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Case Report (Open Access)

A RARE COEXISTENCE OF TUBEROUS SCLEROSIS COMPLEX AND MESIAL TEMPORAL SCLEROSIS IN AN INDIAN ADOLESCENT

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ABSTRACT
Tuberous sclerosis complex (TSC) is a genetic neurocutaneous syndrome commonly associated with epilepsy, cognitive impairment, and dermatological manifestations. Mesial temporal sclerosis (MTS), characterized by hippocampal atrophy and gliosis, is a key cause of focal epilepsy but is rarely reported alongside TSC. We present a 15-year-old male with classical dermatological signs of TSC and MRI findings consistent with both TSC and left MTS. He presented with focal seizures with impaired awareness, responded well to levetiracetam, and had normal cognitive function. Imaging revealed radial migration abnormalities and hippocampal volume loss with increased FLAIR signal. This unique case highlights the possibility of dual pathology in TSC-related epilepsy and emphasizes the need for thorough neuroimaging evaluation.

Keywords*: Tuberous Sclerosis Complex, Mesial Temporal Sclerosis, Dual Pathology, Epilepsy, Adolescent, MRI*

INTRODUCTION

Tuberous sclerosis complex (TSC) is an autosomal dominant disorder caused by mutations in either the TSC1 or TSC2 gene, resulting in widespread development of hamartomas in the brain, skin, and other organs (Curatolo *et al.,* 2008). Neurological involvement, particularly epilepsy, is seen in more than 80% of patients, frequently presenting early in life (Chu-Shore *et al.,* 2010). Mesial temporal sclerosis (MTS), also known as hippocampal sclerosis, involves selective neuronal loss and gliosis in the hippocampus and is one of the most common structural causes of temporal lobe epilepsy (Urbach, 2005).

While epilepsy is a hallmark of TSC, the association between TSC and coexisting MTS is seldom reported. Most patients with TSC-related epilepsy exhibit seizures originating from cortical tubers or subependymal nodules. MTS, if present, may either be a consequence of long-standing epilepsy or represent a separate epileptogenic focus. We present a rare Indian case of an adolescent male with clinical and radiological features of both TSC and MTS, contributing to the limited data on this dual pathology.

THE CASE

A 15-year-old male presented with a two-year history of recurrent focal seizures with impaired awareness. The episodes involved behavioral arrest and orobuccal automatisms, lasted under a minute, and had no postictal confusion. Birth and developmental history were normal. Cognitive performance and academic skills were age appropriate.

On examination, the patient had multiple facial angiofibromas (adenoma sebaceum), hypopigmented macules on the trunk, and a shagreen patch in the lumbosacral region—fulfilling dermatological diagnostic criteria for TSC. Neurological exam was normal. Interictal EEG was unremarkable.

MRI brain (seizure protocol) revealed:

- Linear T2/FLAIR hyperintensities extending from periventricular white matter to subcortical regions—suggestive of radial migration lines.
- Reduced left hippocampal volume with loss of digitations and subtle FLAIR hyperintensity—suggestive of MTS.

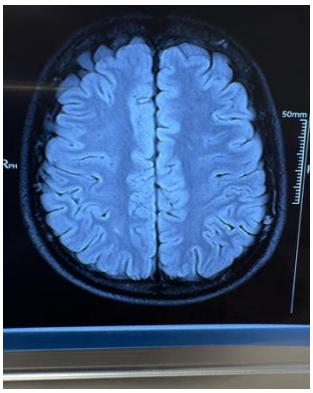

Figure 1: Axial FLAIR MRI showing subcortical linear hyperintensities in the frontal region suggestive of radial migration lines.

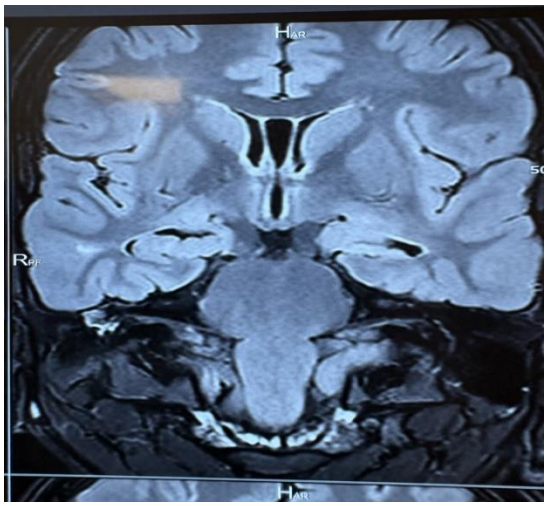

Figure 2: Coronal FLAIR MRI showing reduced left hippocampal volume and loss of digitations – suggestive of mesial temporal sclerosis.

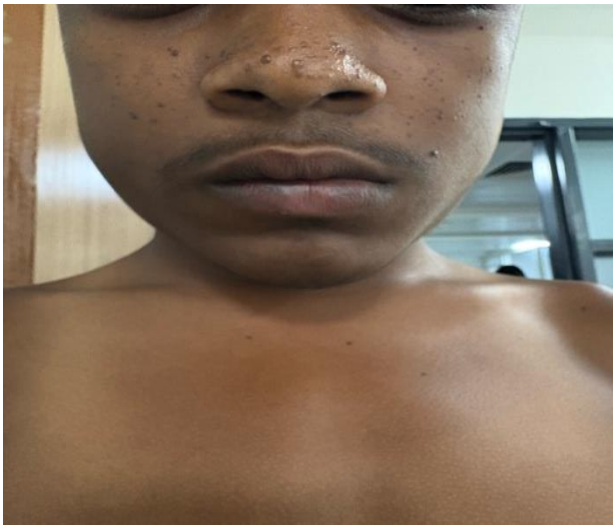

Figure 3: Facial angiofibromas (adenoma sebaceum) over the central face, characteristic of tuberous sclerosis.

Figure 4: Hypopigmented macules (ash-leaf spots) and shagreen patch on the lower back in tuberous sclerosis.

He was started on levetiracetam (20 mg/kg/day), following which he became seizure-free. Renal and cardiac evaluations were normal. Genetic testing could not be performed due to cost constraints

Discussion

TSC is a multisystem disorder characterized by hamartomas affecting the brain, kidneys, heart, and skin. Epilepsy is the most common neurological manifestation, typically associated with cortical tubers and subependymal nodules (Curatolo *et al.,* 2008; Widjaja *et al.,* 2020). Mesial temporal sclerosis is the most frequent cause of drug-resistant temporal lobe epilepsy and is identified by hippocampal atrophy, increased T2/FLAIR signal, and loss of digitations (Urbach, 2005; Cascino, 2009).

Although seizures are common in TSC, coexistent MTS is rare. Some surgical series have described dual pathology in TSC patients undergoing epilepsy surgery (Barba C, *et al.,* 2016; Devlin *et al.,* 2006).

It is postulated that recurrent seizures in early life could contribute to hippocampal sclerosis (Kameyama *et al.,* 2000). However, in our case, the absence of early-onset seizures or developmental delay suggests an independent occurrence.

Recognition of dual pathology is important for prognosis and surgical planning. While our patient remains well-controlled on monotherapy, identifying MTS in TSC patients with refractory epilepsy may influence consideration for selective amygdalohippocampectomy.

CONCLUSION

The coexistence of tuberous sclerosis complex and mesial temporal sclerosis is a rare but clinically relevant combination. High-resolution MRI with epilepsy protocol should be routinely performed in TSC patients presenting with seizures. Recognition of dual pathology can help guide individualized treatment, especially for surgical candidates.

Declaration of patient consent

The authors certify that they have obtained appropriate consent from the patient's guardian. Consent includes the use of images and clinical information for publication while maintaining anonymity.

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