Language in a Brainstorm

Language and Epilepsy
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Language and epilepsy

- Case: A boy who lost language
- Acquired Epileptic Aphasia
- How are language impairment and epilepsy related?
- Language in other epilepsy syndromes
A boy who stopped talking

- 9 year old boy
- Three week history of
  - Complete loss of language (aphasia)
    - Severe problems with comprehension (receptive aphasia) and speech (expressive aphasia)
  - Major behavioural disturbance
- Epileptic seizures
This had happened before ...

- Occurred during a 3 month holiday in Chile
- Lost almost all expressive language
  - Only phrases “I hate you” and “I’ll kill you”
- Aggressive and hyperactive
- Three epileptic (tonic-clonic) seizures whilst in Chile
- Speech and behaviour improved two weeks before return to Australia
Second episode of aphasia

- Three months later ...
- Over a week he lost all spoken language
  - Initially unclear speech
  - Lost all comprehension of speech
  - Completely mute
  - Distressed by normal sounds
- Aggressive and hyperactive
- Neurological examination normal
- Brain MRI normal
Drawing during aphasia

Able to match written words to pictures – sometimes inaccurately
Reading during aphasia

- Able to read with profound receptive aphasia
  - Although some impairments of higher level language
- Implies
  - Dysfunction of verbal decoding (superior temporal gyrus)
  - Relative preservation of occipital (visual cortex) connections with Wernicke’s area (posterior perisylvian)
EEGs and epilepsy

- An EEG measures the brain's electrical activity
  - Records scalp electrical activity coming from the brain beneath the electrode
- The summated electrical fields produced by millions of nerve cells
- Spike
  - The simultaneous discharge of millions of neurones
  - Often followed by a slow wave
  - Focal (one region of the brain)
  - Generalised (many brain regions)
Diagnosis

- Acquired Epileptic Aphasial (Landau-Kleffner Syndrome)
Landau-Kleffner Syndrome

- “Syndrome of Acquired Aphasia with Convulsive Disorder in Children” *Neurology* 1957
- Five children referred to the Central Institute for the Deaf, St Louis Missouri (now > 200 children)
  - Not deaf
  - Previously normal language
  - New fluctuating aphasia
  - Epilepsy
  - Otherwise neurologically normal
Acquired Epileptic Aphasia (AEA): Clinical features

- **Aphasia**
  - Often a receptive aphasia at onset
  - Later children may appear mute
  - **Verbal auditory agnosia** a problem with decoding spoken language
    - Occasionally children have problems recognising common sounds (auditory agnosia)
    - May learn to read, write and use sign language whilst aphasic
Acquired Epileptic Aphasia: Clinical features

- **Seizures**
  - Range of different seizure types (oro-facial, absence, GTC)
  - 20% do not have seizures

- **Behaviour disturbance**
  - Aggression, irritability, attention deficit
  - Lack of socialisation, autistic features

- **Prognosis:** seizures and aphasia usually improve in early adolescence

- **Long term follow up:** most have difficulties with receptive language
  - Varying severity (mild to profound)
  - Poor phonological Short-Term Memory
Acquired Epileptic Aphasia: EEG findings

- **EEG may be normal when awake**
  - Focal temporal or parietal spike or spike and wave discharges

- **EEG in sleep**
  - Almost continuous epileptic activity (Electrical Status Epilepticus of Sleep)
  - Often with a temporal predominance
  - Most children show resolution of ESES prior to language recovery
What causes Acquired Epileptic Aphasia?

- Brain structure is usually normal
  - Rarely focal temporal lobe lesions
- Brain histology
  - Normal (biopsies uncommon)
  - No evidence of brain inflammation/encephalitis
Acquired Epileptic Aphasia: Theoretical Considerations

- AEA is likely to result from bilateral damage or dysfunction of the temporal lobes (*Rapin 1987*)
  - Children with unilateral damage to language cortex can re-locate language to the other hemisphere
  - This does not happen in AEA
Specialised Electrophysiology

- MEG Spikes generated within the Sylvian fissure
  - Spikes may be set off by sound (*Paetau R 1994, J. Clin Neuro*)

- Morrel *et al* (Brain) 1995: single cortical epileptic focus
  - Methohexital test/ Intra-carotid amylobarbitone
  - Electroencephalography: left sylvian discharges (10/14) and bilateral 4/14
Cerebral Glucose Metabolism (Positron Emission Tomography)

- PET studies (acutely)
  - Abnormal metabolism predominantly affecting the temporal lobe
  - Superior and middle temporal gyri (both right and left)
  - Peri-sylvian cortex (decreased)
  - Macquet et al Brain 1995
Theories regarding the cause of Epileptic Aphasia

- **Pathogenesis**
  - 1. Language regression is due to ESES
    - “Persistent convulsive disturbance in brain tissue largely concerned with linguistic communication results in functional ablation of these areas.” *Landau & Kleffner 1957*
  - Transient damage/Inhibition
    - 2. The EEG abnormalities and seizures are epiphenomena secondary to an as yet unidentified cerebral pathology

- **Something inhibits transfer of language**
Treatment

- Standard anti-epileptic medication rarely effective
- High dose steroids frequently effective
  - Powerful anti-epileptic medication
  - Does this treat unrecognised brain inflammation?
  - Relapse after withdrawal
Outcome after MST

Language improvement in 50%
- First words 12 weeks after surgery
- All children with language recovery showed normalisation of EEG
  - Morrel et al Brain 1995

- Caveats
  - These children were at an age when LKS may spontaneously improve
The association between epilepsy and language

- 1. Language impairment and epilepsy may result from focal brain damage
Language and epilepsy: Epileptic encephalopathy

- 2. Functional language impairment during an epileptic discharge
- 3. Functional impairment after a prolonged epileptic discharge
  - Focal weakness after a motor fit
    - Todd’s paresis
  - Language difficulties after temporal lobe seizures
  - Impairment can result from
    - Transient metabolic dysfunction/damage
    - Active cerebral inhibition
Language impairment and epilepsy: A common cause?

- 4. Language impairment may not be due to focal disease
- Language requires
  - Rapid processing
  - Good working memory
  - Attention
- Biological factors that lead to epilepsy may impair these processes
  - Eg Ion channel disorders
Language and epilepsy: Topiramate

- 5. Treatment with topiramate can be associated with language regression
- Word finding difficulties
  - Normal controls show significant language deficits when given topiramate
    - Meador et al Neurology 2005;64:2108-2114
  - Case reports of reversible language regression associated with topiramate
    - Vader et al Neurology 2004;62:299-300
Language in Temporal Lobe and Generalised Epilepsy

- Language impairment in Complex Partial Seizures (CPS)
  - Caplan et al Epilepsia 2004
  - Spoken Language Quotient
    - CPS 90 ± 18.5, controls 104 ± 14.5 p = 0.0001

- Evidence about language impairment in generalised epilepsy is contradictory
Language and generalised epilepsy

- Contradictory information
  - Evidence of abnormal Auditory Event Related Potentials to phonetic and semantic stimuli
  - Henkin et al Epilepsia 2003

- Relative preservation of language despite cognitive impairment in children with childhood absence epilepsy
  - Pataria et al Neurology 2004
Is there an increase in epileptiform activity in children with DLI?
- 50% of children with language impairment found to have epileptic activity on overnight EEG 
  
(Picard et al  Dev Med Child Neuro 1998)

- No history of seizures
- Ascertainment uncertain

Should we treat this activity?
Conclusions

1. Epileptic discharges can produce an acquired language impairment (Acquired Epileptic Aphasia)
   - Verbal auditory agnosia (word deafness)
   - Not always associated with seizures

2. Language impairments are common in Temporal Lobe Epilepsy

3. There is little information about the frequency of language disorders in generalised epilepsy

4. There is probably an increased incidence of epileptic activity in children with Developmental Language Impairment
   - But the significance is unclear
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When to suspect AEA

1. Language regression and seizures +/- behavioural disturbance
2. Verbal auditory agnosia
   - Severe receptive difficulties
   - Otherwise good non-verbal language skills
3. Fluctuating aphasia
Who should undergo language assessment

- 1. Children with clear language delays and epilepsy
- 2. Children with temporal lobe epilepsy
- 3. Children with epilepsy and poor school performance (esp. reading) and difficulty with socialisation