

CASE REPORT

Landau—Kleffner syndrome: a case of a dissociation between spoken and written language

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A case of a girl affected by Landau—Kleffner syndrome (acquired aphasia with convulsive disorder) is reported. The girl showed epileptic seizures and EEG abnormalities. At the age of 7 years 10 months they were followed by onset of aphasic symptoms accompanied with behavioural disturbances. By the age of 9 years she developed a severe verbal auditory agnosia and loss of spoken language expression, but was able to use reading and writing to communicate. This unusual dissociation suggests that spoken or written language can be affected selectively. The girl was followed up to the age of 14 years 6 months and her language recovered well by this age. (Fig. 2, Ref. 42.)

Key words: Landau—Kleffner syndrome, acquired childhood aphasia, verbal auditory agnosia, epilepsy.

In 1957 Landau and Kleffner described a syndrome of acquired aphasia with convulsive disorder in children (1). From then until the early 1990s, nearly 200 cases were reported (2, 3). The prevalence of Landau—Kleffner syndrome (LKS) has not yet been ascertained (4). The original description of LKS by Landau and Kleffner (1) can still be considered essential with regard to the clinical picture (3). LKS is characterized by a gradual or rapid onset of aphasia in a child with previously normal language development, and is associated with clinical seizures and paroxysmal EEG discharges (5). Behavioural disorders are frequently associated with LKS (6, 7).

The pathogenesis of the syndrome remains unclear (3). There is no evidence of structural lesions as usually shown by brain CT or MRI scans (8, 9, 10). Magnetoencephalographic evaluations showed primary intrasylvian or perisylvian spikes (11, 12). Recently, the relationship between LKS and ESES or CSWS have been discussed. The data support the concept that ESES syndrome may also include acquired language defects such as LKS (13). Functional brain imaging studies (PET) have demonstrated unilateral and bilateral metabolic abnormalities in the temporal cortex (10, 14, 15) and an abnormal unilateral perfusion in the temporal or temporoparietal regions (SPECT) (9, 16).

Various findings in LKS have been reported suggesting a possible association with the underlying pathology of the disorder: left temporal lobe astrocytoma (17), cerebral arteritis (18), inflammatory demyelinating disease (19), autoimmune reactions (20, 21).

No standardized treatment regimen has been developed (4). Besides the variable effects of treatment with different antiepileptic drugs, some benefits from corticosteroid treatment have been reported (10, 20), however, the available evidence is inconclusive (9). In the past decade, several studies reported improvements, especially in language recovery, following the use of a surgical technique — multiple subpial transection (22, 23, 24).

The literature reveals a great variability of symptoms in epileptic and behavioural disturbances, as well as in aphasia (4). The most frequent language disturbance is verbal auditory agnosia, which may be so profound that it can be mistaken for a loss of hearing (25). The presence of a number of deviant lingu-

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istic features has led to the conclusion that speech and language in LKS are deviant rather than immature (4, 26). Various findings have been reported concerning the written language in LKS cases (27, 28, 29).

The purpose of this study is to present a case of a child affected by the Landau—Kleffner syndrome in whom a complete dissociation of spoken and written language, in both comprehension and production had been observed for several months.

Case report

The patient, a girl, the younger child of a Hungarian speaking family, was born after an uneventful pregnancy and delivery. Birth weight was 3750 g. There was no family history of convulsive or communication disorders. The girl's psychomotor, speech and language development were reported as normal. Her medical history was unremarkable until the age of 4 years when she had a fainting fit. It was diagnosed as a collapse and she was not treated with any medication. At the age of 5 years she collapsed again and showed clonic seizures. The diagnosis of epilepsy was made and she was treated with antiepileptic drugs in the town of her residence. At the age of 6 years she was enrolled in a regular school. She enjoyed attending school and her first grade school results were very good. At the age of 7 years and 6 months, after awakening in the morning, she complained

of a headache, stomach-ache and showed a generalized clonic seizure. The waking EEG (Fig. 1) showed pathological epileptic activity. Four months later, another episode of clonic seizures occurred. This episode was followed by the onset of language deficits. Her second grade teacher noted difficulties in speech comprehension and recommended clinical examination. She was admitted to the Department of Neurology of the University Children's Hospital in Bratislava. Her expressive language started to deteriorate as well. The girl had word-finding difficulties and her speech was gradually becoming less intelligible. At this period of time, epileptic seizures of different types occurred more often, atypical absences appeared with the frequency up to 15 per day and she showed bilateral generalized seizures during sleep. At that time (8;2 years) the waking EEG (Fig. 2) showed bilateral synchronous discharges. She was placed on a combination of hydantoine and valproate. She stopped attending school because of her comprehension deficits and lack of co-operation. Variable fluctuations were apparent in the following year. She had several epileptic seizures, developed verbal auditory agnosia and behaviour disturbances with ups and downs were observed. Sometimes she was not interested in social contact and demonstrated negativism. She expressed herself by gestures and by finger pointing, and sometimes was able to utter single words. She developed some spontaneous gestural communication with her mother. She said her last intelligible words at the

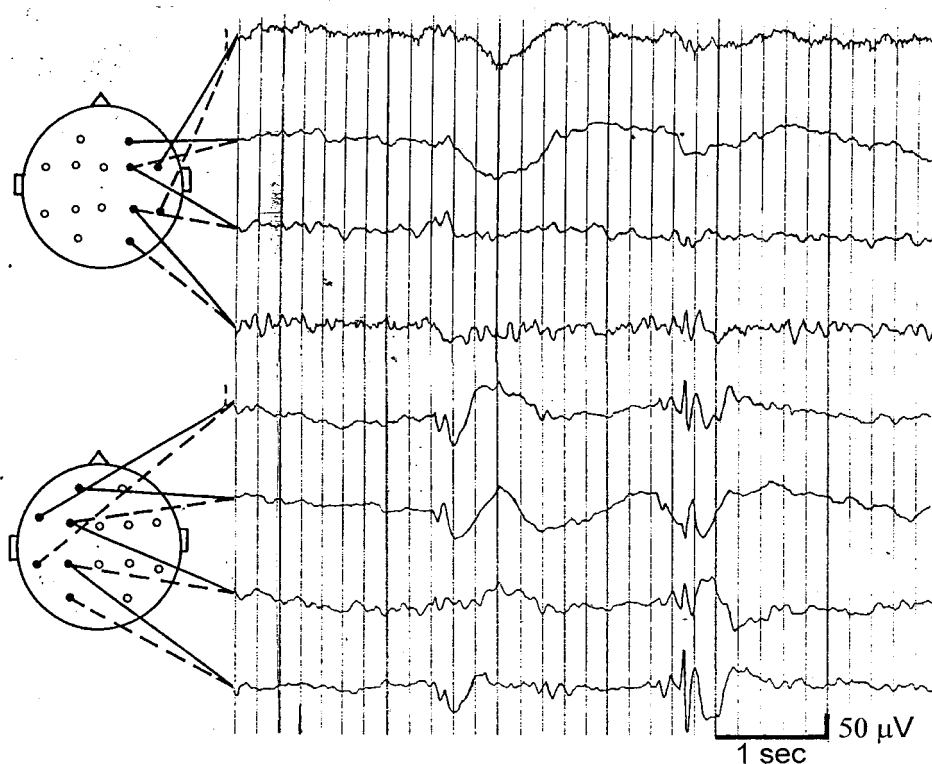


Fig. 1. Waking EEG in the initial phases of the disorder (prior to the language disorder) showing left-sided spike-and-slow-wave discharges over temporal and parieto-occipital regions and right-sided sharp-and-slow-wave over occipital region.

age of 9;1 years. Since then behaviour regression has been observed. Verbal, as well as nonverbal, contact with her was almost impossible. She ceased to dress and feed herself and temporarily she lost sphincter control. She was hyperactive, used to take different objects and toys, smell them and throw them away. She avoided eye contact. Sometimes she was irritable, aggressive, or depressive. Hydantoine was stopped, carbamazepine was added to valproate and a short therapeutic trial with corticosteroids was administered. She became more co-operative and continued her education by learning at home. Her mother, a kindergarden teacher, taught her individually at home under the supervision of the school teacher. They used written language to communicate. At the age of 9;6 years the girl was placed on a high dose of corticosteroids (35 mg/day). She responded well to the therapy and had no further seizures. Her behaviour gradually began to get better. Severe verbal auditory agnosia persisted for six months after the initiation of corticosteroid therapy. At the age of 10;1 years she gradually started to recover her spoken language communication. Her auditory agnosia was becoming less severe, two months later she started to speak again and stopped using writing and reading to communicate. Her mother reported that during the period of the complete verbal auditory agnosia the girl was able twice, immediately after awakening to communicate with her orally. However, after one or two minutes, her oral expression ability disappeared.

At the age of 10;9 years she returned to the regular school and joined the fourth grade class. Her academic achievement

was good with the exception of writing to dictation in both languages, Hungarian and especially in Slovak as her second language. At the age of 10;10 years her waking EEG was back to normal. She continued to take corticosteroids until the age of 11;3 years, took carbamazepine and valproate until the age of 11;8 years and was on carbamazepine until the age of 14;2 years. She has been seizure-free since she started high-dose corticosteroid therapy.

In 2001, at the age of 19;6 years, she graduated from a secondary school of civil engineering.

Neurological examination

The following examinations were performed during her repeated hospitalizations at the Department of Neurology of the University Children's Hospital in Bratislava. Neurological examinations (except for hyperreflexia), oral praxia, cerebrospinal fluid, brain CT scan, ophthalmic background were normal. Sleep EEG recordings are not available. The standard waking EEG recordings during the period of aphasia were abnormal showing epileptic paroxysmal activity.

During the neurological examination at the age of 9;7 years, the following behaviour was observed. It was impossible to communicate with the girl by means of spoken language. She did not speak spontaneously, nor understood or responded to spoken commands. She did not respond to her own name, but responded to

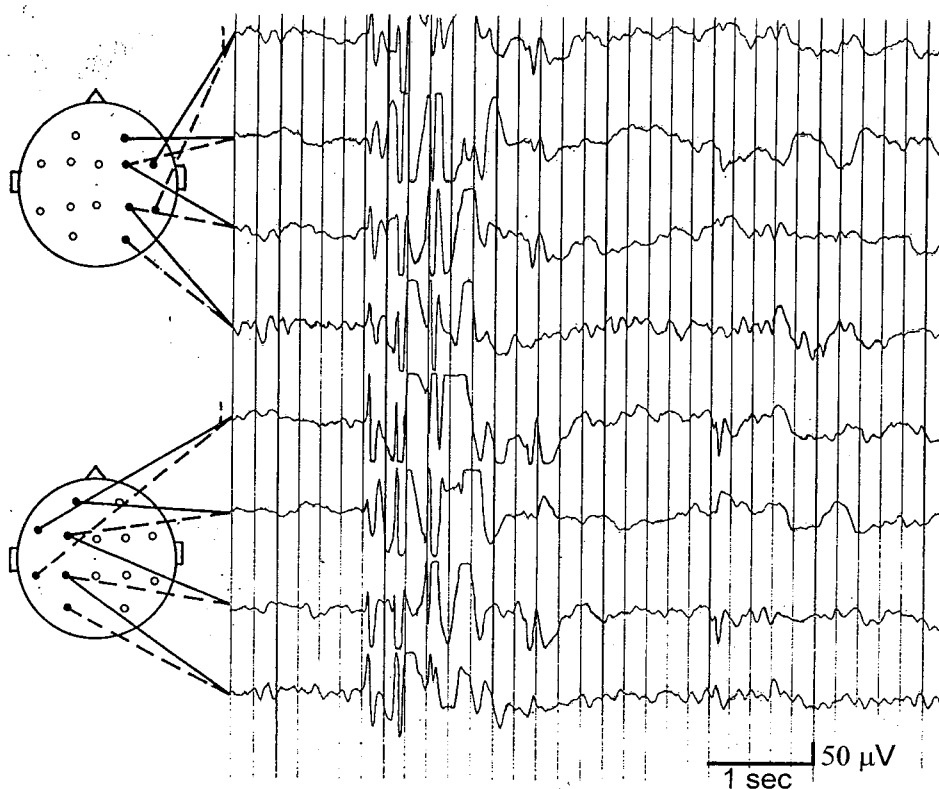


Fig. 2. Waking EEG during the phase of aphasia showing bilateral spike-and-slow-wave discharges with higher left-sided amplitude.

noises out of her sight. She walked around the room, picked up some toys and objects, manipulated objects and toys in a logical manner, her behaviour was characterized by hyperactivity. She communicated with her mother by gestures and by written language. She could copy a written or printed text correctly, written naming of different objects was correct, she used gestures or writing to express her needs. When asked to read aloud a paragraph from her school book she produced unintelligible, unarticulated sounds. She did written calculations — addition, subtraction, multiplication, division, at the second grade level. She enjoyed drawing and putting together jigsaw puzzles.

Neuropsychological examination

During the period of her language recovery, several psychological examinations have been done. At the age of 10;3 years the examination revealed consistent right-handedness as observed in a series of hand preference tasks. The girl's performance on the visual-motor Bender-Gestalt test (30) was at her age level (the 6th sten). As for spoken language comprehension and speech perception, she could understand short and simple commands. She was administered a dichotic listening task using a stop-consonant-vowel nonsense syllables free recall paradigm. She reported the syllable BA to all pairs of syllables presented under dichotic listening condition, but was able to repeat syllables uttered by the examiner. As to expressive language, she did not speak spontaneously. When she was asked to orally name and describe different pictures in a book, on some occasions it was difficult to determine whether she produced jargon or specific attempts to pronounce a word. Sometimes she omitted a syllable or added vowels or syllables within consonant blends. She substituted (l) for (r) and misarticulated sibilants. She was presented pictures of different objects, activities and everyday-life situations and asked to match them to cards with printed names of the pictures (31). She was able to match the pictures to their names almost perfectly. She also performed well in matching pictures of objects to the cards with printed names of different categories (e.g. body parts, vegetables, animals, means of transport). As to written language, she could copy a written or printed text and could write to simple words on dictation. She could read aloud Hungarian as well as Slovak texts, and a text composed of non-words. Apart from mentioned misarticulations she made a few letters or syllables order inversions. She was administered in a nonstandard way PDW — Czechoslovak standardization of the WISC (32). Her teacher translated into Hungarian for her and also translated the girl's responses. She failed to solve e.g. orally presented simple mathematical problems from the Arithmetic subtest, even though she was able to solve easily similar calculations in a written form. On the nonverbal Performance Scale she had an IQ of 96 with the lowest score in the Picture Arrangements subtest and best performances in Block Design and Object Assembly subtests.

At the age of 10 ;10 years she returned to her regular school. Her school achievement was good except for writing to dictation in both languages, in Hungarian and especially in Slovak as

her second language. She was administered (written presentation was used) seven items from an early (prepublication) version of TROG (Test for Reception of Grammar) as published in Bishop (26). The items were translated into Hungarian and she responded correctly to all items. She could read aloud fluently Hungarian as well as Slovak texts, her prosody was good. Her receptive and expressive spoken language was recovering well. On Raven Coloured Progressive Matrices she scored 33 out of 36 which corresponded to the 75th percentile and an IQ of 113 (33).

At the age of 11;3 years she stopped taking corticoids. At this time her performance in copying Rey-Osterrieth Complex Figure fell into the «superior» level and in memory production she performed within the «upper average» level of the 5-point scale of developmental norms based on the Slovak population (34). Her performance on dichotic listening showed no ear advantage. She reported correctly 35 % of stimuli from her right ear as well as from the left one. She did not report correctly any TA and KA syllables under dichotic listening. On the test Reading Sentences for Meaning, Pointing, from the Neurosensory Center Comprehensive Assessment of Aphasia (35) administered in a nonstandard way (translated into Hungarian) she performed at ceiling.

At the age of 14;6 years she had not been receiving any medication for 4 months. In a test of phonological discrimination, all her responses were correct. The dichotic listening revealed a REA with 60 % correctly reported syllables from the right ear and 26.7 % correctly reported syllables from the left one. Her articulation skills, as measured by a test of fast repetition of different syllables, were good. She could read aloud Hungarian, Slovak and a text composed of non-words correctly. Her school performance was good.

Discussion

The clinical picture of the reported case showed main characteristics of LKS. After a period of normal development, seizures were the first manifestation of the disorder and EEG abnormalities were recorded. Later the girl developed aphasia, and behavioural disturbances were observed as well.

A striking feature of the reported case was a complete dissociation of spoken and written language in understanding as well as in production. In the period of a severe verbal auditory agnosia and loss of ability to speak, the girl was able to use reading and writing to communicate — to express her needs by written language, to name objects writing, to copy written and printed texts. She was able to do written calculations as well. Her written language command enabled her to communicate with her mother and her teacher and to learn and continue her education by learning at home. This suggests that the neural circuits of spoken and written language functioned independently.

The variability in the manifestation of language disturbances in LKS is well known. There have been several case studies reporting disturbances of previously acquired written language in children with LKS (5, 9, 29, 36). But there are also cases with relatively preserved written language comprehension and/or pro-

duction and their use for communication (1, 25, 28, 37, 38). In our patient the preserved functional circuit was that of written language. A similar case to ours was reported by Landau and Kleffner (case 5) (1). The girl communicated well through reading and writing, but had also a limited oral expression. Also in her case, the language recovery was complete (36). There was another case of dissociation of spoken and written language in a boy with LKS reported by Denes et al (27). However, the boy started to learn reading and writing after the onset of aphasia. Based on the results of a detailed neurolinguistic analysis of the case, Denes et al (27) proposed that the process of acquisition of written language in their patient could be mediated through direct access to semantic representation of a word, bypassing phonological mediation. They suggested that a central impairment of phonological processing can explain the dissociation between spoken and written modalities.

Another remarkable feature in our patient was her ability to communicate orally for 1–2 minutes immediately after awakening, which her mother observed twice during the period of a complete loss of spoken language. Landau and Kleffner (1) postulated that aphasia may be the result of a functional ablation of primary language areas of the brain by persistent discharges in these regions. The episodes of reversibility of the ability to speak in our patient suggest that the pathological mechanism might have been a sort of functional ablation.

Recent studies of pathophysiological mechanisms involved in LKS point to the importance of the degree of functional integrity and that of increased vulnerability of language circuits in the immature brain (11, 22, 39). Differences in these factors might contribute to the variability of the clinical picture and of the outcome in LKS.

The reported case is one of those that have recovered well. The seizures were the first manifestation and the onset of language regression was observed at the age of 7;10 years. Three years after the onset of the first symptoms of aphasia the language recovered to such a degree that the girl could continue her education at a regular school. However, as revealed also by dichotic listening, even after the active period of LKS, various deficits may persist (40). No ear advantage with relatively low right ear performance on dichotic listening half a year after EEG normalization might have indicated a subtle speech perception deficit in our patient. The later improvement of the right ear performance, as recorded at the age of 14 ;6 years, might have reflected the recovery of speech perception.

Nevertheless, it is questionable to what extent the course was influenced by the corticosteroid therapy or by the age of onset of language regression. Landau (41) pointed out that there was no convincing evidence that corticosteroids or any other pharmaceutical agent contributed surely to clinical improvement. As to the age at onset of language regression, the older the child is at onset, the better the prognosis for language in LKS (42).

Future systematic follow-up studies with detailed analysis of language characteristics in correlation with EEG findings might shed more light on the relation among epilepsy, language disorder, and behaviour disturbances in LKS.

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