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CAS CLINIQUE

MRI and FDG PET/CT findings in a case of probable Heidenhain variant Creutzfeldt-Jakob disease

Aspects en IRM et TEP/TDM au FDG d'un cas de variant Heidenhain de maladie de Creutzfeldt-Jakob probable

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MOTS CLÉS
Maladie de Creutzfeldt-Jakob ;
Variant Heidenhain ;

Summary Creutzfeldt-Jakob disease (CJD) is a neurodegenerative disease caused by the accumulation of a pathogenic isoform of a prion protein in neurons that is responsible for subacute dementia. The Heidenhain variant is an atypical form of CJD in which visual signs are predominant. This is a report of the case of a 65-year-old man with probable CJD of the Heidenhain variant, with topographical concordance between findings on magnetic resonance imaging (MRI) and 18F-fluorodeoxyglucose (FDG) photopenic areas on positron emission tomography (PET)/computed tomography (CT) for cortical parietooccipital lesions.

Résumé La maladie de Creutzfeldt-Jakob (MCJ) est une maladie neurodégénérative causée par l'accumulation d'un isoforme pathogène de la protéine du prion dans les neurones, responsable d'un tableau de démence subaiguë. Le variant Heidenhain est une forme atypique de la MCJ pour laquelle les signes visuels sont au premier plan. Les auteurs rapportent le cas d'un variant Heidenhain de MCJ probable chez un patient de 65 ans, avec présentation des

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Imagerie par résonance magnétique ;
Tomographie par émission de positons (TEP)/TDM au FDG-(18F)

concordances entre les anomalies de signal en imagerie par résonance magnétique (IRM) et l'hypofixation du fluorodésoxyglucose-(18F) (FDG) en tomographie par émission de positons (TEP)/TDM pour les lésions corticales pariéto-occipitales.

Introduction

Sporadic Creutzfeldt-Jakob disease (sCJD) is an infrequent cause of rapidly progressive dementia. Its major symptoms are cognitive dysfunction, myoclonus and pyramidal syndrome. The Heidenhain variant is a rare presentation of sCJD in which visual disturbances such as blurred vision, hallucinations or cortical blindness are at the forefront [1,2].

We report on the case of a probable Heidenhain variant sCJD on the basis of findings from electroencephalography (EEG), fluid-attenuated inversion recovery (FLAIR) imaging and diffusion-weighted images (DWI) on MRI, as well as, on 18F-fluorodeoxyglucose positron emission tomography/computed tomography (18F-FDG PET/CT) of the cortical parietooccipital lesions.

Case report

A 65-year-old male patient was referred to the neurology department because of rapidly worsening dementia. He had no previous history of neurological disorder. The patient presented with recent subacute dementia with anterograde amnesic disturbances and hallucinations, but without fever. Neuropsychological examination revealed disorientation, psychomotor slowdown and anosognosia.

The physical examination showed no somatic abnormality and the initial EEG, performed at the beginning of the symptoms, was normal. One week later, a second EEG (not shown) demonstrated marked slowing of basic activity and a generalized synchronous slow discharge of periodic sharp wave complexes (PSWC). A state of hypovoltage was also observed in the right occipital area.

The patient underwent the following work-up: laboratory assays; contrast-enhanced CT; MRI (1.5-T) and FDG PET/CT. Also, serum tests for *Mycobacterium tuberculosis, Borrelia burgdorferi*, herpesvirus, HIV and syphilis (*Treponema pallidum* hemagglutination [TPHA] and VDRL) were all negative. Analysis of cerebrospinal fluid (CSF) showed one per millimeter cube white cell, normal glucose and protein levels but no 14-3-3 protein.

The contrast-enhanced CT scan (not shown) was normal. MRI axial FLAIR-weighted images showed bilateral hyperintensities in the cortical ribbon of the parietooccipital area, with right dominance. No abnormality was seen in the basal ganglia (Fig. 1A). DWI showed hyperintensities in the parietooccipital cortices (Fig. 1B). The apparent diffusion coefficient (ADC) was decreased in these cortical lesions. Moreover, areas with DWI hyperintensities were in accord with those seen on FLAIR-weighted images (Fig. 1A and B). FDG PET/CT was performed because, as paraneoplastic syndrome was initially evoked, a primitive tumor was

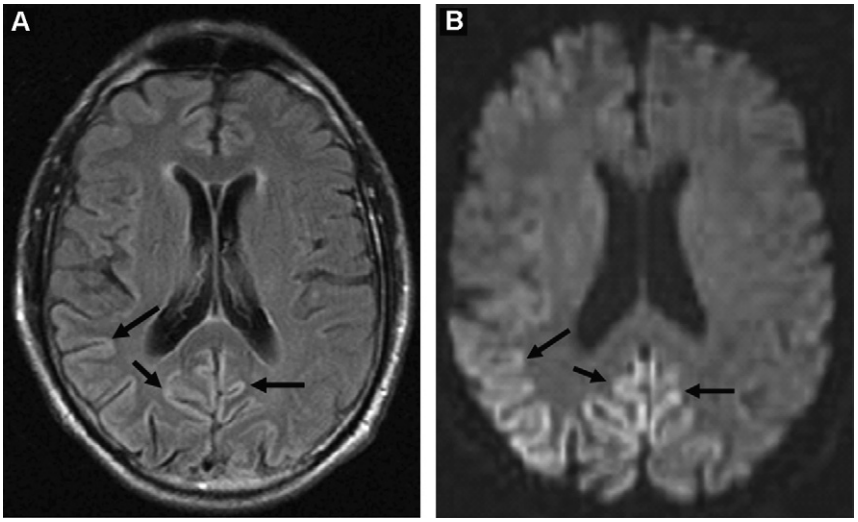

Figure 1 Brain (1.5-T) MRI of a 65-year-old man with rapidly progressive dementia. Axial FLAIR-weighted images **(A)** show slight hyperintense signals that are bilateral and asymmetrical in the parietooccipital cortical ribbon (black arrows). On DWI **(B)**, hyperintense signals are observed at the same locations (black arrows), with a reduction of the apparent diffusion coefficient on ADC mapping (not shown).

Figure 1 Patient de 65 ans présentant une démence rapidement progressive. IRM cérébrale (1,5T). Coupe axiale FLAIR **(A)** montrant de discrets hypersignaux bilatéraux et asymétriques dans le ruban cortical pariéto-occipital (flèches noires). Sur les images en diffusion **(B)**, des hypersignaux sont visualisés dans les mêmes régions qu'en FLAIR, avec réduction du CAD (coefficient apparent de diffusion) sur la cartographie CAD (non présentée).