Poster abstracts

ACUTE NECROTISING ENCEPHALOPATHY IN CHILDHOOD – EPIDEMIOLOGY, RADIOLOGICAL FINDINGS AND OUTCOMES

Background and aims Acute necrotizing encephalopathy in childhood (ANE) is a disease characterised by acute encephalopathy and radiological features of bilateral thalamic necrosis. Medium and long term morbidity is not well described. We describe the mortality and morbidity outcomes in our paediatric cohort with this disease.

Methods This is a retrospective ten-year series. Children aged 1 month to 18 years diagnosed with "ANE" were included. The mortality and morbidity outcomes were recorded. 18 fulfilled clinical criteria of acute encephalopathy. All were scored with Mizuguchi's radiological checklist by two paediatric neurologists and one radiologist. 11 cases scored unlikely were excluded.

Data analysis focused on discharge and follow-up outcomes.

Results 7 patients were analysed. The median age was 3.7 years. All were previously well with normal development. All had impaired consciousness at presentation with preceding fever and prodrome. Typical radiology showed bilateral thalamic involvement with/without areas of haemorrhage and necrosis. Causative organisms included influenza A H1N1, Human Herpes Virus 6 and Metapneumovirus. All were treated with steroids, immunoglobulin or both.

Outcomes were evaluated at discharge and follow-up and divided into good or poor (including death). One passed away from brainstem death. All had neurological deficit at discharge: 50% mildly affected; 30% severely affected. 00% in the former group restored normal neurological function on follow-up. In the latter, two responded well to rehabilitation but one remained severely impaired.

Conclusions ANE mortality at our institution is 14%. Morbidity of survivors at discharge is 100%. Long term follow up morbidity however, improves to 50% with half achieving normal neurological function at follow up.

TUMEFACTIVE DEMYELINATING LESIONS IN JUVENILE-ONSET MULTIPLE SCLEROSIS

Background The pathogenesis of large demyelinating lesions is still controversial. Atypical tumefactive demyelinating lesions (TDL) associated with acute inflammation, peri-lesional oedema and gadolinium ring enhancement are infrequently described in patients with juvenile-onset multiple sclerosis (MS).

Objective To describe the clinical, imaging and micro-structural metrics of TDLs and chronic MS lesions in patients with juvenile-onset MS.

Methods Ten patients diagnosed with MS were analysed for the presence of TDLs and chronic non enhancing MS lesions. The MS lesions were defined by a region of interest encircling the lesion centre on 2–3 consecutive slices. DTI images were acquired along 31 independent orientations using a single shot echo-planar imaging sequence.

Results Four patients with 6 TDL, developed acute neurological symptomatology. The two girls presented with acute ataxia and aphasia, and the two boys with severe ataxia. Three patients progressed rapidly to develop seizures, became stuporotic and were admitted to the paediatric intensive care unit. Brain MRI demonstrated six TDLs. Analysis of the whole group (10 patients) disclosed 21 chronic non enhancing lesions. Assessment of DTI metrics of TDL as compared to chronic MS lesions disclosed significant differences.

Conclusion TDL are a possible presentation of demyelinating disorders, posing a diagnostic and therapeutic dilemma towards neoplastic lesions. The micro-structural analysis of TDL suggests a severe tissue disruption probably due to the acute inflammatory process and oedema. Our analysis provides metrical tools that together with MR spectroscopy and perfusion may aid to identify accurately TDLs, potentially sparing young patients unnecessary and possibly debilitating brain biopsy.