

Reversible aphasia in adolescence: A late-onset form of Landau Kleffner syndrome? Report of a single case

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An adolescent boy developed a long-lasting pattern of global aphasia, concomitant to focal (left temporal) EEG abnormalities; this was followed by complete recovery. Laboratory and neuroimaging studies were within normal limits. The possibility of a late-onset Landau Kleffner syndrome is discussed.

Keywords: Landau Kleffner syndrome – Epilepsy – Aphasia

INTRODUCTION

First described by Jackson (1894), aphasia is sometimes seen in the context of epilepsy, either as an ictal phenomenon or as a postictal language impairment. As an ictal manifestation, epileptic aphasia must be differentiated from the more frequent epileptic speech arrest. The relationship of epileptic speech arrest to aphasia is uncertain since speech arrest can be the clinical expression of a seizure originating in cerebral areas which are not included in the classic language areas (Penfield and Roberts, 1959). The landmarks of ictal aphasia are the presence of clear cut aphasic signs, such as anomias, paraphasias and auditory comprehension disorders, concomitant to seizure activities on the EEG, in a patient whose level of alertness is normal (Rosebaum *et al.*, 1986). A recurrent, reversible pattern of Wernicke's aphasia concomitant to paroxysmal left posterior temporoparietal spike and wave activity was reported in a 62-year-old patient whose symptomatology was repeatedly misdiagnosed as psychotic outbreaks (Knight and Cooper, 1986). More recently, Kirshner *et al.* (1995) presented a case report of a 50-year-old patient who showed a pattern of fluent aphasia of several days duration, secondary to partial status epilepticus arising from the left temporobasal region.

In contrast to the low incidence of epileptic aphasia in adults, acquired aphasias with convulsive disorders in children is reported much more frequently. Since

the first report by Landau and Kleffner (1957), more than 160 cases have been reported between 1956 and 1990 (Paquier *et al.*, 1992). Although a cerebral arteritis (Pascual-Castroviejo *et al.*, 1992) or an inflammatory demyelinating disease (Perniola *et al.*, 1993) has been found in some cases, the aetiology is unknown in the majority of cases. The peak incidence is between ages three and five years, when language is already fully developed. According to the original description of the syndrome, the onset is progressive and differs from the pattern of childhood-acquired aphasia following focal lesions; while the latter is characterized primarily by expressive language disorders, the main symptom of Landau Kleffner Syndrome (LKS) is a deficit in auditory comprehension, often leading to a pattern of word or phonemic deafness (Denes *et al.*, 1986). The prognosis is poorer than in developmental aphasia following focal brain damage. Deonna *et al.* (1989) reported the follow-up of seven LKS patients in adulthood. A severe language disorder, involving both expressive language and auditory comprehension, was present in four of the patients; only one patient showed a complete recovery while the remaining patients were affected by dyslexia or severe language production disorders. A dramatic and persistent improvement of language, including total reacquisition of language after two years of mutism, was reported by Morrel *et al.* (1995) in a subset of LKS children. Epileptogenic

discharges from the perisylvian region of one hemisphere were eliminated surgically by multiple subpial intracortical sections of the epileptogenic cortex. Behavioural disturbances, with aggressive traits, are often described in patients with LKS. Nonverbal cognitive functions are normal.

With the increasing numbers of reported cases, the pattern of clinical and EEG abnormalities has been found to be very variable. Even the landmarks of the syndrome, aphasia and convulsive disorders, are not homogeneous among the reported cases. In some cases, language impairment is predominantly of the motor type. The onset of the language deficit can be gradual or abrupt and it can develop in normal children or in children who have a history of developmental language disorders (Marien *et al.*, 1993). Clinical seizures, most often of generalized motor type, occur in 80% of patients, while the remaining 20% do not develop seizures. Various EEG abnormalities may be found, from bilateral synchronous spikes and wave discharges to unilateral temporal spikes. The pattern of continuous spikes and wave discharge during slow sleep (electrical status epilepticus) in some LKS children appears to be a specific feature of the syndrome. It also appears to give an indication of the severity of the clinical course of LKS (Patry *et al.*, 1971; Tassinari *et al.*, 1985). Unfortunately, the available experimental evidence has been too limited to reach a definite conclusion concerning the specificity and prognostic value of this abnormality. The relationship between the severity of language impairment and clinical course, on the one hand, and frequency of seizures and EEG abnormalities, on the other, is still a matter of debate. The only correlation which appears to be consistent in LKS is that between age of onset and clinical course and recovery. In a meta-analysis of the published case reports, Bishop (1985) found a consistent relationship between age of onset of the syndrome and final outcome. More rapid and complete language recovery occurred in patients who were older at the time of onset of LKS.

This paper is a case report of an adolescent boy who, following a single epileptic seizure, developed a long-lasting aphasia. There was no evidence of structural brain damage. Complete recovery occurred gradually.

CASE REPORT

PD, an 18-year-old right handed boy, was born after an unremarkable pregnancy. His developmental milestones were normal; he walked between the ages of ten and 12 months and he used single words at one year. Further linguistic development was reported to be

normal by his parents. He started school at the age of six and his teacher remembered him as a boy with normal learning abilities, although he was better at written, rather than oral, assignments. He did not have speech therapy or special tutors. After five years of uneventful elementary school and three years of junior high school, he started attending a quite demanding technical high school, where he had failed twice, but he was reported to be doing well at the time of his admission. The patient was described by his relatives as a pleasant boy, fond of sports and member of a sailing team.

On the night of admission, PD went to bed early following a sports coaching session, claiming to be particularly tired and dizzy. A few minutes later, his parents heard a loud noise and found him to be unconscious and pale. Although he was lying on the floor and moving both upper limbs, no definite clonic jerks were reported. The patient was not incontinent. PD regained consciousness after several minutes but he could not remember anything of what happened. On admission to the Department of Neurology, University of Padova, the general physical and neurological examination of PD were normal, apart from some word-finding difficulties. Blood pressure was 160/90. Laboratory investigations showed only a slight increase in leukocyte count (12.9×10^9). Erythrocyte count and erythrocyte sedimentation rate were normal. Examination of the CSF was normal, indicating only a slight pleiocytosis (28 white cells, protein 26 mg, glucose 0.67 mol/L). Serum and CSF virological studies were normal.

Cranial CT, cerebral MRI and cerebral angiography were normal, as were the ECG and echocardiogram. The EEG showed the presence of theta activity, intermixed with spikes over the left temporal region (see Fig. 1).

CLINICAL COURSE

In the days following admission of PD language difficulties progressed to global aphasia with severe output (oral and written) and comprehension disorders. At the same time, slight emotional disturbances appeared, characterized by outbursts of crying and some aggressive traits, mostly directed towards his relatives. The patient was started on high doses of intravenous corticosteroids for three days; treatment was continued at decreasing doses for a further ten days. Phenobarbitone was given orally at doses of 100 mg. The language disturbances started to subside slowly after ten days and mood and behaviour returned to normal. Similarly, the EEG improved to a normal pattern. Prolonged EEG recordings, including sleep, were normal.

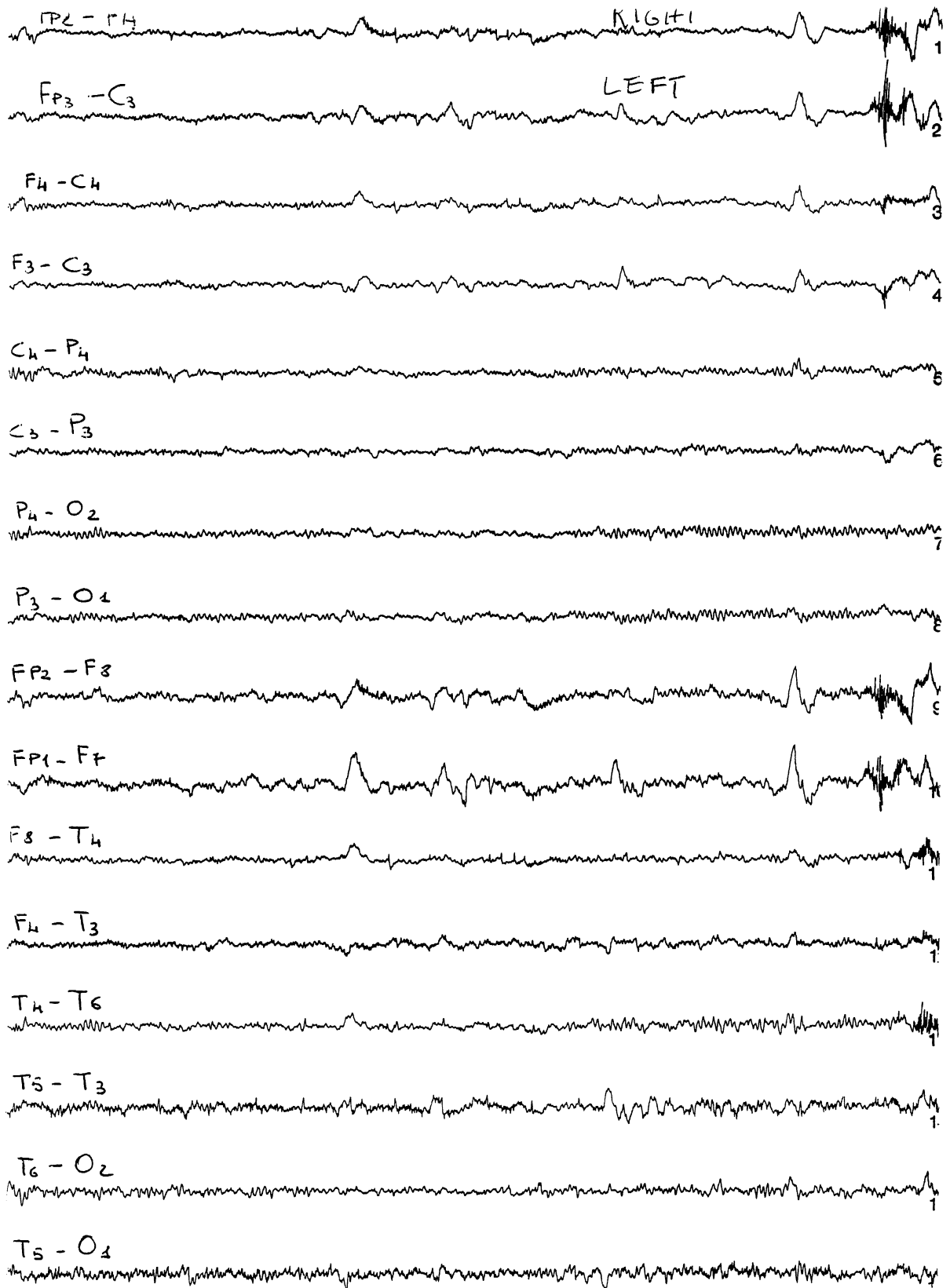


FIG. 1. EEG recording in the acute phase, showing theta and theta delta activity in the left temporal region.

The patient was discharged 20 days after admission; he was free of symptoms. On follow-up examination, neurological and language examination remained normal and he was progressing well at school. Six months after discharge, he had a grand mal seizure which was not accompanied by language disturbances. The EEG showed the presence of a left temporal epileptic focus.

LANGUAGE EXAMINATION

The first formal language examination took place five days after admission using the Italian version of the Aachener Aphasia Test (Luzzatti *et al.*, 1991). Spontaneous language was limited to a few syllables which the patient tended to repeat. In his efforts to communicate, he showed normal use of gestures and tried, without success, to communicate through writing. Repetition and reading aloud were similarly impaired. Auditory and written comprehension were impaired, with some semantic errors in the spoken word-picture matching test; (for example, he matched the word pitchfork with the picture of a rake). On the 50 item version of the Token test (De Renzi and Vignolo, 1962), his performance was grossly defective, with a score of 32 errors. His score for oral naming was zero; although his score for written naming was slightly better, it was still very impaired and contained many orthographic errors. His scores for written naming of compound words and written description were zero. Table I summarizes the relevant findings.

Oral calculation was poor and written calculation, although correct, was extremely slow. Drawing, on command and on copy, was correct. The patient showed some elements of oral apraxia (8/10 correct) while his performance on ideomotor and ideative apraxia tests was flawless. On the 48 item version of the Raven Progressive Matrices, he scored within normal limits (42/48 correct).

In the days following the first formal examination of language, PD's language began to improve. However, some disturbances in language remained, such as in generative naming tasks following semantic cueing. In this test, the patient is asked to produce as many words

as he knows which belong to the following categories: colours, animals, towns, fruits; the time limit is one minute for each category. PD's performance (5.25) was grossly defective (cut off point 19.54; Spinnler and Tognoni, 1987). Reading aloud was characterized by the presence of many phonemic paraphasias.

PD was retested 20 days after admission on the detailed language examination battery developed by Miceli *et al.* (1994) and his score was within normal limits. A psychological evaluation was essentially normal, failing to disclose any important emotional or affective problems. His parents denied the presence of previous emotional or behavioural problems. His previous failures at school were interpreted as being due to the wrong choice of school.

The patient was examined at regular intervals after discharge and his language, memory and calculation were found to be normal.

DISCUSSION

We have documented the case of an adolescent boy who developed, in the days following an epileptic seizure, a severe and progressive language impairment, progressing to global aphasia, followed by a gradual and complete disappearance of symptoms in about one month, resulting in complete recovery.

The linguistic deficit was not concomitant with elementary neurological deficits or with other signs of cognitive impairment, apart from mild calculation disturbances. The awareness, attention, orientation and non-verbal memory of the patient were normal, as was his use of gestures. In the acute phase only, mild emotional disturbances were present but a subsequent psychological report was normal. Previous history was unremarkable; in particular, no developmental language deficits were noted, apart from a higher achievement in written, rather than oral, assignments, suggesting the possibility of a subclinical phonemic output disorder.

The normal values obtained following elementary neurological examination, laboratory studies and neuro-imaging studies rule out the possibility that PD's aphasia could be the expression of a structural disorder of neoplastic inflammatory or vascular origin. From a functional point of view, PD's normal performance in attentional and nonverbal cognitive tasks rules out the hypothesis that the language impairment could be evidence of a more wide-spread cognitive impairment or of a global deficit in communicative abilities. His use of appropriate gestures to try and overcome the consequences of his linguistic impairment indicates that he did not develop a global deficit in his ability to communicate.

TABLE I: The performance of the patient on the Aachener Aphasia Test five days after admission

Test	Range	Score	Global severity	Aphasia severity
Token test (errors)	50-0	31	5	medium
Repetition	0-150	35	2	severe
Written language	0-90	16	3	severe
Naming	0-120	0	1	severe
Comprehension	0-120	61	3	severe

The only abnormal laboratory value noted was during the acute stage of PD's illness. The left temporal focus which was noted on the EEG disappeared following treatment with corticosteroids. This change in the EEG occurred in parallel with the improvement in the language deficit.

On the basis of the clinical course and laboratory studies, it does not seem unreasonable to interpret PD's symptomatology either as a case of aphasia secondary to spontaneous status epilepticus (Rosenbaum *et al.*, 1986; Kirshner *et al.*, 1995), or as a case of late-onset LKS. Given the lack of frequent ictal discharges and the young age of the patient (all reported cases of epileptic aphasia are in adults), it seems more probable that the symptomatology described relates to an adolescent variant of LKS.

To the best of our knowledge, this is the first reported case of LKS in an adolescent. However, this disorder may have occurred in other adolescents in the past. A sudden regression of language followed by complete recovery could be easily interpreted as a transient psychiatric disturbance rather than as a manifestation of a neurological disease, with the consequence of underestimating the incidence of LKS in adolescence (Deonna *et al.*, 1977; Bishop, 1985).

In conclusion, given the persisting lack of knowledge of the aetiology of LKS (Landau, 1992) and the lack of definite diagnostic criteria, the possibility of late-onset LKS should be considered in cases of acquired language impairment in adolescence.

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