Imaging of Bithalamic Pathology in the Pediatric Brain: Demystifying a Diagnostic Conundrum

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BACKGROUND AND PURPOSE

Several conditions are responsible for bilateral, symmetric or near-symmetric involvement of the thalami.

These are often distinct among adult and pediatric patients, although some etiologies are common to both groups.

Certain imaging features are shared among many etiologies; others are unique to a particular entity.

Presented here is an outline of the various causes of bilateral thalamic involvement and imaging findings to formulate a reasonable differential diagnosis for the pediatric and general radiologist.

MATERIALS AND METHODS

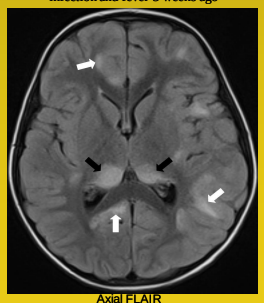
A retrospective review of patients with bilateral thalamic lesions was performed by selecting patients with known bilateral thalamic pathology.

Where available, imaging findings were correlated with clinical and laboratory findings.

Imaging findings were reviewed and key sequences were identified that facilitate the most likely diagnosis or differential diagnoses.

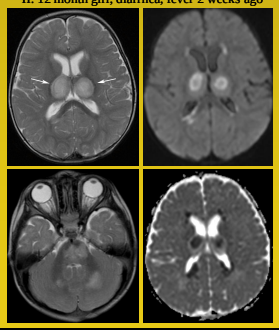
Imaging findings from the most common differentials were identified and are presented below.

I: 6-year-old girl, upper respiratory infection and fever 3 weeks ago



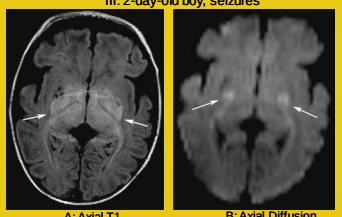
Axial FLAIR

II: 12 month girl, diarrhea, fever 2 weeks ago



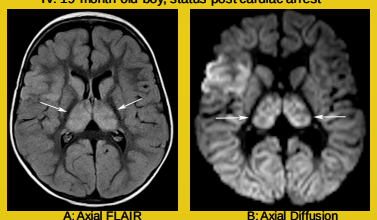
A: Axial T2 | C: Axial Diffusion | B: Axial T2 | D: Axial ADC

III: 2-day-old boy, seizures



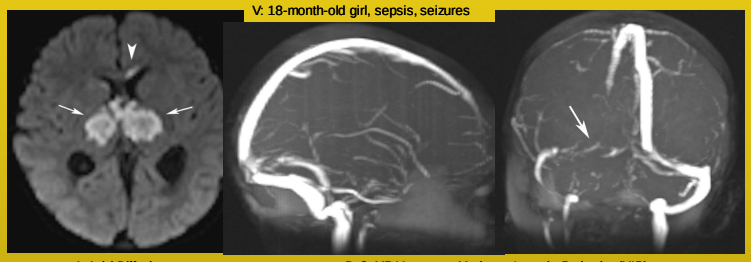
A: Axial T1 | B: Axial Diffusion

IV: 19-month-old boy, status-post cardiac arrest



A: Axial FLAIR | B: Axial Diffusion

V: 18-month-old girl, sepsis, seizures



A: Axial Diffusion | B, C: MR Venogram, Maximum Intensity Projection (MIP)

RESULTS

I. Acute Disseminated Encephalomyelitis (ADEM). FLAIR image reveals abnormal signal in bilateral thalami (black arrows) with scattered signal abnormalities involving subcortical white matter (white arrows). All abnormalities resolved on follow up imaging obtained four months later.

Usually a monophasic illness, immune-mediated sequela of viral (respiratory) infection. Patchy T2/FLAIR hyperintense foci involving gray and white matter, supra- greater than infratentorial. May affect thalami (12%). More acute lesions exhibit enhancement and/or restricted diffusion. Necrosis portends poor prognosis.

II. Acute Necrotizing Encephalopathy of Childhood (ANEC). A: There is bithalamic T2-weighted hyperintensity (A, arrows). The central thalami exhibit greater signal on both sequences. B: T2 image through the posterior fossa demonstrates hyperintense lesions in the pons and deep cerebellar white matter. C: Diffusion and D: ADC map demonstrate restricted diffusion in the thalami and periventricular white matter.

Variant of ADEM, more aggressive, reported mainly from Southeast Asia. Like ADEM, usually monophasic, immune-mediated demyelination following viral infection/vaccination. Bilateral symmetric thalamic lesions with edema, necrosis/ring enhancement, restricted diffusion, petechial hemorrhage. Other findings similar to ADEM.

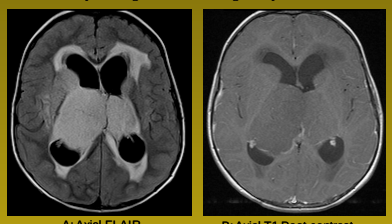
III. Early Hypoxic Ischemic Encephalopathy (HIE). A: T1 non-contrast image demonstrates hyperintensity (arrows) of the thalami and basal ganglia, with bilateral symmetric involvement. There is loss of normal T1 hyperintense myelin signal from the posterior limbs of internal capsules (PLICs) bilaterally. B: Posterior lentiform nuclei depict restricted diffusion (arrows).

IV. Full-blown HIE. A: FLAIR image demonstrates edematous, hyperintense thalami (arrows) and caudate heads, with right frontal predominantly cortical FLAIR signal abnormality. B: FLAIR hyperintense areas demonstrate restricted diffusion suggesting infarcts (arrows).

Result of severe cerebral hypotension. Basal ganglia, thalami, brainstem, cortical mantle with or without white matter typically affected. Early (birth) – T1 hyperintensity in deep gray structures, loss of normal neonatal T1-hyperintense/T2-hypointense PLIC myelin. Late (7-14 days) – T2/FLAIR hyperintensity within deep gray and white matter. ADC map useful, since DWI not always abnormal, and DWI sequence detects white matter abnormalities better than it detects deep gray abnormalities.

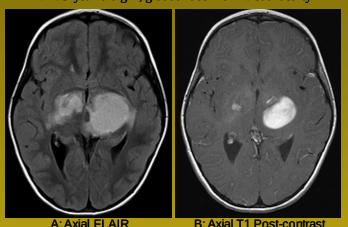
V. Venous Sinus Thrombosis. A: DWI reveals restricted diffusion within bilateral thalami (arrows) as well as genu of the corpus callosum (arrowhead). B and C: Lateral and frontal MIPs from a 2-D time-of-flight (TOF) MRV depicts loss of flow in the right transverse sinus (C, arrow). Sagittal image reveals diffuse thrombosis of the inferior sagittal, straight sinuses, and internal cerebral veins.

VI: 5-year-old girl, word stuttering, left eye deviation



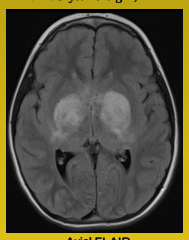
A: Axial FLAIR | B: Axial T1 Post-contrast

VII: 5-year-old girl, gradual decline in visual acuity



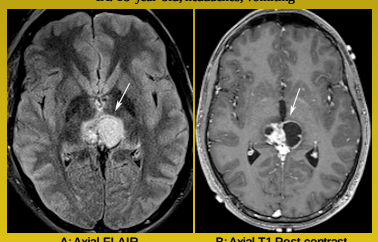
A: Axial FLAIR | B: Axial T1 Post-contrast

VIII: 5-year-old girl, NF1



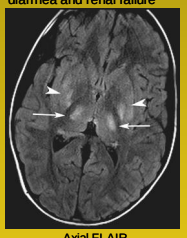
Axial FLAIR

IX: 18-year-old, headaches, vomiting



A: Axial FLAIR | B: Axial T1 Post-contrast

X: 10-year-old, acute hemorrhagic diarrhea and renal failure



Axial FLAIR

RESULTS

Etiologies: perinatal complications, dehydration, sepsis, coagulopathy. Thalami may be affected in conjunction with basal ganglia and/or PLIC. Usually vein of Galen or straight sinus thrombosed. Venous infarcts may or may not exhibit restricted diffusion. Hemorrhage noted in most venous infarcts. Useful sequences: MR venogram, gradient-recalled echo (GRE), post-contrast T1 with thin-sections.

VI. Tumor: bilateral fibrillary astrocytomas (WHO Grade II). A: FLAIR image shows right larger than left bithalamic masses effacing the third ventricle with obstructive hydrocephalus and transependymal CSF resorption. B: T1-weighted post-contrast image shows no intralesional enhancement.

Most often WHO Grade II, however III or IV grade tumors may occur. Typically involve bilateral thalami in a symmetric, diffuse manner. T2/FLAIR hyperintense, nonenhancing if WHO II, may enhance if higher grade. MR spectroscopy (MRS) – elevated creatine-phosphocreatine peak is specific. Definitive diagnosis necessitates biopsy.

VII. Tumor: optic pathway gliomas (non-NF1). A: FLAIR-hyperintense masses involve bilateral thalami, posterior basal ganglia, and the medial temporal lobes. T1-weighted post-contrast image at the same level (B) exhibits heterogeneous nodular enhancement.

VIII. Tumor: optic pathway gliomas (known NF1). FLAIR-hyperintense masses involve bilateral thalami symmetrically, extending into the PLICs and lentiform nuclei bilaterally.

Often occur in setting of neurofibromatosis type I (NFI) but may occur sporadically. Affect thalami by direct extension as tumors involve optic pathways. FLAIR sequence most sensitive for detection. May be solid or cystic, often patchy enhancement.

IX. Pineal region tumor: dorsal midbrain germinoma involving thalami. A: FLAIR image shows hyperintense tumor (arrow) involving bilateral thalami, left greater than right. B: T1W post-contrast image demonstrates a cystic-solid neoplasm with heterogeneous enhancement (arrow).

3-8% of pediatric brain tumors. Generally divided into germ-cell (60%) origin (germinoma most common) and pineal cell origin (pineoblastoma, pineocytoma). May involve the thalami by direct extension. May cause hydrocephalus by obstructing the aqueduct of Sylvius, precocious puberty, or paresis of upward gaze (Parinaud syndrome).

X. Metabolic abnormalities: Hemolytic Uremic Syndrome (HUS). FLAIR image exhibits bithalamic and posterior lentiform hyperintensity (arrows). DWI (not shown) depicted restricted diffusion. Signal abnormalities completely resolved in both patients with correction of metabolic derangement.

Most common cause of pediatric acute renal failure. Uremia, thrombocytopenia, hemolytic anemia. Typically affects basal ganglia and dorsolateral lentiform nuclei. Occasionally affects thalami, cerebellum, brainstem. T2/FLAIR hyperintensity involving these regions, with possible restricted diffusion and/or hemorrhage. Signal normalizes following correcting underlying metabolic derangement.

CONCLUSIONS

Neuroimaging features of the abnormal thalami in the pediatric population are varied, and overlap exists when compared to similar lesions in adults.

The diagnostic workup includes a thorough history, clinical and lab exams.

Conventional MRI and, wherever applicable, advanced MR imaging techniques may greatly facilitate diagnosis and clinical management.

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