¹⁸F-FDG PET/CT in pachygyria during evaluation for seizure disorder

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(Received 31 May 2020, Revised 2 November 2020, Accepted 9 November 2020)

ABSTRACT

Pachygyria or incomplete lissencephaly is a developmental condition due to abnormal migration of neurons. The association of seizures in this condition warrants investigation like electroencephalogram (EEG) and magnetic resonance imaging (MRI). ¹⁸F-flurodeoxyglucose positron emission topography computed topography (¹⁸F-FDG PET/CT) has a potential role in commenting of wide distribution of abnormal metabolism in affected brain parenchyma as well as hypermetabolic epileptogenic focus during ictal phase. Scan during interictal phase will give an idea of affected regions in brain in the form of hypometabolic areas. Although treatment is long-term antiepileptic drugs however if the epileptogenic foci are localized to same region then surgical interventions like hemispherectomies are the options. We present a case in which ¹⁸F-FDG PET/CT proved beneficial in pachygyria with seizure disorder.

Key words: Pachygyria; Seizure; Epileptogenic focus; ¹⁸F-FDG PET/CT

Iran J Nucl Med 2021;29(1):49-51 Published: January, 2021 http://irjnm.tums.ac.ir

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INTRODUCTION

The cerebral cortex is responsible for conscious movement and thought, and should have deep convolutions (gyri) and grooves (sulci), which are formed by "infolding" of the cerebral cortex. Malformations of cortical brain development that occur can be broadly classified as per the underlying mechanisms like: abnormal cell proliferation, abnormal neuronal migration and abnormal cortical organization [1].

During normal embryonic growth, cells that later develop into specialized nerve cells (neurons) migrate to the brain's surface, making several layers of cells. Disruption in normal migration of cells to their locations, results in too few cell layers, absence or incomplete development of gyri. Environmental factors that contribute to the condition may include intrauterine infection during pregnancy (such as a virus), and insufficient flow of oxygenated blood to the brain (ischemia) during fetal development [2].

A spectrum of disease caused due to disruption in normal migration of post mitotic neurons from the region of ventricles to cortical plate leading to relative smoothness of sulci appreciated in imaging modalities and is termed as lissencephaly-pachygyria. The spectrum includes agyria (no gyri), pachygyria (broad gyri) and lissencephaly (smooth brain surface) [3].

CASE PRESENTATION

A 6 year child was referred for PET-CT by department of neurology for imaging in seizure disorder

The child was born out of non-consanguinous marriage and delivered at 7 months of gestation. There were no pre, peri and post-natal complications. Developmental milestones were delayed, there was poor muscle tone, motor functions and intellectual disability. The child had recurrent seizures since the age of 03 months. EEG revealed – continuous runs of spikes and polyspike discharges lateralizing to left hemisphere with occasional suppression of background electrical activity.

MRI brain revealed focal hamartomatous overgrowth of left occipital lobe, parietal lobe and superior part of temporal lobe with pachygyria. Loss of grey white differentiation was noted and associated chinking of occipital horn and temporal horn of left lateral ventricle [Figure 1]. He was diagnosed as a case of left hemispheric cerebral palsy and was managed for recurrent seizures with antiepileptic medications of no great help.

Inter ictal PET-CT was performed and findings revealed megaencephaly of left cerebrum with pachygyria. PET revealed hypometabolic areas in middle and superior temporal lobe and in the area adjacent to central sulcus on the left cerebral hemisphere – correlating with areas of abnormal electrical activity in EEG [Figures 2 and 3].

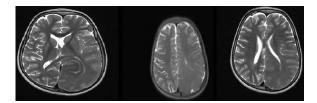


Fig 1. MRI images suggestive of focal hamartomatous overgrowth of left occipital lobe, parietal lobe and superior part of temporal lobewith broad gyriand shallow sulci

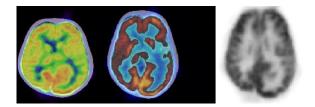


Fig 2. ¹⁸F-FDG PET/CT axial sections of brain show that left cerebrum appears bulky as compared to contralateral side. Distinction of the gyri and sulci are lost in the left cerebrum. Chinking of anterior, lateral and posterior horn of left lateral ventricle is seen and midline shift towards right is noted. The MIP images in axial section show hypometabolism is seen in the middle and superior temporal gyrus on left side and in areas adjacent to left central gyrus (SUV max=4.2 with respect to SUV max=5.4 on contralateral side).

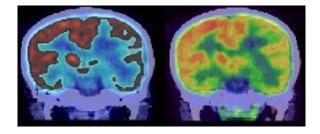


Fig 3. ¹⁸F-FDG PET/CT in coronal section of brain show that left cerebrum appears bulky and distinction of the gyri and sulci are lost. There is relative hypometabolism on left side.

DISCUSSION

Pachygyria is a developmental condition in which abnormal migration of nerve cells takes place with incidence rate of 1 in 100000 [4]. It leads to developing brain with less number of gyri that are broad and flat. This is also known as incomplete lissencephaly. It can occur alone or associated with underlying syndrome. Symptoms include moderate to severe developmental delay, seizures, poor muscle tone and control and feeding/swallowing difficulty [5].

As per existing literature, only 3 cases of occipital pachygyria and 7 cases of frontotemporal pachygyria

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have been reported [6]. Pachygyria presents in early childhood with delayed milestones and recurrent seizures. Facial dysmorphism can be associated with normal head circumference or associated with microcephaly, telecanthus, esotropia and/or exotropia. Mental retardation and hypotonia with or without pyramidal and cerebellar signs can be seen [7]. Traditionally, the diagnosis of such conditions was based on post-mortem examination of the brain. Although lissencephaly/pachygyria can be identified on all cross-sectional modalities (antenatal and neonatal ultrasound, CT and MRI), MRI is the modality of choice to fully characterise the abnormalities.

Because of the varied spectrum of symptoms of this condition, treatment is symptomatic, often with antiseizure medication. Special education and training consisting of physical, occupational, and speech therapies are helpful in improving quality of life [8].

Surgical procedures like hemespherectomies and multilobar resections are the options for sulcal-gyral abnormalities including polymicrogyria, pachygyria, lissencephaly and schizencephaly. Although MRI scan will show, the extent of abnormality but usually ¹⁸F-FDG PET/CT show more extensive cortical involvement in comparison to MRI because it covers the subtle areas with metabolic abnormalities in the affected cortex.

Abnormal neuroelectric discharges can be localized during ictal ¹⁸F-FDG PET/CT as abnormal area of hypermetabolism thus localizing area of dysfunctional pachygyria [9, 10]

In conclusion, neuroimaging findings have a varied pattern and play a critical role in pachygyria. In this case, MRI showed regions of anatomical abnormality in the affected hemisphere and conventional investigation could give an idea of abnormal focus of seizure. In contrast, ¹⁸F-FDG PET/CT showed the pattern of metabolic distribution of abnormal foci within the involved cerebral hemisphere. ¹⁸F-FDG PET/CT had an added benefit of providing unequivocal imaging results with high degree of confidence corresponding to the areas of abnormality in MRI thus guiding further management.

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