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Case Report

Cerebral hypometabolism in a pediatric patient with clinically resolved posterior reversible encephalopathy syndrome ☆,☆☆

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ABSTRACT

A 4-year-old boy with Nuclear factor-kappa B Essential Modulator deficiency syndrome presented with encephalopathy post haematopoietic stem cell transplantation. MRI demonstrated T2/FLAIR-hyperintensities in the posterior cerebral cortex concerning for posterior reversible encephalopathy syndrome. Clinical improvement was appreciated following withdrawal of the suspected offending pharmacological agent (Cyclosporine). An ^{18}F-FDG PET/CT performed 2 months later to screen for post-transplant lymphoproliferative disease demonstrated markedly reduced FDG uptake in the posterior cerebral cortex, involving the parietal and occipital lobes. We describe, to the best of our knowledge, the first case of profound cerebral hypometabolism in a child with clinically resolved posterior reversible encephalopathy syndrome.

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Introduction

Posterior reversible encephalopathy syndrome (PRES) is a neurological disorder characterized by reversible, bilateral cerebral oedema. Magnetic resonance imaging (MRI) changes are thought to normalize following clinical resolution in the vast majority of pediatric cases of PRES [1]. Our aim is to report the first pediatric case of profound cerebral hypometabolism detected on 2-deoxy-2-[^{18}F]fluoro-D-glucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) in a 4-year-old patient with clinically resolved PRES to alert pediatric health care providers and imaging specialists about this phenomenon and possible long term implications.

Case presentation

A 4-year-old boy with Nuclear factor-kappa B Essential Modulator (NEMO) deficiency syndrome presented with encephalopathy and hypertension 4 months post hematopoietic stem cell transplantation. There was no clinical or electrographic evidence of seizures. CT Brain demonstrated parenchymal hypoattenuation involving the cortex and subcortical white matter in bilateral parietal and occipital lobes (Fig. 1A). MRI Brain demonstrated hyperintense T2-weighted/fluid-attenuated inversion recovery foci in the aforementioned regions (Fig. 1B), supporting the diagnosis of PRES. Following management with anti-hypertensive medication and withdrawal of the suspected offending agent

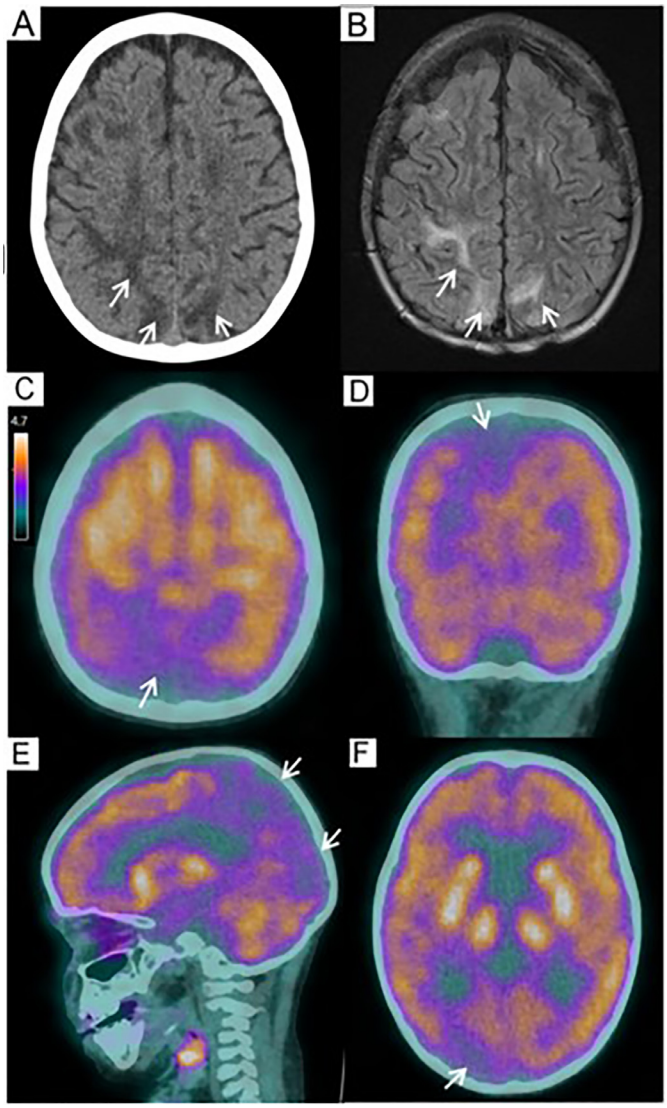

Fig. 1 – Axial computed tomography brain (A) at initial presentation demonstrated parenchymal hypoattenuation (white arrows in A) involving the cortex and subcortical white matter in bilateral parietal lobes. Axial magnetic resonance imaging T2-weighted/fluid-attenuated inversion recovery image (B) also demonstrated hyperintense foci (white arrows in B) in the aforementioned regions, supporting the diagnosis of posterior reversible encephalopathy syndrome (PRES). ^{18}F-FDG PET/CT was performed 2 months following clinical resolution of PRES. Fused axial (C), coronal (D), sagittal and further axial PET/CT demonstrated markedly reduced FDG uptake in the posterior cerebral cortex involving the posterior parietal and occipital lobes (white arrows in C-F), more extensive on the right.

(Cyclosporine), clinical resolution ensued within 1 week. Cyclosporine was re-introduced at a lower dose 1 month later without recurrence of symptoms. An ^{18}F-FDG PET/CT scan was arranged 2 months later to screen for post-transplant lymphoproliferative disease (PTLD) in the setting of respiratory deterioration and Epstein-Barr viraemia (Figs. 1C-F). Of note, the patient had remained seizure free. There was no scan evidence of hypermetabolic lymphomatous disease to suggest the presence of PTLD. However, there was markedly reduced FDG uptake in the cerebral cortex involving the posterior parietal and occipital lobes despite the absence of clinical encephalopathy. Neither progress MRI Brain nor formal cognitive testing had occurred thus far.

Discussion

PRES is a neurological disorder characterised by reversible, bilateral cerebral oedema. Pathogenesis is thought to involve dysregulation of physiologic auto-regulatory vasoconstriction [2]. PRES commonly presents with headache, encephalopathy, seizures and visual disturbance [3]. Well identified triggers include hypertension and immunosuppressive agents [2]. Prompt recognition, leading to targeted management and withdrawal of offending agents often results in clinical reversal within days to weeks. However, delayed diagnosis and lack of treatment may result in permanent brain damage [4].

Molecular imaging findings of PRES, particularly in the pediatric population, remain sparsely documented. Both increased and decreased focal FDG uptake in the posterior cerebral cortex have been described during the acute phase, though there is no published literature on residual PET changes [5,6]. MRI changes are thought to resolve in the majority of cases. In a prospective study of 24 children with PRES, Darwish reported residual MRI changes at 2 months in 2/24 (8.3%) patients and at 24 months in 1/24 patients with clinically resolved, first episode PRES [7]. Additionally, there was a significant association between recurrent PRES, residual MRI changes and the development of epilepsy [7]. On subsequent formal cognitive testing, only 1 patient with neuroblastoma had below average intelligence though prior chemotherapy may have contributed to this [7]. While our patient has not had further MRI Brain and the significance of FDG PET abnormalities following clinical resolution of PRES is unknown (and has not been reported previously), close clinical follow up (paying particular attention to seizures) and cognitive testing is prudent in view of the cerebral metabolic abnormalities.

Conclusion

To the best of our knowledge, the present case is the first report of cerebral hypometabolism detected on ^{18}F-FDG PET/CT in clinically resolved, pediatric PRES. It is vital that pediatric health care providers and imaging specialists are aware of this phenomenon, so as to provide close follow up. Additional research is required to understand the long term impacts of metabolic changes on ^{18}F-FDG PET/CT and how these are associated with initial or residual MRI changes.

Patient consent

Personal patient information was removed from the presented radiology images and appropriate patient consent was acquired.

CRediT authorship contribution statement

Karan Singh: Conceptualization, Writing – original draft, Visualization. **Jeanette Taylor:** Writing – review & editing. **Adam Nelson:** Writing – review & editing. **Richard Mitchell:** Writing – review & editing. **Ivan Ho Shon:** Conceptualization, Writing – review & editing, Visualization, Supervision.

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