

## **Diffusion Tensor imaging findings in a patient with Panayiotopoulos syndrome**

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### **Introduction:**

Panayiotopoulos syndrome or benign epilepsy with occipital paroxysms is a common benign epilepsy syndrome described in children. Patients generally have infrequent seizures, mainly occurring at night with multitude of autonomic symptoms such as emesis, pallor, tachycardia and visual hallucinations. Inter-ictal EEG findings most commonly demonstrate nearly continuous unilateral or bilateral high voltage spike-wave discharges from the occipital or posterior temporal regions.

We present a patient with clinical symptoms and electroencephalographic (EEG) findings consistent with Panayiotopoulos syndrome and diffusion tensor imaging of her brain showing attenuation of fibers over the right temporal-occipital area and decreased thickness of the right occipital lobe. Previous scientific literature on Panayiotopoulos syndrome has discussed the brain MRI and functional MRI findings. However, to the best of our knowledge, diffusion tensor studies in patients with Panayiotopoulos syndrome have not been reported. Our case report suggests that subtle structural lesions may be a part of the pathophysiology of Panayiotopoulos syndrome.

### **Case report:**

A 6-year-old right-handed previously healthy Caucasian female woke up one night, after being asleep for about an hour, and told her father that she had a bad dream. She was retching and vomited out partially digested food. Her father described her as staring and unresponsive with a dazed, confused look in her eyes. The episode lasted for about 30 minutes and she had incoherent speech for about an hour. No generalized tonic-clonic movements were witnessed. After about an hour she returned to her baseline.

She had similar episodes 2 weeks and also three months prior to the current event. Her developmental, birth and maternal and family history were unremarkable. Her general physical and neurological examination was unremarkable.

She was initially taken to a nearby hospital and was given ondansetron and lorazepam. The initial non-contrast head CT was unremarkable. Lab work-up was unremarkable; except for an elevated white cell count of 16,800. Urine drug screen was negative.

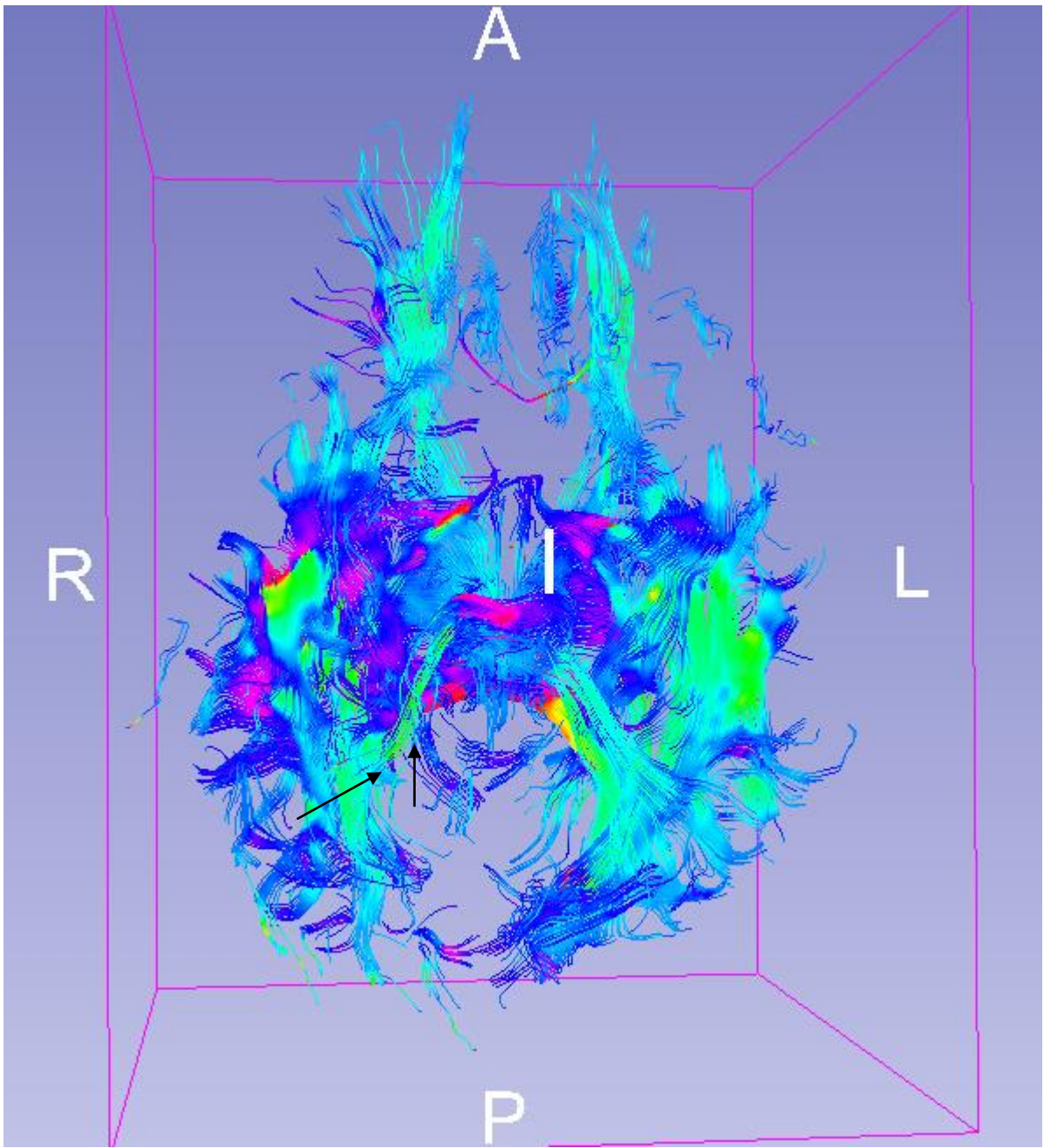
EEG (Figure 1) showed frequent, 200-300 microvolt spike and wave discharges, predominantly seen over the bilateral occipital derivations occurring synchronously and occasionally independently. They were seen much more prominently during photic stimulation as well as during sleep. Both the clinical semiology of her spells and EEG findings were consistent with the diagnosis of Panayiotopoulos syndrome.

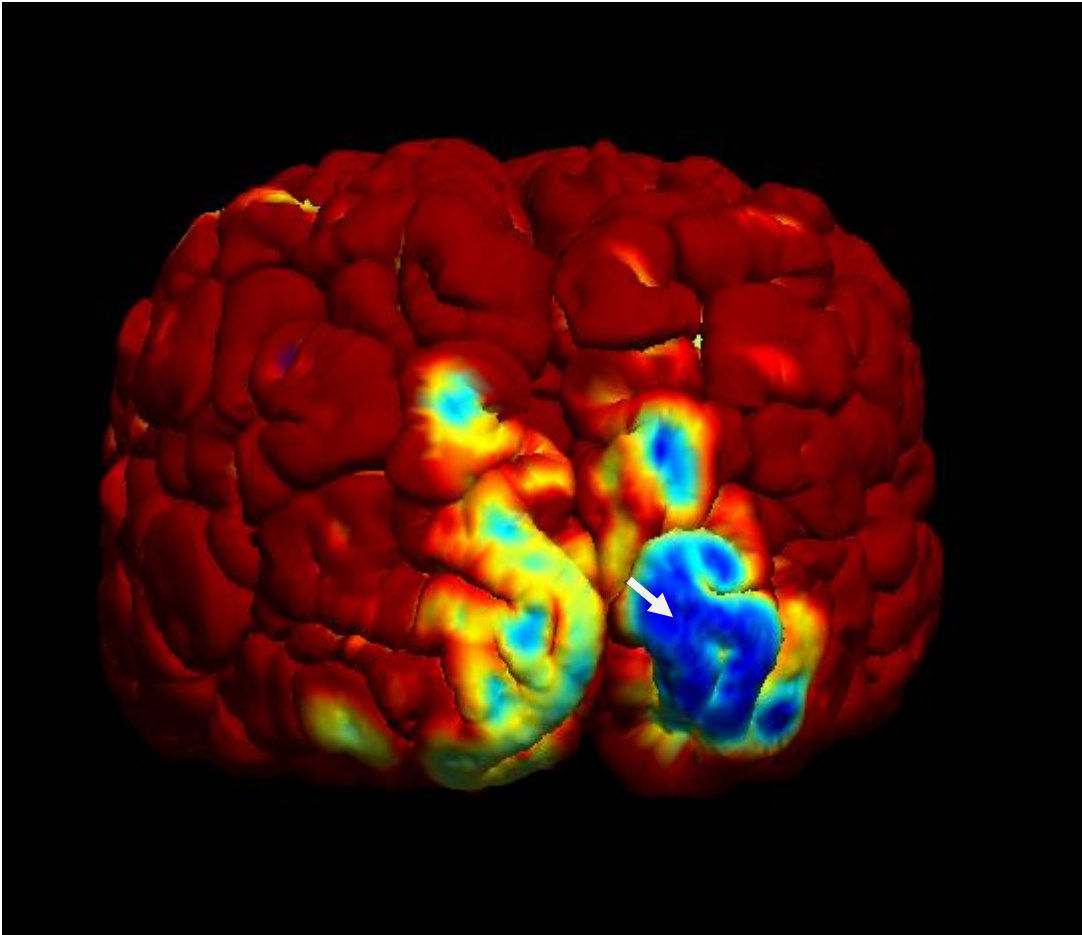
Non-contrast brain MRI was unremarkable. A Diffusion tensor imaging was performed and showed attenuation of fibers over the right temporal-occipital area (Figure 2a) and decreased thickness of the right occipital lobe (Figure 2b). She was started on Oxcarbazepine and is

currently being followed up in the clinic. Since then she had a total of 4 similar events in a period of over one year after the initial event.



**Figure 1:** Routine EEG (longitudinal bipolar montage) showing frequent occipital epileptiform discharges





**Figures 2a and 2b:** DTI showing attenuation of white matter tracts over the right occipital lobe

#### **Discussion:**

Panayiotopoulos syndrome or benign epilepsy with occipital paroxysms is an uncommon benign epilepsy syndrome described in children. While there are several case reports and case series describing the MRI findings in patients with Panayiotopoulos syndrome, there is no literature on the diffusion tensor imaging (DTI) in these patients.

Lada C et al [1] reported 43 patients with Panayiotopoulos syndrome, where they described that most had normal neuroimaging. Yeom et al [2] reported a 4 year old girl with Panayiotopoulos syndrome whose MRI showed a slightly high signal on T2 weighted images in the left hippocampus.

Iannetti et al [3] described an 8 year old girl with Panayiotopoulos syndrome and a normal MRI, whose ictal EEG showed a diffuse onset of the ictal discharge despite an interictal electroencephalography showing infrequent multifocal spikes, favoring the idea of a diffuse and multifocal cortical epileptogenicity as the underlying mechanism in this syndrome.

On the other hand Leal et al [4] & [5] proposed that a rapid spread of posterior epileptic activity in the Panayiotopoulos syndrome is responsible for the variable and poorly localized discharges seen on EEG, with the lateral occipital cortex being the primary generator of the epileptic activity. This theory is supported by Leal et al's description of the functional MRI findings in a patient with the syndrome showing that the spike-related blood oxygen level dependent (BOLD) effect was multifocal, with clusters in the lateral and inferior occipital lobe, and the lateral and anterior temporal lobe. Regional dipole seeding in BOLD clusters strongly suggested that the origin of

epileptic activity was in the lateral occipital area with the spread to the occipital pole and lateral temporal lobe. However, a more recent report also by Leal et al [6] described a 5 year old girl with a lesion on the inferior parietal lobe, with seizure semiology suggestive of Panayiotopoulos syndrome.

Yalcin et al [7] described 3 out of 53 patients with Panayiotopoulos syndrome with coincidental brain lesions, namely a neuroepithelial cyst, a right occipital encephalomalacia and an arachnoid cyst located in the cisterna magna associated with colpocephaly, thus suggesting a focal generator as well.

An article by T.Dura-trave et al [8] discussed the imaging findings among 32 patients of Panayiotopoulos syndrome where MRI was done in 15 patients. Two of the 15 patients had abnormalities, including a pineal cyst and right parietal white matter atrophy.

Sakagami et al [9] described the SPECT findings in a 7 year old girl with Panayiotopoulos syndrome whose MRI and MR angiography were normal. Interictal single photon emission computed tomography (SPECT) using  $^{99m}\text{Tc}$ -ethyl-cysteinate dimer revealed decreased cerebral blood flow in the right occipital region corresponding to the epileptogenic focus on EEG, again suggesting a focal seizure generator.

#### **Teaching Points:**

Patients with Panayiotopoulos syndrome were reported to have varied focal pathologies on MRI scans. In our patient while the MRI was unremarkable, DTI sequencing found decreased white matter fibers in right occipital lobe. Whether this is a coincidental finding or suggestive of a focal pathology contributing to the pathogenesis of Panayiotopoulos syndrome is yet unclear. The major limitation of our case report is whether a single case showing DTI abnormality, in the absence of normal MRI brain abnormality, can explain the neuroanatomical basis of this unique epilepsy syndrome. In addition, asymmetry of the DTI images can lead to misinterpretation and can be a limitation in assessing the fiber density. We suggest that more DTI sequencing data from Patients with Panayiotopoulos syndrome may help in clarifying the underlying abnormality in this unique epilepsy syndrome.

#### **References:**

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