Landau-Kleffner Syndrome

Synonyms: acquired epileptic aphasia

Landau-Kleffner syndrome is an acquired aphasia secondary to an epileptic disturbance affecting a cortical area involved in verbal processing.[1] Affected children who have developed age-appropriate speech then experience language regression with verbal auditory agnosia, abnormal epileptiform activity, behavioural disturbances, and sometimes overt seizures.[2] Landau and Kleffner initially described acquired epileptic aphasia in 1957.[3] Landau-Kleffner syndrome has three features:[4]

- An acquired receptive aphasia.
- Temporoparietal spike-wave discharges in the awake state.
- Frequent generalised spike-wave discharges in sleep (electrical status epilepticus in sleep (ESES)).

Epidemiology

- The syndrome has been thought to be very rare but, with improved diagnostic techniques, the known prevalence may increase significantly.
- Currently, over 200 cases have been described in world literature.
- There is a slight increase in incidence in boys.
- Most cases do not have a well-defined cause. A few cases appear to have been secondary to low-grade brain tumours, closed head injury, neurocysticercosis and demyelinating disease.

Presentation

- Onset of aphasia is usually between the ages of 3 and 8 years.
- The syndrome is typically characterised by an abrupt or gradual loss of language ability and inattentiveness to sound (auditory agnosia).
- Receptive language is often severely impaired.
- Reading and writing may be preserved.
- The child may be completely mute or have severe expressive speech problems.
- Seizures occur in most cases and usually present between the ages of 4 and 10 years.
- Seizures may be partial, generalised tonic-clonic, absence or myoclonic.
- Many affected children have behavioural disturbances, including hyperactivity and decreased attention span, aggression and attacks of rage.

Differential diagnosis

- This is very broad and includes other causes of epilepsy, autism, hyperactivity and reduced attention span, aggressive (eg rage attacks), oppositional, or psychotic behaviour and mental retardation. Some patients may appear deaf, autistic or psychotic, or have some features suggesting attention deficit hyperactivity disorder (ADHD).
- Head injury, brain neoplasms (eg low-grade astrocytoma), and cerebrovascular thromboembolism and neurocysticercosis may all be associated with acquired aphasia, an epileptiform electroencephalogram (EEG) and seizures.
- Neurodegenerative disorders, especially adrenoleukodystrophy, and acute disseminated encephalomyelitis may also produce a similar presentation.

Investigations

- MRI is essential to rule out cerebrovascular thromboembolism, brain tumours, demyelination, neurodegenerative disease and central nervous system infections.
- Fluorodeoxyglucose positron emission tomography (FDG-PET) imaging reveals decreased metabolism in one or both temporal lobes. Single-photon emission computed tomography (SPECT) of the brain demonstrates decreased perfusion of the left temporal lobe.
- EEG: abnormalities are present in this syndrome but no consensus exists about what constitutes typical abnormalities.
- Brainstem auditory evoked potentials and hearing tests.

Management
Patients have special educational needs and require speech therapy. Speech therapy, including sign language, and a number of classroom and behavioural interventions, is beneficial. Psychotherapy may be indicated. A ketogenic diet has been recommended but experience is very limited.

Drugs

- Anticonvulsant medications have variable success.
- As initial therapy, valproic acid or diazepam is often used. Subsequently, other anti-epileptic drugs, corticosteroids, or intravenous immunoglobulin (IVG) therapy are often used.
- Various corticosteroid regimens including oral prednisone, high doses of intravenous pulse corticosteroids, and adrenocorticotrophic hormone (ACTH) have been reported to be effective.

Surgical

Multiple subpial transection (MST) has been used:

- The cortex is sliced in parallel lines in the midtemporal gyrus and perisylvian area in order to prevent the spread of the epileptiform activity without causing cortical dysfunction.
- This treatment is reserved for patients who have not responded to multiple medical therapies, but has been followed in selected cases by a marked improvement in language skills and behaviour.
- There is currently no accepted consensus about suitable candidates for this procedure or evidence for its effectiveness.

Prognosis

The long-term follow-up shows that epilepsy and EEG abnormalities do not always disappear. Language disturbances tend to persist in most patients. Patients tend to have an overall poor quality of life, mostly due to language difficulties.

- The overall long-term prognosis for language development is poor. Between 25% and 50% maintain language function at a level good enough to hold a job and have a normal social life.
- Patients with an onset of language regression before the age of 5 years may have a worse prognosis and symptoms persisting for more than one year are predictive of poor language recovery.
- Fluctuations in clinical severity and EEG changes are not unusual and short-term remissions may occur.

Further reading & references

- Landau-Kleffner Syndrome, Online Mendelian Inheritance in Man (OMIM)
  3. Neiman ES et al; Landau-Kleffner Syndrome, eMedicine, Jul 2010
  4. Landau-Kleffner Syndrome, Epilepsy in action

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