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A Defying Case of Progressive Multifocal Leukoencephalopathy

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Abstract: ***Progressive Multifocal Leukoencephalopathy is a diffuse demyelinating disease of the central nervous system, caused by infection by the JC virus, usually associated with immunosuppression. The authors present the case of a 78 years old male, that was brought to the emergency department due to a confusional state. On the emergency department a Brain Computed Tomography was performed, revealing"Multiple cortical-subcortical and deep focal hypodensity areas in both hemysferes, associated with dimming of the grooves, of unknown nature, likely secondary." The patient was admitted and a Brain Magnetic Resonance Image was performed, and confirmed the previous results, even though it didn't exclude the possibility of a demyelinating disease. Considering the likelihood of a Lymphoma, corthicoids where initiated, but failed to improve the Brain lesions, as confirmed by a second Brain Magnetic Resonance. The clinical status of the patient began to deteriorate, and the patient started to become lethargic and with dysphonia. A lumbar puncture was performed and found no neoplasic cells. Facing the images and fast evolution of the disease, the diagnosis of Progressive Multifocal Leukoencephalopathy was assumed.***

Keywords: Progressive Multifocal Leukoencephalopathy; Brain lesions; Confusional State

1. Learning Points

The first clinical impression obtained from complementary diagnostic exams is not always the correct one

Even though Progressive Multifocal Leukoencephalopathy (PML) is usually associated with immunosuppression, in this patient that was not the case

It is not always possible to pursue a complete diagnostic approach, since the patient and family may not approve all interventions. Communication is essential, but not always enough.

2. Case Description

The authors present the case of a 78 year old male, previously independent, with a personal history of arterial hypertension, dyslipidemia and prostatic surgery of unknown cause 20 years earlier, medicated with a statin, a beta blocker and an Angiotensin Conversion enzyme inhibitor, that was brought to the emergency department due to a period of disorientation and confused speech during the night, while attempting to go to the bathroom. During that episode, the patient fell, but had no major head trauma, and denied loss of consciousness.

At evaluation on the emergency department, the patient was alert, oriented in time and space, without speech alterations, without meningeal signs, without visual alterations, without loss of muscular strength, but with apparent minor pronator drift on the right arm.

The remainder physical examination showed no alterations, and the patient had normal blood pressure and cardiac frequency, and a temperature of 36.7ºC.

The blood samples revealed no significant anomalies, and the Brain Computed Tomography (CT) revealed "Multiple cortical-subcortical and deep focal hypodensity areas in both hemispheres, associated with dimming of the grooves, of unknown nature, likely secondary." (Image 1)

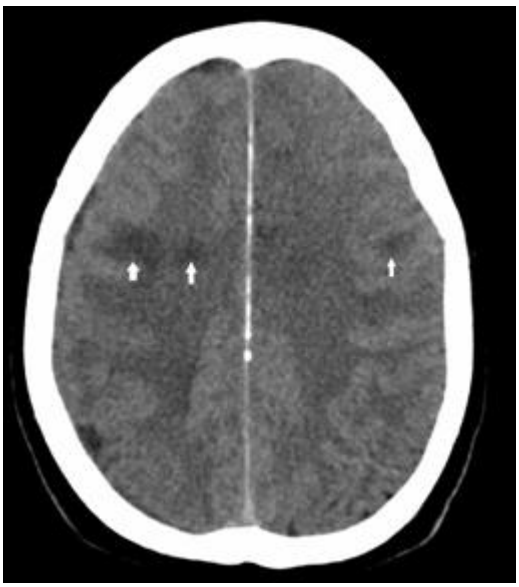

Image 1: CT Scan

The patient was admitted to study. Initially the patient remained asymptomatic, without further changes to the physical examination. A Brain Magnetic Resonance Image (MRI) was performed and confirmed the "Presence of

multiple occupying space lesions on the cortico-subcortical interface of both hemispheres, predominantly on the frontal regions but also on the basal ganglia and corpus calosum. Thelesions exhibit no diffusion restriction nor anomalous highlight after contrast. Their multiplicity may suggest a secondary lesion, even though the possibility of inflammatory disease (demyelinating) cannot be excluded. The lesions cause mass effect but no ventricular compression nor midline shift". (Image 2) Having this image, the clinical state f the patient was surprising, exhibiting no focal signs at all.

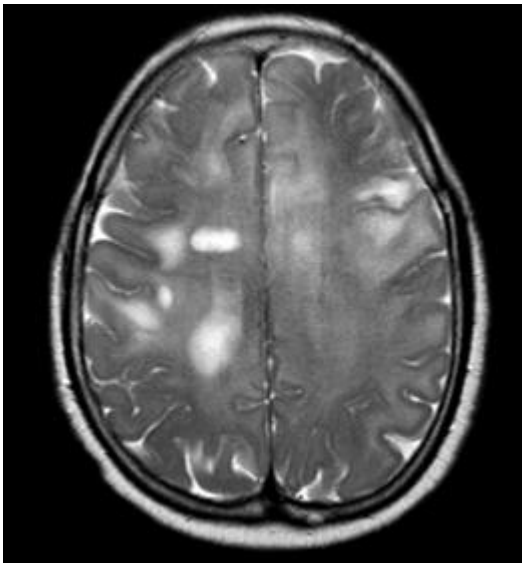

Image 2: First MRI

Having in mind the possibility of a secondary lesion, a Thoraco-Abomino-Pelvic CT was performed, and showed no primary lesions. A Positron Emission Tomography was also performed and revealed only "some heterogeneity on the distribution of FDG-F18 on the brain, without clearly suspicious focal changes".

A lumbar puncture was performed, with normal protein level and glucose, a weak positive antineuronal anti-SOX antibody, normal immunophenoty page, and no neplasic cells.

The possibility of a lymphoma was considered, and the patient initiated corthicosteroids. At this point, the patient behavior started to shift, and the patient began to act differently, running on the yard and performing exercises.
After 1 week of corticotherapy a second MRI was performed, to access drug response, but showed no alterations at all. (Image 3).

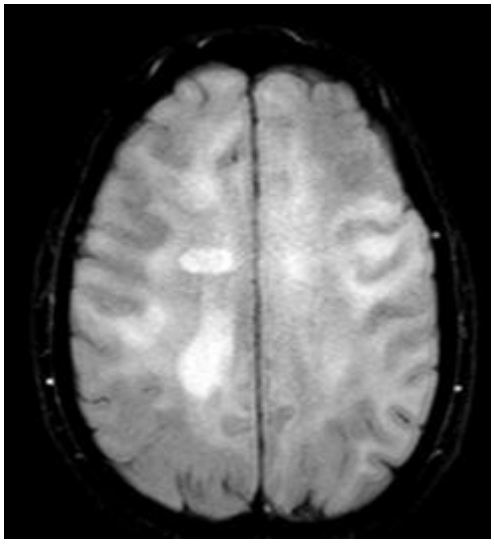

Image 3: Second MRI

The possibility of a PML was then considered, and the case discussed multidisciplinary with a neurologist, neuroradiologist and infecciologist, which corroborated the possibility. The cerebrospinal fluid was tested for JC virus, but was negative.

The clinical state of the patient then began to deteriorate, with generalized weakness and dysphonia, and the patient became bedridden. An electroencephalogram was performed, and revealed "diffuser brain disfunction, without specific etiological findings".

A brain biopsy was then considered, but the patient and family denied, becoming thepossible diagnose of PML assumed based on the clinical evolution and imageological appearance of the lesions.

The patient remained on rehabilitation, without significant improvements.

3. Discussion

Even though PML is associated with JC virus infection, false-negative PCR-based results for JC virus in cerebrospinal fluid have been reported. [1, 2]

Most cases are associated with immunosuppression, mainly with HIV infection, but there are reported cases of PML and JC infection in immunocompetent patients.[1,2] The patient in this case had a prostate surgery, performed in a foreign country where he used to work, and had no information on the cause. The possibility of a previous neoplasm has to be considered.

The above case, does not allow a definite diagnose of PML, due to the lack of JC infection confirmation, but with the suggestive clinical findings and imageologicalfindings, a possible diagnose can be achieved.[3]

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