Autoimmune Encephalitis: A Pictorial Essay

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INTRODUCTION

Autoimmune encephalitis is a family of disease entities that involve antibody-mediated inflammation of the brain. They demonstrate overlapping but variable clinical and imaging manifestations. Although definitive diagnosis relies on serum or CSF antibody testing, it is important for the radiologist to recognize and suggest the possibility in the absence of routine antibody screening to enable timely serological diagnosis and treatment.

ANTI-NMDAR ENCEPHALITIS

Anti-N-methyl D-aspartate receptor (NMDAR) encephalitis presents with cognitive and behavioral symptoms, seizures, and abnormal movements. There is an association with tumors, most frequently ovarian teratomas.

Initial MR brain is normal in 49-89% of patients. T2-FLAIR hyperintense lesions are seen with an apparent predilection for the hippocampus (Fig 1). Non-hippocampal lesions are most commonly found in the frontal and temporal lobes. Mesial temporal atrophy is seen in some patients. Leptomeningeal, cortical, and parenchymal enhancement are sometimes present (Fig 2).

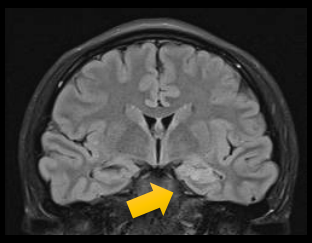

Fig 1. Hippocampal hyperintensity in a 26 year old female patient with ovarian teratoma.

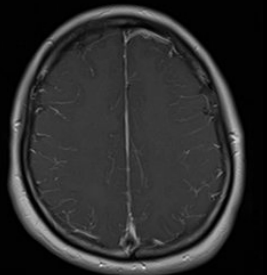

Fig 2. Diffuse leptomeningeal enhancement in a 36 year old female patient.

MOG ENCEPHALITIS

Myelin oligodendrocyte glycoprotein (MOG) antibody disease predominantly causes inflammation and demyelination of the optic nerve and spine. When the brain is involved, its clinical picture is similar to acute disseminated encephalomyelitis. About half of patients on presentation have abnormal MR brain. Most patients have lesions in the supratentorial white matter (Fig 4A&B), where they are commonly found in the juxtacortical and periventricular/juxtaventricular white matter. Mesial temporal lesions have also been reported. Around one third of patients demonstrate lesions in the brainstem, corpus callosum, thalamus, and/or basal ganglia. Cranial nerve involvement is common in MOG encephalitis, with concomitant optic neuritis found in at least half of patients (Fig 4C).

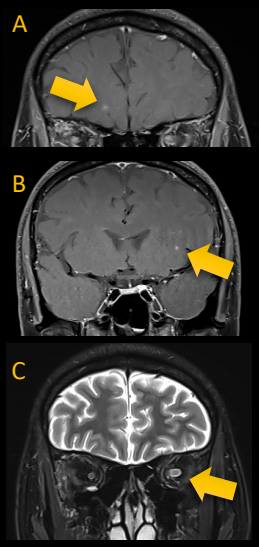

Fig 4. Enhancing supratentorial lesions (A, B) and left optic neuritis (C) in a 28 year old male patient.

VOLTAGE-GATED POTASSIUM CHANNEL (VGKC) ENCEPHALITIS

VGKC encephalitis is generally non-paraneoplastic. Cognitive impairment and seizures are the most frequent presentations.

Limbic abnormalities dominate on imaging, presenting as amygdala and/or hippocampal enlargement with T2 hyperintensity (Fig 3). It can also mimic Creutzfeldt-Jakob Disease with cortical T2/FLAIR hyperintensities.

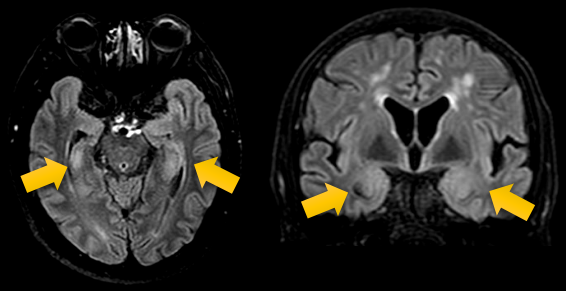

Fig 3. Hippocampal hyperintensity and swelling in a 53 year old male patient.

VOLTAGE-GATED CALCIUM CHANNEL (VGCC) ENCEPHALITIS

VGCC encephalitis is a rare subtype with variable clinical manifestations, ranging from seizure, confusion, psychiatric symptoms, vertigo and tremors, to rapidly progressive dementia.

MR findings can be normal. Abnormal T2-FLAIR signals can be found in the hippocampus, and less frequently in the parieto-occipital lobes and cerebellum. Juxtacortical and periventricular T2-FLAIR hyperintense lesions mimicking multiple sclerosis, as well as migrating gyriform enhancement and cortical laminar necrosis have also been reported.

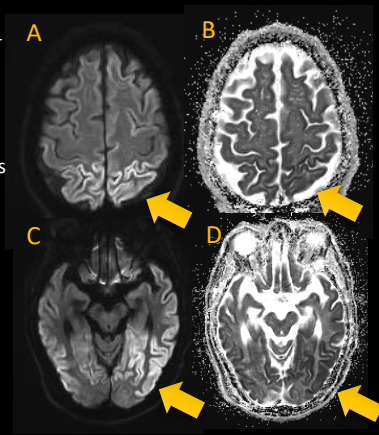

Fig 5. Atypical presentation with cortical restricted diffusion on DWI (A,C) and ADC maps (B,D) mimicking CJD in a 79 year old male patient.