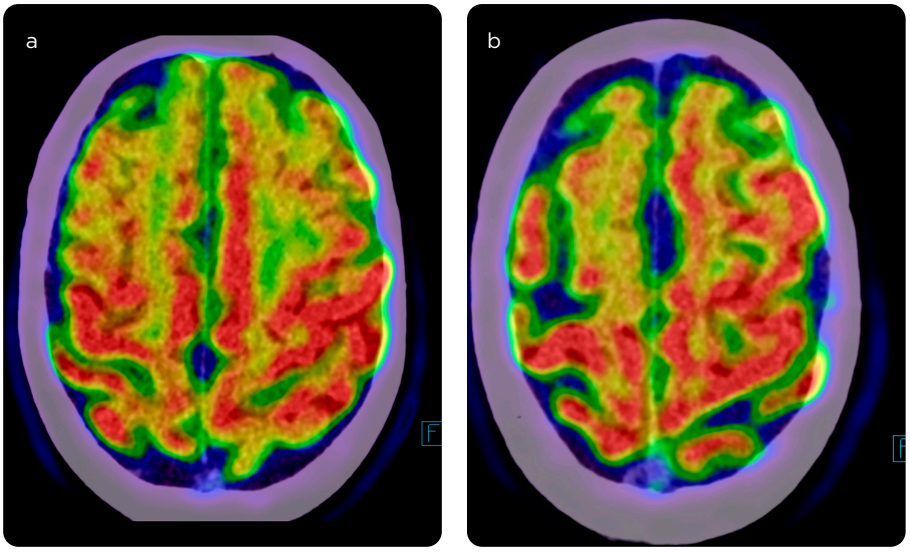

Figure 3. a, b) Axial slices of 18F-FDG-PET/CT image showing areas of hypometabolism in bilateral frontal cortex, right predominant.

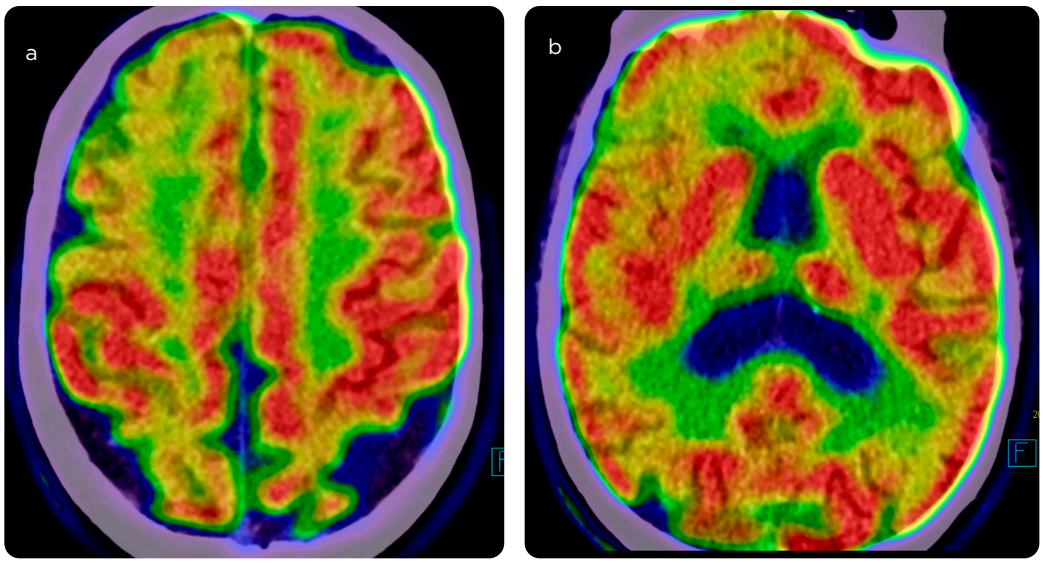

Figure 4. a, b) Axial slices of 18F-FDG-PET/CT image showing areas of hypometabolism in the right frontal and parietal cortex.

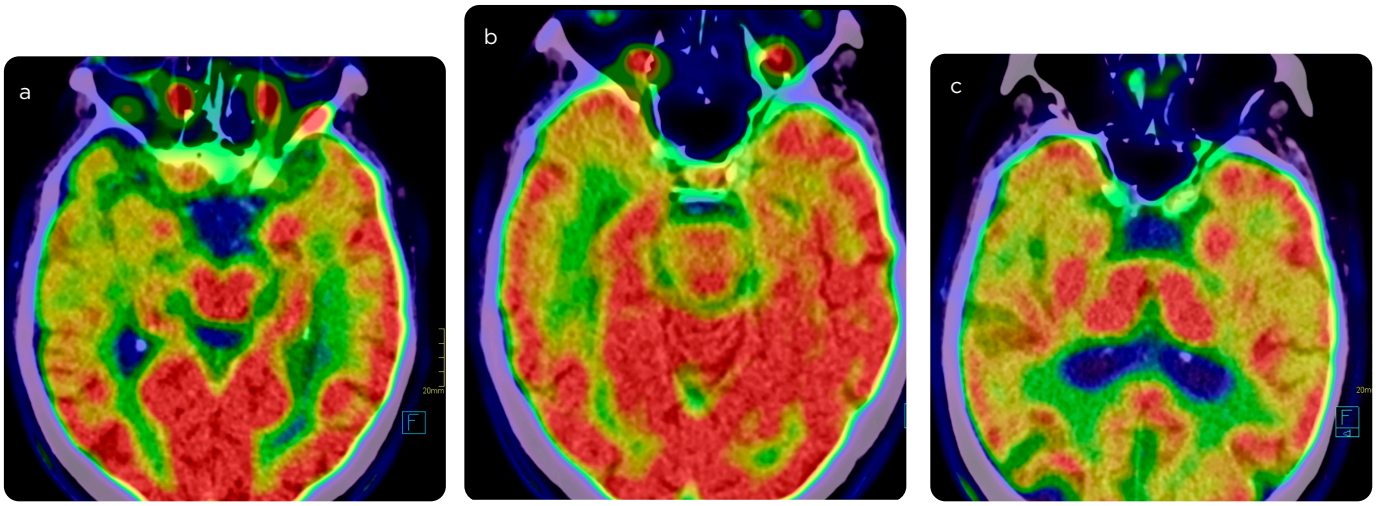

Figure 5. a, b, c) Axial slices of 18F-FDG-PET/CT image showing areas of hypometabolism in the bilateral temporal cortex, right predominant.

The patient consulted for rapidly progressive symptoms of two weeks of evolution consisting of behavioral changes, hypoprosexia, depressed mood, hyporexia, visual and auditory hallucinations, delusions and spatial disorientation, which coincided with the voluntary discontinuation of psychiatric medication, so decompensation of his underlying disease was suspected. The electrocardiogram, chest x-ray and initial laboratories showed no alterations.

During hospitalization she presented rapid progression of symptoms, hypoactivity, spatial and temporal disorientation, bradypsychia, fluctuation of alertness, bradykinesia, slow gait, distal intermittent tremor in lower limbs and right Babinski. Computed axial tomography (CT) of the skull showed involutional changes, CT of the neck, thorax and abdomen ruled out underlying solid neoplasm, and electroencephalogram (EEG) showed mild nonspecific encephalopathy.

Magnetic resonance imaging (MRI) of the brain with contrast medium was performed on suspicion of limbic encephalitis of possible paraneoplastic origin, which showed areas of diffusion restriction in the cortex of both frontal lobes of left predominance and in the cingulate gyrus (Figure 1).

These areas and the cortex of both temporal lobes showed high signal in FLAIR (Figure 2). No contrast enhancement or depiction was demonstrated in the other pulse sequences. Autoimmune encephalitis of paraneoplastic origin and prion disease were considered as diagnostic possibilities.

In order to rule out paraneoplastic involvement, a body 18F-FDG-PET/CT was performed, negative for moderate/poorly differentiated tumor viability. The 18F-FDG-PET/CT brain showed hypometabolism of the bilateral frontal cortex, predominantly right and in the ipsilateral parietal and temporal lobes (Figures 3-5), with adequate cerebellar and basal ganglia metabolism, without alterations suggestive of tumor viability. The findings found have been described in prion disease, so this was considered the most probable diagnosis (1, 2).

Subsequently, a cerebrospinal fluid study showed 14-3-3 protein, T-Tau protein and RT-QUIC positive, resulting in probable CJD.

Discussion

CJD is a neurodegenerative pathology caused by prions, with a low incidence and no curative treatment so far, with a mortality rate of 100 % (3).

It has been classified into: acquired forms -iatrogenic and variant-, genetic and sporadic (sCJD) -the most frequent of the three, representing 85-95 % of cases- (3).

Probable sCJD is diagnosed by the sum of the patient's clinical symptoms and positive findings in RT-QUIC, 14-3-3 protein and electroencephalogram or MRI (2,5). The definitive diagnosis is obtained with the pathological study of brain tissue (1).

The findings frequently described in MRI are: diffusion restriction or high signal in FLAIR, in the cortex -mainly in the insula, cingulum and frontal lobes- or in basal ganglia -mainly, caudate and putamen-, frequently bilateral and asymmetric. These findings have been documented even before the onset of symptoms (1, 2, 6).

As for 18F-FDG-PET/CT, characteristic findings have been reported that can be found even before MRI abnormalities are observed (2) and consist of hypometabolic areas in the affected areas, usually bilateral and asymmetric. The medial temporal region, cerebellum and basal ganglia are often less involved (1, 7).

The patient in this case met the clinical and MR imaging criteria for sCJD, but because of the suspicion of autoimmune encephalitis of paraneoplastic origin, 18F-FDG-PET/CT was performed, which supported the diagnosis of sCJD.

Although 18F-FDG-PET/CT is not part of the diagnostic criteria for CJD, it is a very useful tool, especially in patients in whom MRI cannot be performed or is inconclusive. It is also useful to rule out some differential diagnoses. However, more studies are needed for it to become part of the comprehensive study of rapidly progressive dementias, and to be included in the diagnostic criteria.

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