Case 17814

Eurorad

LGI1-abs Encephalitis

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Section: Neuroradiology
Area of Interest: Neuroradiology brain
Imaging Technique: MR
Case Type: Clinical Cases
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Patient: 71 years, male

Clinical History:

A 71-year-old male presented with episodes of confusion and disorientation lasting a few minutes. Clinical examination revealed choreiform movements. The patient had no recent illness, fever, headaches, nausea or vomiting episodes. Blood results and CT brain were normal, therefore, the underlying reason for the confused and disoriented presentation remained unexplained.

Imaging Findings:

Two MRI Head scans were subsequently performed one month apart.

During the first scan, the patient was uncooperative allowing for only the T2 weighted sequences and diffusion-weighted imaging to be performed. Bilateral high signal intensity within the caudate nuclei, putamina as well as the globus pallidi were present, right more than left. The affected basal ganglia were slightly swollen. There was no evidence of restricted diffusion in the affected regions (Figures 1 and 2).

On the second MRI scan performed one month later, bilateral symmetrical swelling and high FLAIR/T2 signal of the hippocampi was present. There was restricted diffusion in both hippocampi. The basal ganglia appeared normal on T2 weighted images with some high signal on T1 weighted imaging in the right basal ganglia and anterior commissure (Figures 3–5).

Discussion:

Background

Encephalitis often results secondary to an infection or an autoimmune response. LGI1-abs (leucine-rich glioma inactivated 1 - antibodies) is an uncommon autoimmune encephalitis. This pathology is more commonly found in men than women (2:1) and is very rare in children.

Clinical perspective

Signs and symptoms include mental disorders and hyponatremia. severe anterograde amnesia, psychiatric symptoms, and seizures [1–2].

Seizures are one of the first signs in patients with LGI1-abs encephalitis. In these cases, faciobrachial dystonic seizures (FBDS) are the most common seizure type and are seen in around 60% of cases [3]. Initially, FBDS may be a rare episode of occasional involuntary muscle jerk movements of their arm and their face. However, this may increase to occur more frequently.

In addition, some patients would have lowered blood sodium levels (hyponatremia) which might be indicative of this illness [4].

Imaging perspective

MRI scans may show changes in the affected parts of the brain, including the temporal lobe and the hippocampus [5]. 70% of patients would have increased T2 and FLAIR MRI signals within these affected areas [5]. In addition to supporting the diagnosis, MRI has the ability to quantify disease severity [6].

Outcome

Patients suffering from LGI1-abs would require early and long-term effective immunotherapy of anti-LGI1 encephalitis for better cognitive functional prognosis [7–8]. Patients are given first-line therapies (e.g. corticosteroids, intravenous immunoglobulin, or plasma exchange). The first-line therapies have a rapid onset of action and relatively low associated risks. It is sometimes necessary to add second-line immunotherapy (e,g, cyclophosphamide or rituximab) [9]. These are administered if the patient does not respond to the first-line immunotherapy agents. The second-line immunotherapy are higher-risk medications due to their longer duration of action and increased immunosuppressive nature [9].

Approximately 25% of patients who suffered from LGI1-abs encephalitis would relapse [10]. This is most commonly seen upon treatment withdrawal. In addition, the mortality rate of LGI1-abs encephalitis is between 6–19% [10–11].

Take-home message/Teaching Points

The diagnosis of LGI1-abs encephalitis may be challenging, especially since patients might present to psychiatrists or movement disorder specialists prior to autoimmune neurologists. However, it is important to note that psychiatric symptoms and seizures are the most common signs of this pathology. LGI1-abs can be confirmed in spinal fluid and blood results, and then early treatment is necessary to improve the prognosis.

Differential Diagnosis List: LGI1-abs encephalitis, Viral encephalitis, Toxoplasmosis

Final Diagnosis: LGI1-abs encephalitis

References:

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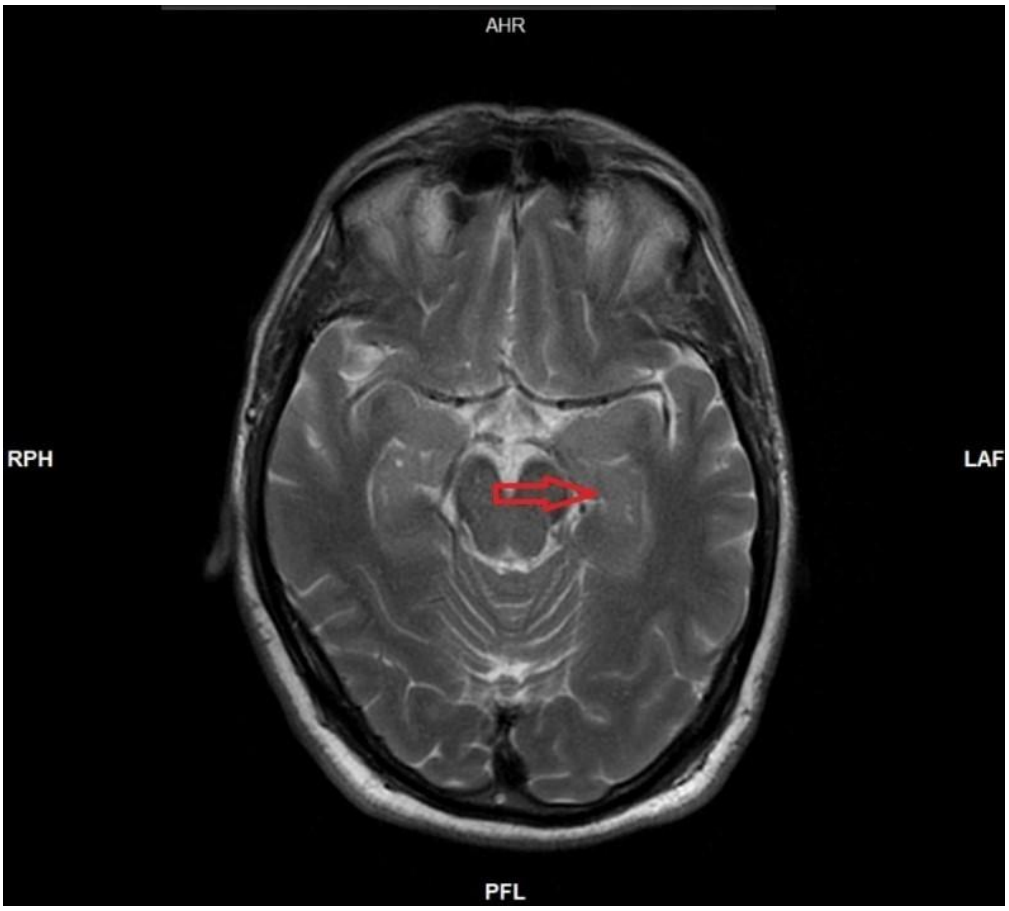
Figure 1

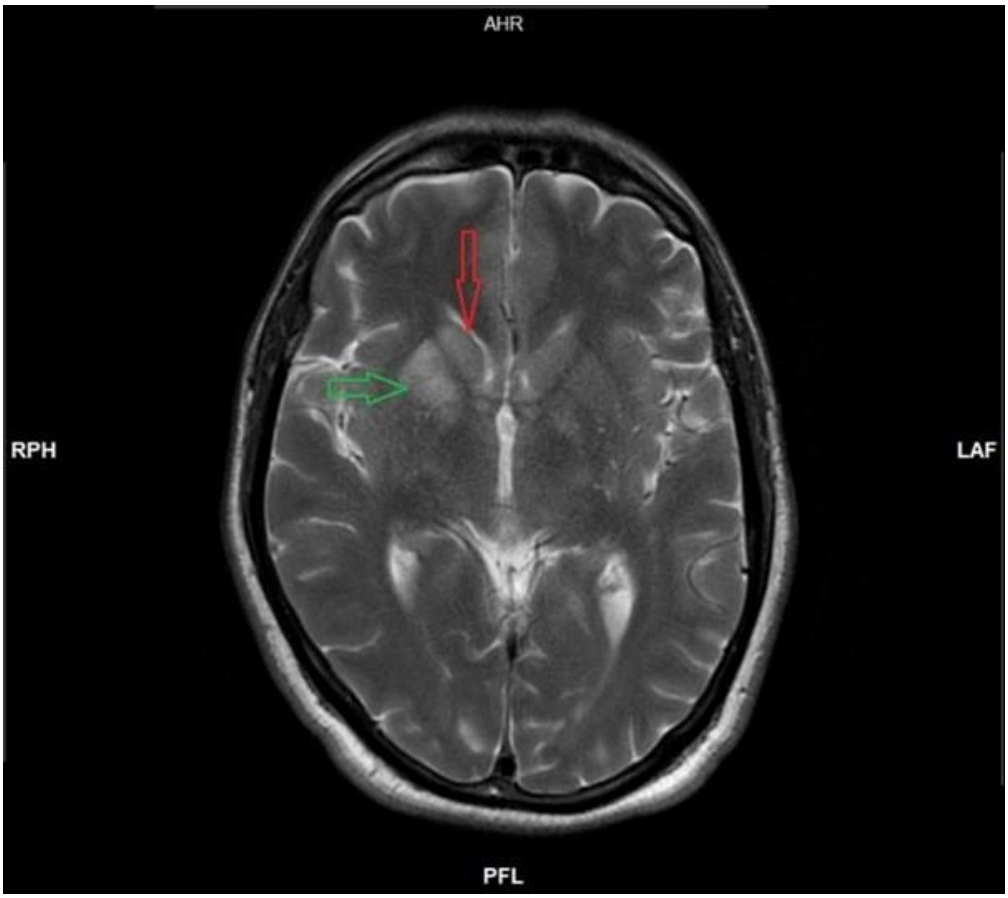
a



Description: On the initial images, at the level of the hippocampi - there is no restricted diffusion on the left **Origin:** Medical Imaging Department, Mater Dei Hospital, Msida, Malta

Figure 2

a



Description: There is bilateral asymmetrical T2 hyperintensity in the basal gangli (right > left). Both the putamen (green arrow) and caudate nuclei (red arrow) are affected. There is no corresponding restricted diffusivity in the affected areas **Origin:** Medical Imaging Department, Mater Dei Hospital, Msida, Malta

Figure 3

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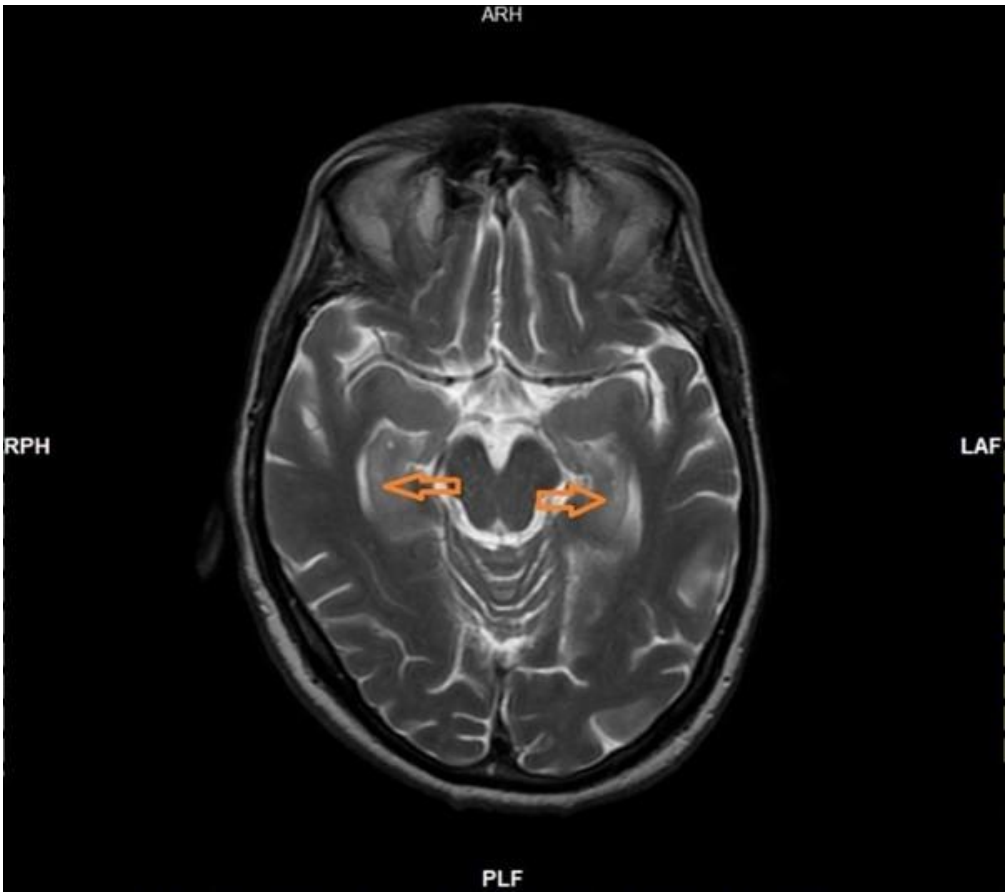

Description: Follow-up MRI (Axial T2W and Axial DWI) show interval development of bilateral hyperintensity within the body in the hippocampi bilaterally (orange arrows)**Origin:** Medical Imaging Department, Mater Dei Hospital, Msida, Malta

Figure 4

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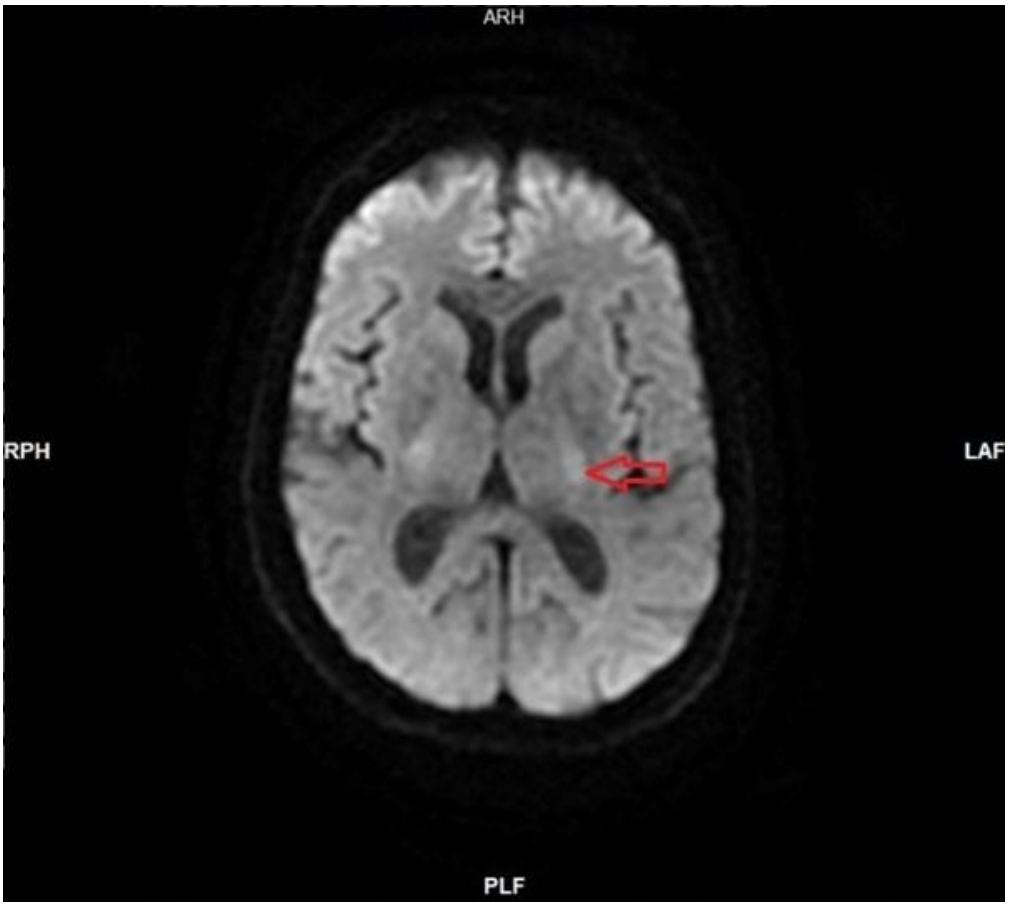
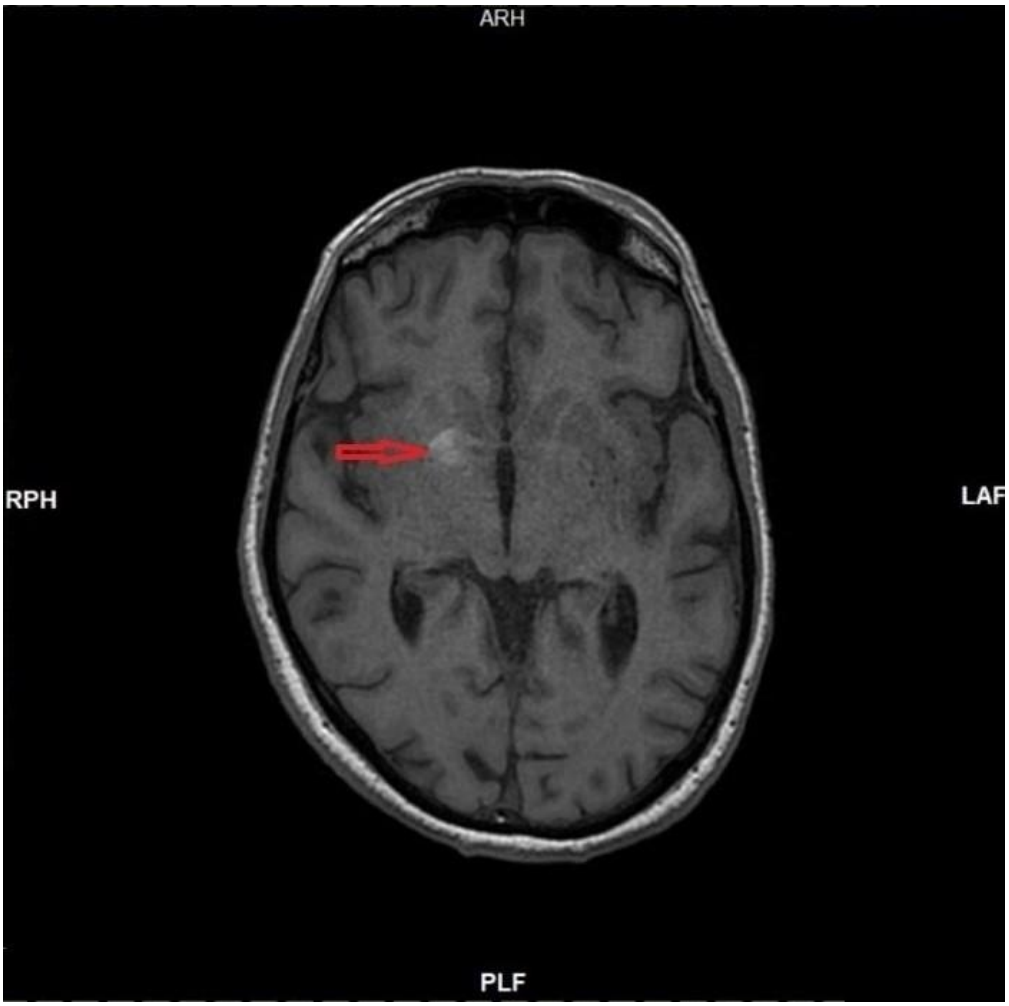


Description: Follow-up image showing evidence of restricted diffusion in the internal capsules bilaterally (red arrow) seen bilaterally **Origin:** Medical Imaging Department, Mater Dei Hospital, Msida, Malta

Figure 5

a



Description: Follow-up image shows intrinsic (pre-contrast) T1 hyperintensity (red arrow) in the right basal ganglia region. There was no corresponding haemosiderin deposition on SWI **Origin:**
Medical Imaging Department, Mater Dei Hospital, Msida, Malta