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Case Report

Dyke–Davidoff–Masson Syndrome with Right Mesial Temporal Sclerosis and Mammillary Body Atrophy: A Case Report

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Name of Author:
Dr. Harsh Kushwaha, MDRD[1], Dr. Vishnu Prasanth, MDRD[2] and Dr. Sachin Shetty, MDRD[3]

Affiliation: *[1]Postgraduate Resident, Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, India*
[2]Senior Resident, Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, India
[3]Associate Professor, Department of Radiology, Sree Balaji Medical College and Hospital, Chennai, India
Corresponding Author:
Dr. Harsh Kushwaha

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Abstract: Dyke–Davidoff–Masson syndrome (DDMS) is a rare neurological disorder characterized by unilateral cerebral hemiatrophy with compensatory calvarial thickening and sinus hyperpneumatization, typically presenting with seizures, contralateral hemiparesis, and cognitive impairment. We report a 35-year-old woman with a long-standing history of epilepsy secondary to hemiconvulsion–hemiplegia–epilepsy (HHE) syndrome who presented with recurrent seizures and headaches. Magnetic resonance imaging revealed marked atrophy of the right cerebral hemisphere with compensatory calvarial thickening and hyperpneumatization of the ipsilateral frontal and maxillary sinuses, consistent with DDMS. Additionally, the right hippocampus demonstrated atrophy and T2/FLAIR hyperintensity indicative of mesial temporal sclerosis, and the right mammillary body was notably atrophic. The coexistence of DDMS with ipsilateral mesial temporal sclerosis and mammillary body atrophy is exceptionally rare and, to our knowledge, has not been previously reported. This combination likely reflects an early, severe brain insult during infancy—in this case attributable to HHE syndrome, resulting in both widespread cortical damage and selective limbic system involvement. This case underscores the importance of comprehensive MRI evaluation in patients with chronic epilepsy, as multifocal structural abnormalities may coexist and significantly influence diagnostic precision, prognostic assessment, and therapeutic decision-making.

Keywords: Dyke–Davidoff–Masson syndrome, cerebral hemiatrophy, hemiconvulsion–hemiplegia–epilepsy syndrome, mesial temporal sclerosis, mammillary body atrophy, epilepsy, MRI.

INTRODUCTION

Dyke–Davidoff–Masson syndrome (DDMS) is a rare neurological disorder first described in 1933 by Dyke, Davidoff, and Masson, who characterized the triad of unilateral cerebral hemiatrophy, ipsilateral calvarial thickening, and hyperpneumatization of the paranasal sinuses [1,2]. The syndrome results from an insult to the developing brain during fetal or early childhood life, leading to arrested cerebral growth and subsequent compensatory osseous changes as the skull continues to develop in the absence of normal cerebral expansion [3]. Common etiologies include intrauterine vascular events, birth trauma, prolonged febrile seizures, infection, and hemorrhage [4].

Clinically, DDMS typically presents in childhood with contralateral hemiparesis, seizures, facial asymmetry, and varying degrees of cognitive impairment [5]. The left hemisphere is more frequently involved, and a male predominance has been reported, though right-sided and female cases occur [6]. Adult presentations are uncommon, and diagnosis in adulthood often occurs during evaluation for chronic, medically refractory epilepsy [7].

Hemiconvulsion–hemiplegia–epilepsy (HHE) syndrome, first described by Gastaut in 1957, is a rare condition characterized by prolonged unilateral febrile seizures in infancy followed by the development of ipsilateral hemiplegia and subsequent

epilepsy [8]. HHE syndrome is known to cause severe, unilateral brain injury and is recognized as one of the potential precursors to DDMS when the insult occurs during critical periods of brain and skull development. Mesial temporal sclerosis (MTS) is the most common cause of temporal lobe epilepsy and is characterized by neuronal loss and gliosis within the hippocampus, amygdala, and adjacent structures [9]. While MTS may occur as an isolated finding, it can also coexist with other pathologies when extensive brain injury involves both neocortical and limbic structures.

We present a unique case of DDMS in a 35-year-old female with a history of HHE syndrome, demonstrating the classic imaging findings of right cerebral hemiatrophy with compensatory skull changes, accompanied by ipsilateral mesial temporal sclerosis and mammillary body atrophy—an association not previously documented in the literature. This case highlights the importance of comprehensive neuroimaging in chronic epilepsy to identify multifocal pathology that may influence management and prognosis.

CASE PRESENTATION

A 35-year-old woman presented to the neurology outpatient department with complaints of recurrent seizures and headaches. She had a lifelong history of epilepsy, initially diagnosed with hemiconvulsion–hemiplegia–epilepsy (HHE) syndrome during childhood following a prolonged febrile illness associated with unilateral convulsions and subsequent right-sided weakness. The most recent seizure episode occurred two days prior to evaluation and involved loss of consciousness for approximately 30 minutes. According to her family, her typical seizure semiology included right-sided motor activity with secondary generalization. She experienced seizures approximately once per month despite being on regular antiepileptic medication. There was no family history of epilepsy or neurocutaneous disorders, and she had not undergone any neurosurgical procedures.

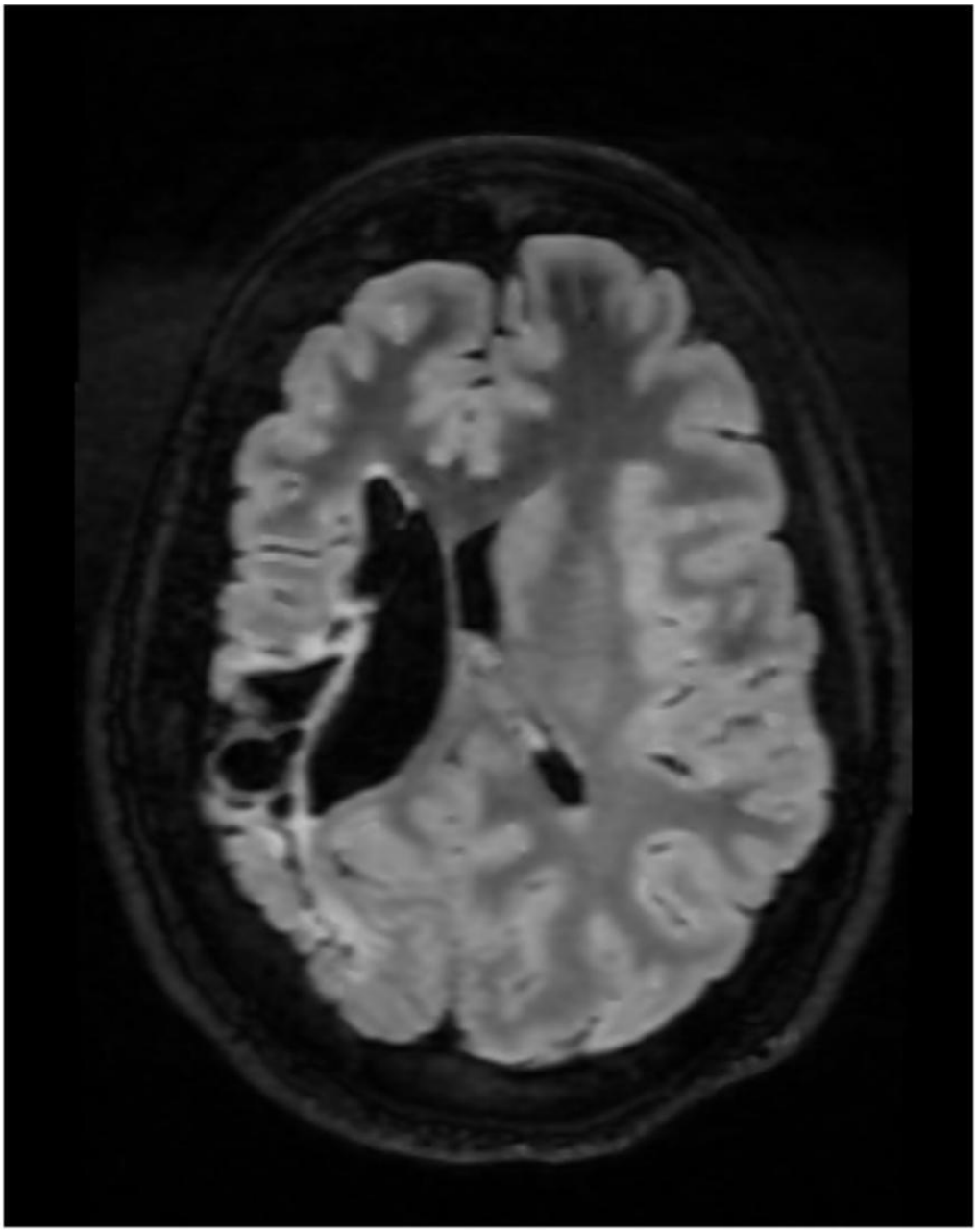

Figure 1 T2-FLAIR axial section MR Image showing right sided cystic encephalomalacia with atrophy of right cerebral hemisphere and midline shift towards right

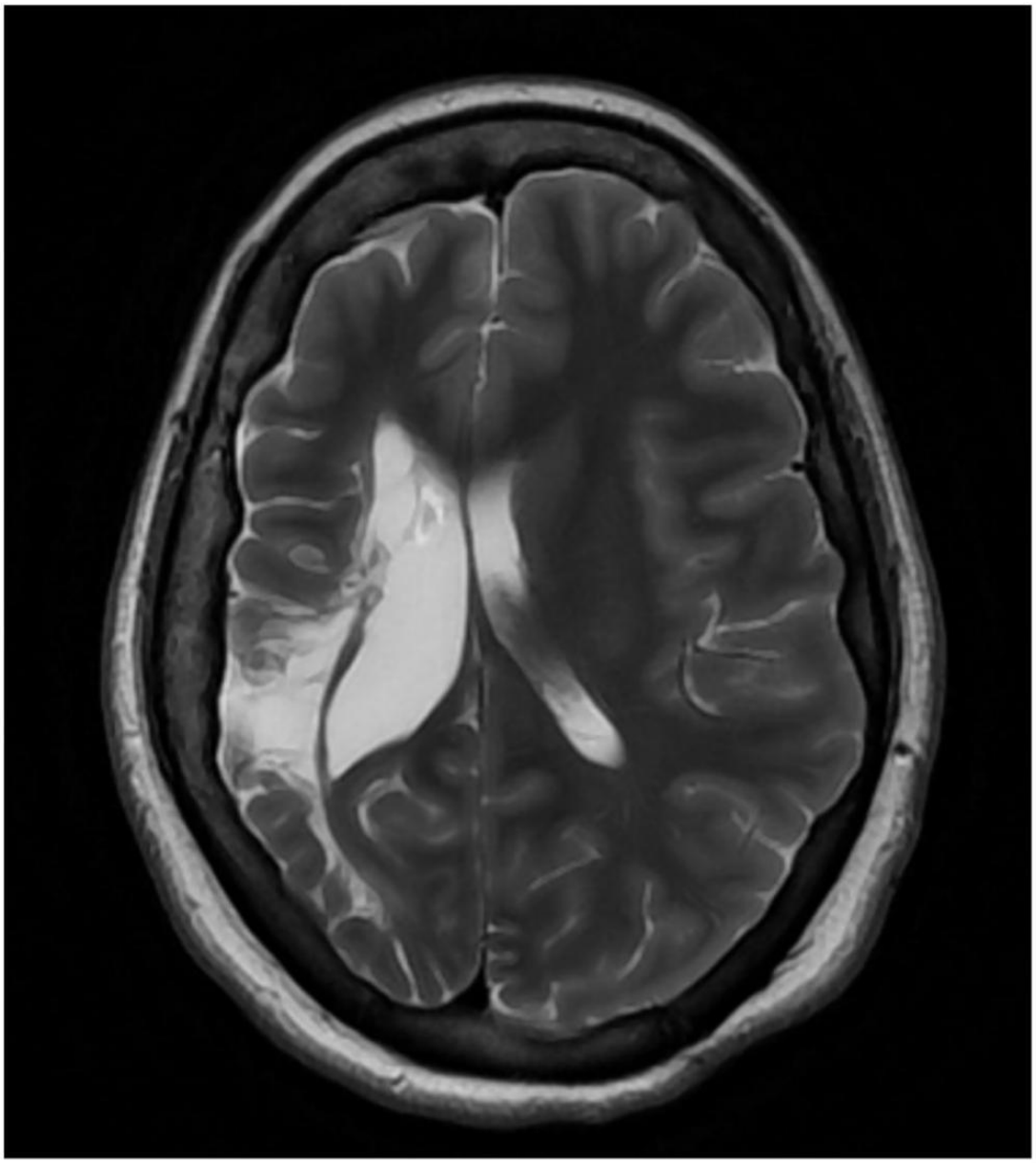

Figure 2 **T2-Weighted axial section MR Image showing right sided calvarial thickening.**

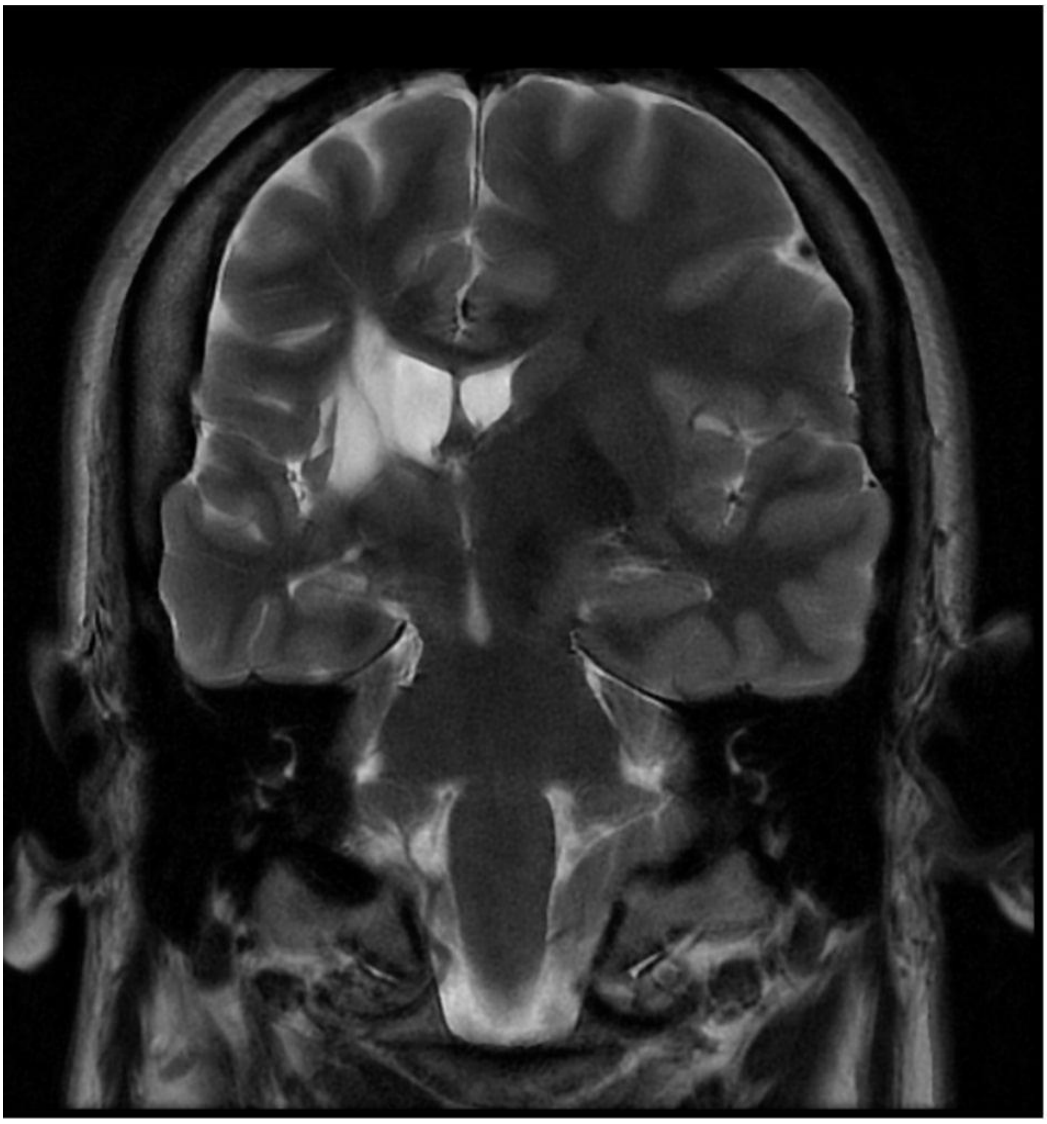

Figure 3 ***T2-Weighted coronal section MR Image showing atrophy of the right hippocampus in right MTS***

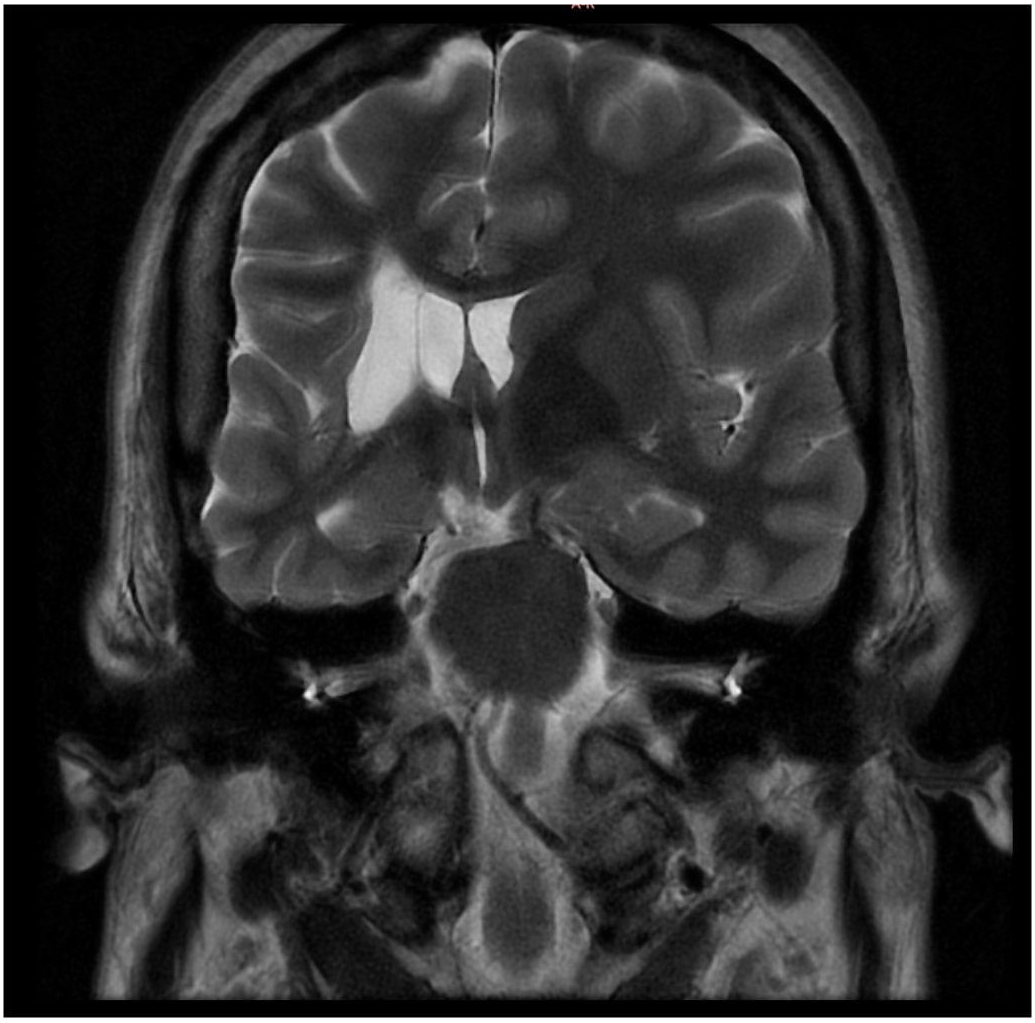

Figure 4 ***T2-Weighted coronal section MR Image showing Atrophic right mammillary body***

Clinical Examination: General physical examination was unremarkable. Neurological examination revealed mild right-sided hemiparesis (Medical Research Council grade 4/5 in both upper and lower extremities) with associated right-sided facial asymmetry. Deep tendon reflexes were brisk on the right side (grade 3+) compared to the left (grade 2+), and an extensor plantar response was elicited on the right, indicating upper motor neuron involvement. There was no sensory deficit, and cerebellar signs were absent. Cognitive function assessed by mini-mental state examination was within normal limits (score 28/30). Cranial nerve examination was unremarkable except for the previously noted facial asymmetry.

Imaging Findings: Magnetic resonance imaging (MRI) of the brain was performed using an epilepsy protocol including thin-section coronal T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences through the temporal lobes. The study revealed marked atrophy of the right cerebral hemisphere, most pronounced in the frontoparietal and occipital regions (Figure 1). Compensatory calvarial thickening was noted on the right side, along with hyperpneumatization of the right frontal and maxillary sinuses (Figure 2). Chronic gliotic changes and cystic encephalomalacia were observed in the right temporoparietal region, consistent with remote ischemic or inflammatory injury.

On dedicated temporal lobe imaging, the right hippocampus demonstrated significant atrophy and abnormal T2/FLAIR hyperintensity, meeting diagnostic criteria for mesial temporal sclerosis (Figure 3). Additionally, the right mammillary body was notably smaller than its left-sided counterpart, indicating ipsilateral mammillary body atrophy (Figure 4). No enhancing lesions, vascular malformations, or leptomeningeal angiomas were identified. The left

cerebral hemisphere, left hippocampus, and left mammillary body appeared normal.

Based on these imaging findings, a diagnosis of Dyke–Davidoff–Masson syndrome (right cerebral hemiatrophy with compensatory skull changes) with coexisting right mesial temporal sclerosis and right mammillary body atrophy was established.

Clinical Management: The patient was continued on her existing antiepileptic regimen with dose optimization. Given the presence of medically refractory seizures and the identification of unilateral mesial temporal sclerosis, surgical options including anterior temporal lobectomy were discussed with the patient and family. However, they elected to continue medical management at this time. Regular follow-up was advised for seizure monitoring and consideration of surgical intervention if seizure control remained inadequate.

DISCUSSION

Dyke–Davidoff–Masson syndrome represents a rare but important cause of chronic epilepsy and hemiparesis, with characteristic imaging findings that distinguish it from other progressive neurological disorders [3]. The pathogenesis involves an insult to the developing brain during a critical window when cerebral growth and skull development are occurring in parallel. Following unilateral cerebral injury, the affected hemisphere fails to achieve normal growth, while the skull continues to develop, resulting in compensatory ipsilateral calvarial thickening and hyperpneumatization of the paranasal sinuses as the skull expands into spaces normally occupied by brain parenchyma [4].

In our patient, the antecedent history of hemiconvulsion–hemiplegia–epilepsy (HHE) syndrome provides a compelling etiological explanation for the observed structural abnormalities. HHE syndrome typically occurs in infants and young children following prolonged unilateral febrile seizures, leading to acute hemispheric edema and subsequent atrophy [8]. The initial insult damages not only the cerebral cortex but also deep gray matter structures and the hippocampus, which is particularly vulnerable to prolonged seizure activity due to its high metabolic demand and excitatory amino acid receptor density [9]. Bhargava and Dwivedi documented that HHE syndrome frequently results in permanent structural damage, with MRI evidence of hemispheric atrophy and gliosis developing over months to years following the acute event [8].

The coexistence of DDMS with mesial temporal sclerosis is uncommon but has been reported in a small number of cases. Rondão and colleagues, in their systematic literature review of DDMS, identified that while hemispheric atrophy is universal, associated hippocampal sclerosis is documented in only a minority of cases, and its presence correlates with more severe epilepsy phenotypes [3]. The mechanism linking these two pathologies likely involves the extension of the initial insult from the neocortex to the mesial temporal structures. The hippocampus, given its anatomical location and vulnerability to excitotoxic injury, may be damaged during the same acute event that causes hemispheric injury, or it may undergo secondary damage through repeated seizure activity originating from the damaged hemisphere [9].

The additional finding of ipsilateral mammillary body atrophy in our patient further extends the spectrum of limbic system involvement. Mammillary body atrophy is most commonly associated with Wernicke–Korsakoff syndrome due to thiamine deficiency, but it can also occur via trans-synaptic degeneration following hippocampal damage [10]. The mammillary bodies receive major afferent projections from the hippocampus via the fornix, and chronic hippocampal sclerosis can lead to Wallerian degeneration of these connections, resulting in secondary mammillary body atrophy. To our knowledge, the combination of DDMS, mesial temporal sclerosis, and mammillary body atrophy has not been previously reported, making this case unique and expanding the recognized phenotypic spectrum of post-injury brain malformations.

The differential diagnosis for unilateral cerebral hemiatrophy includes several conditions. Rasmussen's encephalitis is a progressive inflammatory disorder characterized by unilateral hemispheric atrophy and intractable epilepsy, but it typically lacks the compensatory calvarial changes seen in DDMS and demonstrates progressive rather than static findings [3]. Sturge–Weber syndrome features leptomeningeal angiomatosis, facial port-wine stains, and characteristic tram-track calcifications on imaging, none of which were present in our patient. Crossed cerebral hemiatrophy with contralateral cerebellar involvement (crossed cerebellar diaschisis) may occur following large hemispheric strokes but does not produce the skull changes characteristic of DDMS [5].

The clinical implications of identifying multifocal pathology in patients with DDMS are substantial. Patients with both hemispheric atrophy and mesial temporal sclerosis may have more medically refractory epilepsy than those with hemispheric atrophy alone, as two distinct epileptogenic zones may be present [9]. Recognition of hippocampal involvement opens the possibility of surgical intervention; anterior temporal lobectomy or

amygdalohippocampectomy may be considered in patients with unilateral temporal lobe epilepsy who fail medical therapy [7]. In our patient, this option was discussed but declined at present.

The mammillary body atrophy observed in this case, while not directly contributing to seizure generation, serves as an important imaging biomarker of the extent of limbic system disruption. Its presence suggests that the original insult affected not only the hippocampus but also its efferent connections, potentially contributing to the chronicity and severity of the epilepsy syndrome [10].

This case has several limitations. The retrospective nature of the report and the absence of histopathological confirmation limit definitive conclusions about the precise etiology and temporal evolution of the findings. Additionally, detailed cognitive assessment and formal video-EEG monitoring were not performed, which would have provided valuable data regarding the functional correlates of the structural abnormalities. Long-term follow-up will be necessary to determine whether surgical intervention ultimately becomes necessary.

Despite these limitations, this case contributes importantly to the literature by documenting a previously unreported combination of findings and emphasizing the value of comprehensive neuroimaging in chronic epilepsy. For clinicians, this case reinforces several key principles: (1) patients with HHE syndrome require long-term surveillance for the development of secondary structural abnormalities; (2) dedicated epilepsy-protocol MRI with thin-section temporal lobe imaging should be performed in all patients with medically refractory epilepsy to identify surgically treatable lesions; (3) the presence of one structural abnormality does not exclude coexisting pathology; and (4) mammillary body assessment should be included in the systematic evaluation of patients with temporal lobe epilepsy, as its atrophy may provide insights into the extent and chronicity of limbic system involvement.

CONCLUSION

This case presents a rare coexistence of Dyke–Davidoff–Masson syndrome, right mesial temporal sclerosis, and ipsilateral mammillary body atrophy in a 35-year-old woman with long-standing epilepsy secondary to hemiconvulsion–hemiplegia–epilepsy syndrome. The combination of findings suggests an early, severe brain insult during infancy that produced widespread damage to both the cerebral hemisphere and the limbic system, with subsequent trans-synaptic degeneration contributing to mammillary body atrophy. Recognition of this triad emphasizes the importance of comprehensive neuroimaging evaluation in patients with chronic epilepsy, as multifocal structural abnormalities may coexist and significantly influence diagnostic accuracy, prognostic assessment, and therapeutic decision-making. This case expands the known phenotypic spectrum of post-injury brain malformations and highlights the value of dedicated epilepsy-protocol MRI in uncovering clinically significant coexisting pathology.

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