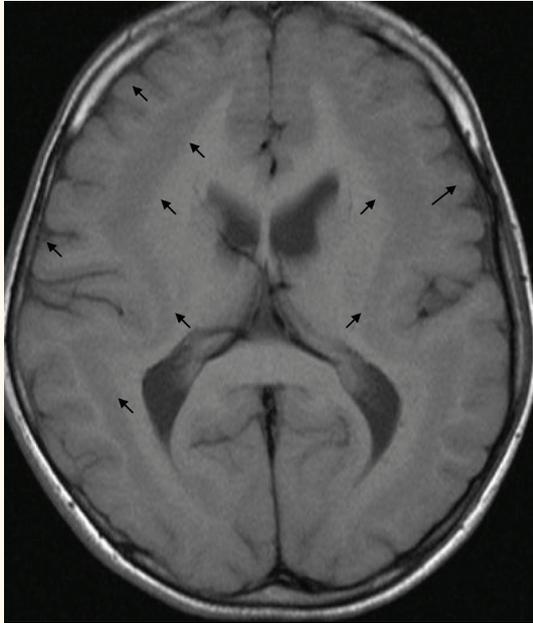


## A Ten year-old Girl with Band Heterotopia or Double Cortex (two brains)

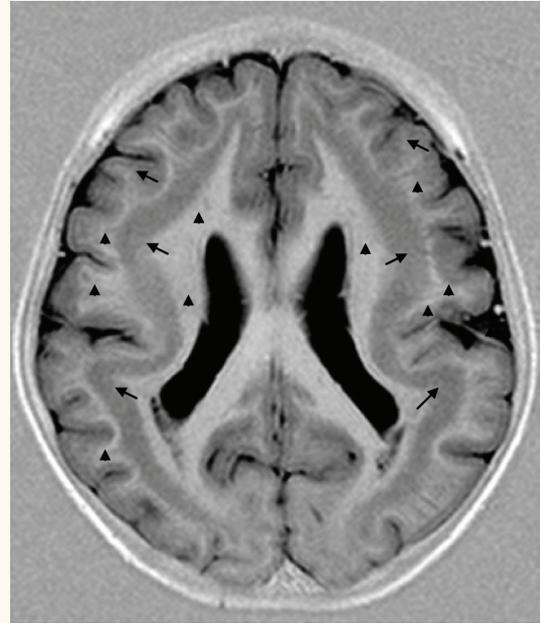
\*Roshan Koul,<sup>1</sup> Amna Alfutaisi,<sup>1</sup> Faisal Alazri,<sup>2</sup> Rajeev Jain<sup>3</sup>

طفلة عمرها عشر سنوات بتوضع مغاير شريطي أو قشرة مزدوجة  
(دماغين)

روشان كول، أمينة الفطيسي، فيصل العزري، راجيف جين



**Figure 1:** Magnetic resonance imaging T1 weighted axial image of the brain shows double cortex. Arrows indicate two layers of grey matter separated by bright looking white matter.



**Figure 2:** Magnetic resonance imaging T1 weighted axial image (inversion recovery), shows bilateral band heterotopia (double cortex). Large arrows denote grey matter and small arrows white matter.

**A**TEN-YEAR GIRL PRESENTED AT SULTAN Qaboos University Hospital with poor school performance and generalised tonic clonic seizures in the previous three months. Her birth history and antenatal history were normal. She also had poor school performance. Her other siblings were normal. Her general physical examination and neurological examination was unremarkable. There were no neurocutaneous lesions. An assessment revealed an IQ of 57, which

is very low. The routine blood work up was normal. An electroencephalogram showed left temporal region seizure discharges with generalisation. A magnetic resonance imaging scan of the brain revealed complete band heterotopias in both hemispheres (double cortex) [Figures 1 and 2].

Neuronal migration disorders (NMD), are seen in children with psychomotor developmental delay, epilepsy and mental retardation. Physical examination is usually normal in these children

except neurocutaneous disorders. Minor forms of NMD are picked up only when imaging is done. The normal human brain goes through three major phases of morphogenesis which are neuronal production, neuronal migration and differentiation. This mechanism is complex and chemical signals and guides control this.<sup>1</sup> Neuronal production and migration starts at six weeks of gestation and proceeds until twenty-six weeks. After this phase, differentiation and maturation continue until the age of 15 years.<sup>1</sup> Heterotopias are one of the NMDs, where in ectopic grey matter cells get arrested in inappropriate sites in the brain. The cells may be arrested between the leptomeninges and periventricular region. The cells arrested in the subependymal region are called subependymal nodular heterotopias. The cells arrested below the proper cortex and separated from the cortex by a thin band of white matter, are called subcortical band heterotopias (SBH). When this band is circumferential beneath the cortex, it is called a double cortex as was seen in our child. This condition is very rare with only about 120 cases reported in the world medical literature.<sup>2</sup> The syndrome is usually associated with mutations in the doublecortin (DCX) (Xq22.3-q23) gene, and much less frequently in the LIS1 (17p13.3) gene. The majority of patients with SBH are sporadic, most

patients are females and few patients with familial X-linked inheritance have been seen.<sup>2</sup> Heterotopias may be isolated or part of a syndrome. The exact incidence of heterotopias in the general population is unknown; however, heterotopias formed 11.6% of cases of NMD in our series.<sup>3</sup> Only one of them had SBH, forming about one percent of NMD.

#### ACKNOWLEDGMENT

This case was presented on 12<sup>th</sup> November 2008 in the SQU Clinical-Pathological Conference Series (SCRAPS) as: "Are two brains better than one?"

1. Caviness VS Jr, Goto T, Tarui T, Takahashi T, Bhide PG, Nowakowski RS. Cell output, cell cycle duration and neuronal specification: a model of integrated mechanisms of the neocortical proliferative process. *Cerebral Cortex* 2003; 13:592–8.
2. D'Agostino MD, Bernasconi A, Das S, Bastos A, Valerio RM, Palmini A, et al. Subcortical band heterotopia (SBH) in males: clinical, imaging and genetic findings in comparison with females. *Brain* 2002; 125:2507–22.
3. Koul RL, Alfutaisi A, Sankhala DK, Javad H, William RR. Pattern of childhood neuronal migration disorders in Oman. *Neurosciences* 2009; 14:158–62.