Remember the Limbic System?: MR Anatomy and Pathology Review of Structures Involved in Emotion and Memory Formation

Jane Ball,BS; David Sawyer,BS; Adam Blanchard,MD; Krystle Barhaghi,MD; Enrique Palacios,MD; Jeremy Nguyen,MD.
Tulane University School of Medicine Department of Radiology

Introduction

Rather than a single, defined structure within the brain, the limbic system is a collection of interrelated structures involved in learning, memory, emotional responses, homeostasis and primitive drives. Different reference sources include and exclude structures within the limbic system. Some structures share formations or groupings and have additional functions beyond their roles in the limbic system. Generally, the hippocampal formation, amygdala, hypothalamus and limbic cortex form the limbic system. The limbic cortex includes the fornix, cinngulate gyrus, prefrontal cortex, septal area, and parahipocampal gyrus. Closely related structures include the basal ganglia, thalamus, and mamillary bodies. This poster reviews the functions, anatomy and neuroimaging appearances of limbic system components in disease states.

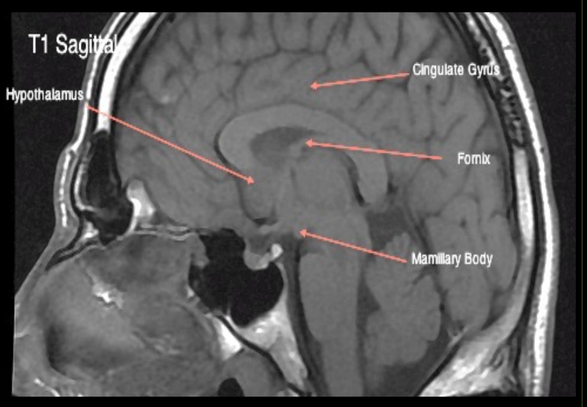
Functional Review

In this section, the limbic-related related functions of the structures are reviewed. Other functions such as their influence on the autonomic nervous system and hormonal secretions not explicitly related to the limbic system are beyond the scope of this poster.

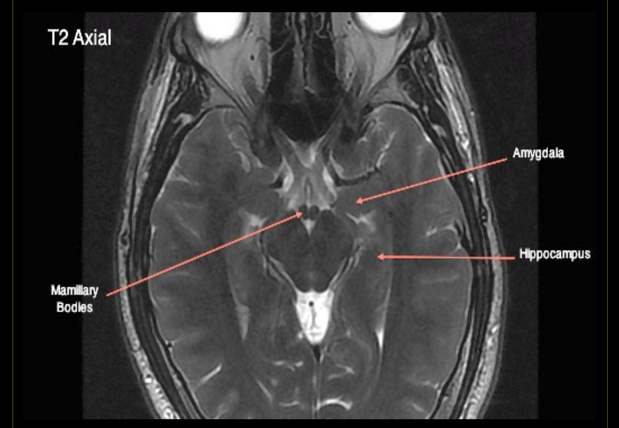
- Hippocampal formation: short-term and immediate explicit memory consolidation into long-term storage[1]
- Amygdala: emotional interpretation of external stimuli and internal states, including fear and aversion responses[1]
- Limbic Cortex
  - Fornix: encoding and recall of new information, as one part within the Papez circuit[2]
  - Cingulate gyrus: selection of appropriate responses to stimuli and projections to other structures within the limbic system [1]
  - Prefrontal cortex: integrates information from many areas of the brain including limbic structures and may influence complex behaviors based on that information[5]
  - Septal area: roles in behavior, attention, and memory
  - Parahippocampal gyrus: processes contextual associations roles in episodic memory and visuospatial processing[6]
  - Entorhinal cortex: sensory input modulation and integration with the hippocampus memory circuits[1]
- Hypothalamus: response coordination to internal and external stimuli, especially in regards to homeostasis and primal drives such as hunger; role in sleep and alertness[1]

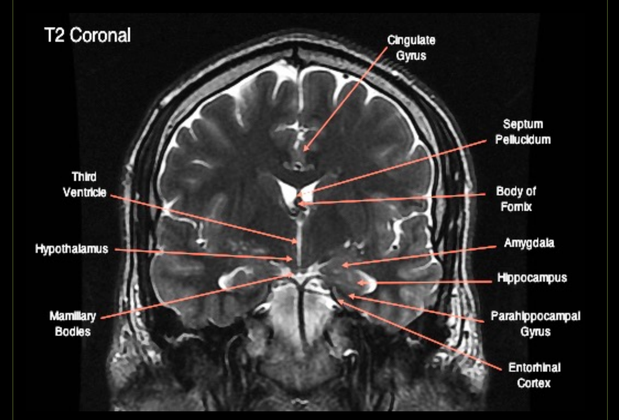
Structural Review

MR images that demonstrate the previously described structures:



Structural Review





MR Pathology

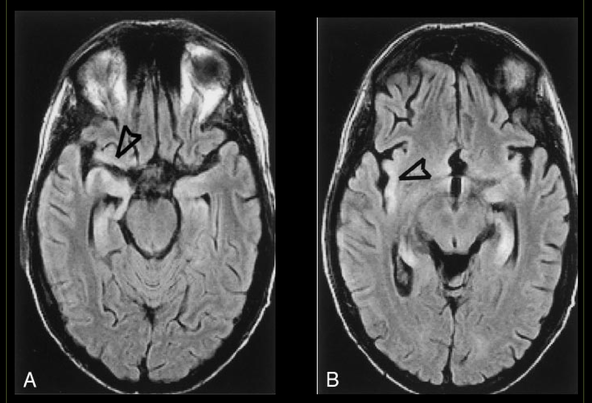
MR is the preferred method in evaluation of disease processes that affect soft tissue structures of the brain. MR allows for greater soft tissue contrast between structures than CT allows. Pathologies affecting the limbic system may mimic one another at first inspection. Some methods to clarify a diagnosis include optimized imaging such as different sequences, history and physical exam correlations, or laboratory testing such as serum antibodies, CSF antibodies, CSF cell analysis, and biopsy.

Limbic Encephalitis

Limbic Encephalitis (LE) presents with seizures, confusion, memory loss, anterograde amnesia, and/or irritability manifesting over days to weeks to months. LE is often considered a paraneoplastic syndrome, most commonly associated with SCLC, breast cancer, and germ cell tumors of the testis. However, there is some evidence that other autoimmune processes, including post-infectious states, may cause LE as well.[7,8,9] Serum antibodies can support the diagnosis. Anti-neuronal antibodies such as anti-Hu (associated with SCLC) and anti-Ta (associated with testicular cancer) can suggest a paraneoplastic syndrome etiology. VGKC (voltage-gated potassium channel) antibodies are associated with non-paraneoplastic etiologies.

Characteristic MR findings include increased signal intensity changes in bilateral temporal lobes on T2-weighted images, especially in FLAIR sequences, as well as in the hippocampi, amygdalae, and the hypothalamus.[9,10]

Limbic Encephalitis



FLAIR images of autopsy-proven paraneoplastic limbic encephalitis. Images from Thuerl et al.[10]

A. Axial FLAIR image with findings that include increased signal intensity in the medial temporal lobes bilaterally as well as in the right frontobasal cortex (arrowhead).

B. Axial FLAIR image with findings that include increased signal intensity in the medial temporal lobes bilaterally as well as in the right insular cortex (arrowhead).

Hippocampal Sclerosis

Hippocampal sclerosis, also known as mesial temporal sclerosis, is a condition characterized by neuronal cell loss and gliosis in the hippocampus, particularly in the cornu ammonis regions.[11] It is commonly found in asymptomatic persons at autopsy, but is clinically most associated with temporal lobe epilepsy. Patients with longstanding epilepsy can also demonstrate abnormalities of the structures connected to the hippocampus along the Papez circuit, such as the fornix and mammillary bodies.[12] This suggests that hippocampal sclerosis is in fact a disease involving the entire limbic system.

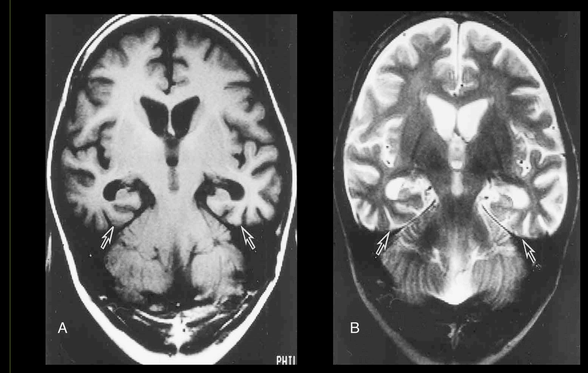
Hippocampal sclerosis is best demonstrated on MR imaging using a dedicated temporal lobe epilepsy protocol. [13] Characteristic findings include atrophy of the affected hippocampus and hyperintensity on T2-weighted images.[14] Gadolinium is not useful in evaluating hippocampal sclerosis.

A. Coronal T2-weighted MR image of the brain in a patient presenting with seizures.The left hippo-campus demonstrates both atrophy and a hyperintense T2 signal, characteristic of hippo-campal sclerosis. Image from A.Prof Frank Gaillard, Radiopaedia.org, rID: 2618.

B. Axial FLAIR image of the brain in a patient presenting with new onset seizures. There is hyperintense signal demonstrated in the region of the left hippo-campus, consistent with the final diagnosis of hippocampal sclerosis. Image from Dr Arthur Daire, Radiopaedia.org, rID: 31005.

Klüver-Bucy Syndrome

Klüver-Bucy Syndrome (KBS) is a clinical diagnosis characterized by visual agnosia, hyperorality, hypersexuality, placidity, abnormal dietary changes, hypermetamorphosis, dementia, and amnesia. Limbic encephalitis is the most common cause of KBS, and KBS has been associated with other neurological disorders including traumatic brain injury, anoxia-ischemic encephalopathy, Pick's disease, Alzheimer's disease, bilateral temporal lobectomy, tuberculous meningitis, and neurocysticerosis. MR findings in KBS include bilateral temporal lobe atrophy, bilateral hippocampi atrophy, atrophy of other limbic structures, cystic or necrotic foci in the limbic areas and edema within and surrounding the temporal lobes.[15-17]



Images of a 3 year-old female with Klver-Bucy syndrome from Ozawa et al.[15]

A. Coronal T1 image demonstrating diffuse brain atrophy (DBA), including in the
bilateral temporal lobes and hippocampi (arrows).

B. Coronal T2 image demonstrating DBA, especially in the bilateral hippocampi.

Conclusions

Limbic System pathology often presents with a clinical picture that correlates with the imaging findings on MR. Knowledge of the clinical presentation may aid in distinguishing similar imaging findings and vice versa.

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Tulane University
School of Medicine
Department of Radiology
1430 Tulane Ave. 8654
New Orleans, LA 70112-2699
Poster assistance by Jeremy Nguyen & Donald Olivares