RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 1 | Jan 2015 | ISSN - 2249-555X

SUBCORTICAL BAND HETEROTOPIA(SBH) - A RARE CAUSE OF REFRACTORY EPILEPSY

KEYWORDS: Subcortical band heterotopia,refractory epilepsy,anti-epileptic drugs,electroencephalogram

Dr.S.LakshmiBai
Associate Professor of Medicine,Government General Hospital, Kurnool

Dr.VasanthaRao
Assistant Professor of Medicine, Government General Hospital, Kurnool

DR.TELIKI VINIL
Jr resident, Government General hospital ,Kurnool

Dr.Narasaram
Neurologist, Kurnool

ABSTRACT *We presnet a case of 16 years old girl with 10 years history of uncontrolled multiple generalised seizures inspite of adequate Anti-epileptic drugs(AED'S).Electroencephalogram(EEG) showed generalised discharges with relatively normal background activity.Magnetic resonanace imaging showed Lissencephaly type 1 with SBH .*

Introduction:
Band heterotopia also known as double cortex syndrome is a form of diffuse grey matter heterotopia.It is best thought as being on the lissencephaly type 1.Subcortical band heterotopia spectrum,as genetic defects are in many cases the same and results from arrested migration of neuron ,such that they form a grey matter layer within the centrum semiovale (or)subcortical white matter.

The majority of patients with double cortex syndrome are female onaccount of genetic abnormality often being located on x chromosome.seizures and delayed development are the most common presentation,usually evident in the first decade.The majority of band heterotopia syndrome cases have noebwwn attributed to an abnormality of hte DCX gene (also known as XLIS gene) located on the long arm of chromosome X.MRI is the imaging modality of choice[6].Positron emission tomography reveals metabolic abnormalities.

Case report:
A 17 year girl presented with history of recurrent unprovoked seizures since age of 5 years.Patient had delayed milestones,history of frequent falls and recent visual impairment.On neurological examinatiocognitivrimpairmrnt present with spasticity of all four limbs,bilateralplantarsext ensors.EEG showed generalised sharp and slow waves.MRI brain showed a band of grey matter located deep to and roughly paralleling,the cortex and signal intensities is the same as normal cortex on all sequences[2].Patient was initially started on phenytoin 100mg od at night at age of 5 year later changed to divalproate and clobazam due to recurrent seizures.Laterleviteracetam 500mg bd was added. .

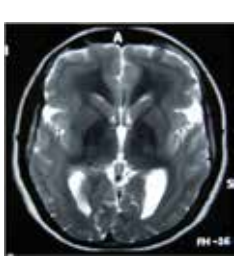

fig.01.MRI brain showing a band of grey matter located deep to and roughly paralleling,the cortex and signal intensities is the same as normal cortex on all sequences

Discussion:
Double cortex syndrome (or) diffuse cortical dysplasia is a rare cause of refractory epilepsy.As per clinical and epileptic spectrum in 10pts of double cortex syndrome published in neurology journal 1991,all patients are female,all have uncontrolled epilepsy,age of onset between 2 months to 11 years,all have mental retardation.The case described above have all the above features.Ideally patient should have positron emission tomography/computerised tomography(PET/CT) study[3].The pattern of metabolic abnormality in diffuse band heterotopia and useful for presurgical evaluation[4].the above patient may be a ideal candidate for palliative surgery with corpus callostomy.

Conclusion:
subcorticalband heterotopia are very cause of neurological and developmental disability and epileptic seizures. The seizures may arise at any age, but epilepsy will usually commence in childhood and is often resistant to anticonvulsant medications. Surgery may have a role in the treatment. Double cortical malformation syndromes with specific pathological, clinical, imaging, and genetic syndromes are being defined, and this knowledge has improved the clinician's ability to provide more accurate prognostic and genetic counseling to affected families, including prenatal testing for certain disorders. The study of these disorders has provided researchers with a unique opportunity to investigate the mechanisms of epileptogenesis.

Volume : 5 | Issue : 1 | Jan 2015 | ISSN - 2249-555X

REFERENCE

• 1)Madhavitripathi,'double cortex'sign on FDG-PET/CT in diffuse band heterotopia,Indian journal.med.2013oct. | • 2) BarkovichAJ,jacksonDE,Jr. BoyerRS.Bandheterotopia,A newly recognised neuronal migration anamoly.Radiology 1989. | • 3) Fong J,WuG,WyllieE,GuptaA. interictallhypermetabolic subcortical band on brain FDG-PET in double cortin mutation.neurology.2008. | • 4)FalconerJ.WadaJA,martinW.LID.PET,CT and MRI imaging of neuronal migration anamolies in epileptic patients.canJawnol sci.1990;1. | • 5) DevolderAG,GodisseuxJF,michelCJ,MaloteauxTm,BolAC,GrandinCB,et al.Brain glucose utilization in band heterotopia:synaptic activity of "double cortex"pediatr neurol.1994; . | • 6) Franicgallard,"double cortex syndrome" radiopedia. org MRI image. |