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Caught Between Stroke and Vasculitis: A Complex Case in Pediatric Thalassemia

Phylicia Lim Yi-Tynn, Medical Officer[1]
Tan Chung Yung, Emergency Physician[1]
Abdul Kursi Bin Latif, Emergency Consultant[1]
[1]Hospital Raja Permaisuri Bainun, Ipoh, Perak, Malaysia

Introduction

Cerebrovascular accidents (CVAs), though uncommon in children, may occur in the setting of hematologic disorders associated with a hypercoagulable state. Beta thalassemia intermedia, while generally milder than thalassemia major, carries a risk of thromboembolic complications, including silent and overt cerebral infarctions. The pathogenesis is multifactorial, involving chronic anemia, endothelial dysfunction, thrombocytosis (especially post-splenectomy), and iron overload. We present a case of acute ischemic stroke in a 9-year-old boy with transfusion-dependent beta thalassemia intermedia, highlighting the diagnostic complexity in distinguishing thromboembolic stroke from primary central nervous system (CNS) vasculitis.

Case Description

A 9-year-old boy with transfusion-dependent beta thalassemia intermedia and iron overload presented with acute right lower limb weakness. On arrival, he was hemodynamically stable; neurological examination revealed flaccid paralysis of the right lower limb with hypotonia and areflexia. Cardiovascular exam noted a grade 3 ejection systolic murmur. Blood investigations were unremarkable. Urgent CT brain showed a left frontal acute infarct. MRI revealed multifocal acute infarcts in several arterial territories with occlusions in both anterior cerebral arteries (ACAs) and the left middle cerebral artery (MCA), suggesting a thromboembolic event. Echocardiography showed preserved ejection fraction (69%) with trace tricuspid regurgitation and no thrombus. Carotid Doppler identified narrowing of the right internal carotid artery. He was started on aspirin (75 mg daily). Multidisciplinary consultations supported a working diagnosis of primary CNS vasculitis. He was managed conservatively and discharged in stable condition after four days.

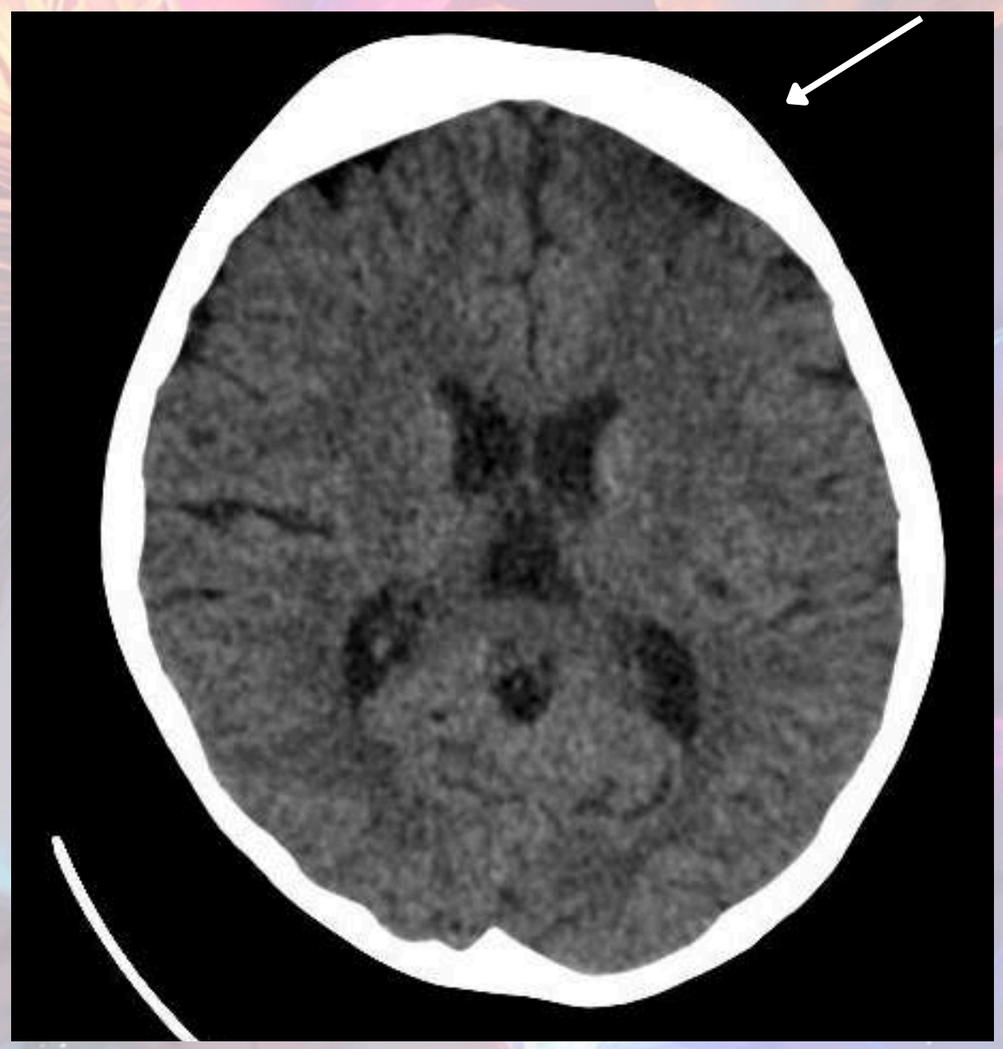

Figure 1: Left frontal lobe hypodense area represents acute infarct

Discussion

This case highlights a rare but significant neurological complication of beta thalassemia intermedia—an acute ischemic stroke in a pediatric patient with overlapping features of thromboembolism and primary CNS vasculitis. While beta thalassemia intermedia is often considered a milder phenotype, it is increasingly recognized to carry a prothrombotic risk, particularly in transfusion-dependent patients with iron overload and post-splenectomy thrombocytosis. In this case, the presence of multifocal infarcts across multiple arterial territories, alongside angiographic evidence of vascular narrowing, raised the possibility of primary CNS vasculitis. Differentiating thromboembolic stroke from vasculitis in thalassemia patients is inherently challenging due to the overlap in clinical and radiographic findings. The concordance of large-vessel occlusions, normal cardiac evaluation, and segmental arterial narrowing ultimately supported a working diagnosis of primary CNS vasculitis, prompting conservative management with antiplatelet therapy and close monitoring. It also raises important questions about the role of cerebrovascular screening in high-risk thalassemia patients. While routine neuroimaging is not standard in asymptomatic children, early identification of silent infarcts or vasculopathy may facilitate timely intervention.

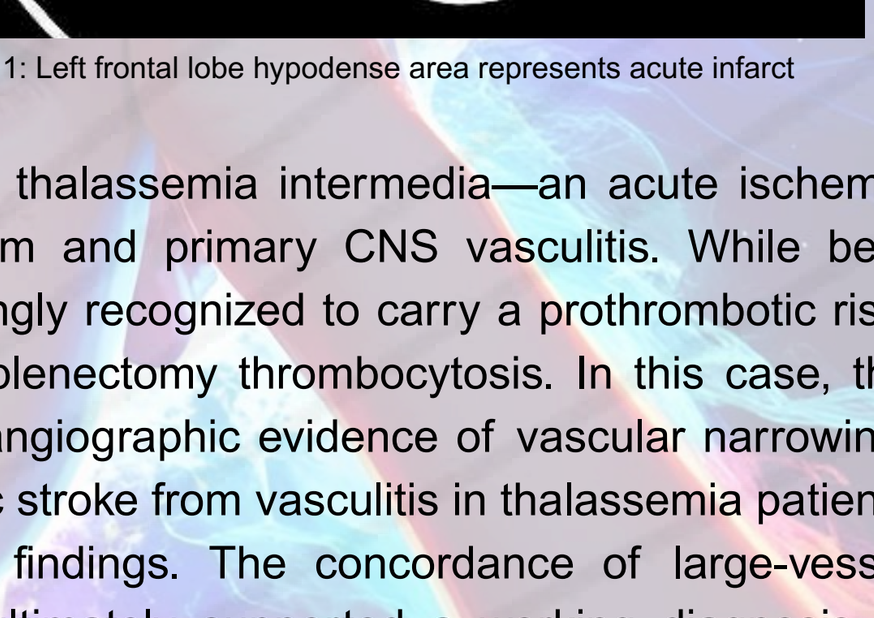

Figure 2: Challenges in Diagnosing Pediatric Stroke in ED

Conclusion

Pediatric stroke in beta thalassemia intermedia presents significant diagnostic and therapeutic challenges, necessitating individualized, multidisciplinary care and vigilance for vascular complications.

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