Acquired childhood aphasia with convulsive disorder (Landau-Kleffner syndrome)

A case report

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Summary

The Landau-Kleffner syndrome is a rare form of acquired childhood aphasia associated with convulsive disorder. These children are typically misdiagnosed. A 15-year-old girl whose aphasia was not diagnosed until the age of 13 is described. Pitfalls in diagnosis are discussed, particularly with regard to interpretation of the results of audiometric testing. Attention is drawn to the existence of this rare form of aphasia and to the lack of appropriate educational facilities for aphasic children in general.

Acquired childhood aphasia with convulsive disorder, 1 although a rare form of language disorder in childhood, has become increasingly well recognised as a separate clinical entity since the first case was described in 1957 by Landau and Kleffner. 2 It is characterised by a number of features related to onset and course that overlap with a number of other childhood syndromes and thus presents a challenge to the diagnostician.

The hallmarks of the syndrome are normal development until the age of 3-5 years followed by a sudden or gradual loss of language skills with electro-encephalography (EEG) typically showing bilateral paroxysmal spike and wave discharges from the temporal lobes. Seizures and/or behavioural and psychomotor disturbances occur in about 70% of cases. 3-5 There is considerable variability in outcome for such children, 2,6,7 and there tends generally to be a poor correlation between the frequency of the seizures and the severity of the aphasia, and control of the epilepsy often has no effect on the aphasia.

The language characteristics of such children typically include an auditory comprehension disorder with a sudden or gradual loss of speech. 8,9 Behavioural characteristics may include attention deficit, hyperactivity disorder and sometimes a withdrawal from social situations. Audiologically, such children are often difficult to test and typically give the impression of being deaf.

The differential diagnosis includes severe deafness, mental retardation, autism, developmental language delay and other forms of acquired aphasia, e.g. those resulting from trauma, neoplasm and stroke and the temporary intermittent aphasia typically giving the impression of being deaf. 8,9 There is considerable variability in outcome for such children, 2,6,7 and there tends generally to be a poor correlation between the frequency of the seizures and the severity of the aphasia, and control of the epilepsy often has no effect on the aphasia.

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The differential diagnosis includes severe deafness, mental retardation, autism, developmental language delay and other forms of acquired aphasia, e.g. those resulting from trauma, neoplasm and stroke and the temporary intermittent aphasia associated with ongoing epilepsy. 10 The problems in differential diagnosis of such cases have important implications for management of the child as will be illustrated in the following case report, which underlines the need for team diagnosis and the danger of inappropriate labelling.

Case report

A 15-year-old right-handed girl from a Portuguese-speaking home presented as an unusual case of school refusal, after 10 years as a boarder at a school for the deaf.

The developmental history was uneventful until the age of 22 months when she began to have seizures (tonic-clonic and absent seizures). These decreased in frequency and occurrence over the next two years and anticonvulsant medication was eventually stopped at the age of 13 after a 2-year seizure-free interval.

Six months after the onset of epilepsy, she began to lose her speech and language abilities and appeared deaf. By the age of 3 years, she had lost all expressive language and no longer responded to any verbal stimuli.

The history of different assessments is a long and complex one and is summarised in Table 1. The patient was re-evaluated on 16 separate occasions over a period of some 12 years and the labels offered included: brain damage, deafness, autism, mental retardation and aphasia.

Repeated neurological examinations revealed no physical abnormality apart from apparent deafness. An EEG at the age of 7 years was reported to show a grossly abnormal petit mal variant tracing with 1-2 Hz spike complexes and frequent dominating bursts. More recent EEG tracings (at the age of 13 years and 14 years) revealed episodic sharp-tipped and slow-wave activity in the left frontotemporal areas (Fig. 1). At this stage computed tomography (CT) with temporal lobe views was normal. Repeated hearing tests over the years revealed a very variable picture with regard to hearing status. A hearing aid was inappropriately prescribed at 4 years of age and the patient was placed in a school for the deaf, in which the educational policy focuses on the development of oral linguistic skills.

Speech and language development remained very restricted and incompatible with the normal response of the deaf child. By the age of 12½ years the patient used no recognisable words, but had developed a simple gesture system together with some functional reading and writing. At the age of 13 years she was referred for a reassessment because she refused to wear a hearing aid and because a response to environmental sound was noted, and at this point brainstem evoked response (BSER) indicated normal hearing bilaterally.

Present test results

Medical psychiatric assessment

The parent interview revealed an understandably distressed mother bewildered by the variety of diagnoses that had been offered over the years. The child presented as dull, making poor eye contact and did not co-operate with the interviewer.
**Non-verbal IQ**

66

**Autism**

Seizure-free for 2 years; medication stopped

Normal hearing; referred for alternative placement (outside school for the deaf)

Mild loss with conductive component

WISC-R non-verbal IQ 96; profound aphasia; does not qualify for admission to school for brain-injured children; hearing aid discontinued

Left frontotemporal dysfunction

Non-verbal IQ 66

Neurology assessment

EEG

Psychological assessment

Bilateral hearing loss

Speech therapy

Hearing again

Paediatric

Neuropsychological assessment

Psychology; OT; speech, language and hearing

Neuropsychological; psychiatric; speech and language

TABLE I. ASSESSMENT HISTORY OF PATIENT WITH ACQUIRED CHILDHOOD APHASIA

<table>
<thead>
<tr>
<th>Date</th>
<th>Type of assessment</th>
<th>Findings and recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>1973</td>
<td>Medical</td>
<td>Epilepsy; anticonvulsants prescribed</td>
</tr>
<tr>
<td>1974</td>
<td>ERA (audiometric)</td>
<td>Severe deafness; hearing aid prescribed</td>
</tr>
<tr>
<td>1974</td>
<td>ERA</td>
<td>Normal hearing. Placement in school for the brain-injured recommended, but placed in a school for the deaf</td>
</tr>
<tr>
<td>1977</td>
<td>Hearing test</td>
<td>Severe bilateral deafness</td>
</tr>
<tr>
<td>1978</td>
<td>Speech therapy</td>
<td>Deafness</td>
</tr>
<tr>
<td>1978</td>
<td>Neurology assessment</td>
<td>No other neurology deficits</td>
</tr>
<tr>
<td>1979</td>
<td>EEG</td>
<td>'Child living in subconscious'</td>
</tr>
<tr>
<td>1980</td>
<td>Psychological (Snijders-Ooman)</td>
<td>Generally abnormal — petit mal</td>
</tr>
<tr>
<td>1982</td>
<td>Snijders-Ooman</td>
<td>Non-verbal IQ 70</td>
</tr>
<tr>
<td>1983</td>
<td>Medical (Portugal)</td>
<td>Seizure-free for 2 years; medication stopped</td>
</tr>
<tr>
<td>1984</td>
<td>Neurology</td>
<td>Normal hearing; referred for alternative placement (outside school for the deaf)</td>
</tr>
<tr>
<td>1984</td>
<td>Brainstem audiometry</td>
<td>Mild loss with conductive component</td>
</tr>
<tr>
<td>1984</td>
<td>Pure tone audiometry</td>
<td>WISC-R non-verbal IQ 96; profound aphasia; does not qualify for admission to school for brain-injured children; hearing aid discontinued</td>
</tr>
<tr>
<td>1985</td>
<td>EEG</td>
<td>Left frontotemporal dysfunction</td>
</tr>
<tr>
<td>1986</td>
<td>Neurology</td>
<td>Aphasia</td>
</tr>
<tr>
<td>1986</td>
<td>CT</td>
<td>NAD</td>
</tr>
<tr>
<td>1987</td>
<td>Hearing</td>
<td>Bilateral hearing loss</td>
</tr>
<tr>
<td>1987</td>
<td>Paediatric</td>
<td>? autism</td>
</tr>
<tr>
<td>1987</td>
<td>Neuropsychological; psychiatric; speech and language</td>
<td>Severe aphasia manifested in a profound verbal auditory agnosia and oral apraxia</td>
</tr>
</tbody>
</table>

However, she appeared to relate meaningfully with her mother using gesture to communicate with her.

The mother reported that the child enjoyed watching videos and listening to tapes of her favourite pop group and that she was able to participate spontaneously in shopping and cooking activities and showed good learning ability in everyday situations. Physical examination was normal.

**Neuropsychological assessment**

Because the patient presented with a dense oral apraxia and auditory agnosia, all neuropsychological testing was carried out at a visual level, the child responding in a written form or in terms of a drawing. The mother acted as an interpreter, conveying instructions through gesture. The results of the testing were: the non-verbal IQ appeared borderline-normal.

The patient showed basic preservation of constructive praxis (as tested in the copying of simple figures), memory function for both verbal and non-verbal forms (tested on the Webster Memory Scale), naming (through gesture and drawing), graphic skills, calculation and object recognition (through the visual mode). However, a marked degradation of performance was noted with increased task complexity and there were some signs of frontal lobe dysfunction (as seen in the copying of the Rey Complex Figure; Fig. 2).11

This constellation of signs is compatible with a left temporoparietal lesion that has left the occipitoparietal cortex intact. There is probably no involvement of axial structures, such as the hippocampus or mamillothalamic systems, although frontal lobe dysfunction seemed to be present. In view of the preserved arithmetic function and immediate memory, the lesion is relatively anterior probably involving the frontotemporal cortex.

Fig. 1. EEG at 13 years of age showing irregular and episodic 5-7 Hz sharp-tipped activity in the left frontocentrotemporal area.
Speech and language assessment

The speech and language assessment confirmed the diagnosis of severe aphasia and profound verbal auditory agnosia. There was very little co-operation during formal testing and the patient became antagonistic and left the situation. However, observation of her communication pattern with her mother revealed an apparent competence in relation to gesture. Her mother reported that she was able to communicate well with deaf friends using sign language. The patient's vocalisations during the assessment were limited to single vowel sounds and inappropriate laughing. Vocal quality was nasal and uninflected, sounding very similar to that of profoundly deaf children. The attention span and concentration for a chosen activity was felt to be appropriate. Hearing was normal in that she responded quickly and accurately to environmental sounds, thus confirming the previous objective test results.

Discussion

The diagnosis of the Landau-Kleffner syndrome was made on the basis of the neurological findings, diagnostic testing and the case history. The patient's profile of features excluded certain other syndromes. For example in other types of acquired childhood aphasia there is a clear, temporally related aetiology, recovery is fairly rapid, particularly with preschool children, and frequently one modality only is affected (usually the expressive side). This patient's relatively good interpersonal skills throughout childhood, not only with the mother but also with school peers, use of imaginative play, and the relative absence of stereotyped movements and rituals, together with the preservation of memory function makes a diagnosis of autism highly unlikely, although it is clear that the differentiation between autism and verbal auditory agnosia of this type is often very difficult. Deafness was mooted as a possible cause for a number of years and this label, in fact, influenced the patient's entire educational placement. The difficulty in diagnosing hearing loss is a particularly pronounced feature of this group. Such children respond inconsistently on normal behavioural audiometry and speech audiometry is clearly inappropriate. Cortical testing (e.g. using electro-encephalographic response audiometry) can often be confounded by neurological variables, including abnormal EEG patterns and medication. BSER seems the diagnostic procedure of choice in such cases, but it is important to remember that even if audiometric findings are strongly suggestive of hearing loss, a diagnosis of deafness should be reserved in children with a history of convulsions and aphasia, for such a label, as we have seen in the case of our subject has critical implications for placement.

Placement of such children in an educational setting presents a problem, particularly in a South African context. In this patient's case, however, the inappropriate placement in a deaf school can be viewed as fortuitous for she was able to learn some sign language, which (together with reading and writing) was the only channel available to her. The fact that learning of sign language was, because of local educational policy for the deaf, restricted to casual extra-classroom contact with deaf children, severely restricted both opportunity and prognosis for learning a formal symbol system. The formal teaching of an alternative communication system such as sign language, Blissymbols or Makaton vocabulary has proved useful in previously documented cases and may well have been effective with our patient had it been introduced at an early age.

The question of aetiology of the disorder for this patient is, as it is with most documented cases, very difficult to determine. Several theories have been postulated. In most cases, there is evidence of bilateral temporal lobe lesions. This accounts for the fact that such individuals seem unable to develop adequate compensatory mechanisms as in other childhood aphasias. The attention disorders and the nature of the EEG abnormality have been posited as evidence of a subcortical problem. Some suggestions have been offered of a subacute encephalitis caused, for example, by the herpes simplex virus or the possibility of autoimmune reaction. In the Moor House series of studies some of the children with this syndrome had a definite antecedent condition such as encephalitis or measles. Very few studies have, in fact, undertaken the necessary diagnostic testing (e.g. cerebrospinal fluid viral analysis) at the time of onset of seizures. This is an important implication for future research with such cases.

It is hoped that this case study has highlighted a relatively rare and unique childhood syndrome and the importance not only of team diagnosis but also team collaboration in deriving an appropriate diagnosis and subsequent treatment.

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REFERENCES

Chemotherapy for metastatic non-small-cell lung cancer

A case report

R. P. ABRATT, P. A. WILLCOX, E. BOLDING, R. H. HEWITSON

Summary

The role of chemotherapy for patients with metastatic non-small-cell lung carcinoma (NSCCL) is controversial. A patient with intrathoracic metastatic NSCCL, who was treated by moderate dose cisplatin combination chemotherapy and who remained clinically free of disease more than 5 years after presentation, is described. This treatment has not previously been reported. A trial of moderate-dose cisplatin combination chemotherapy in selected patients with good performance status seems justified.

Case report

A 60-year-old man presented to hospital in August 1984 with a 6-week history of a 'flu-like illness and marked weight loss. He was an ex-smoker. Chest radiography revealed bilateral pulmonary masses (Fig. 1). He was Eastern Co-operative Oncology Group (ECOG) performance status 1 and the biochemical profile was normal. Fibre-optic bronchoscopic results were normal and the patient underwent open biopsy, which revealed multiple masses in all lobes of the right lung.

Histological examination of a 3 x 2 cm wedge of lung tissue showed it to be infiltrated by several solid nodules of undifferentiated carcinoma composed of solid sheets and nests of large vesicular cells with prominent nucleoli and scanty cytoplasm. Numerous mitoses were noted, many of which were atypical and there was marked tumour necrosis. No erythrophagocytosis was present. Mucin staining was negative and immunoperoxidase stains for leucocyte common antigen and cytokeratin were also negative, although chorio-embryonic antigen showed focal positivity. The features were those of undifferentiated large-cell carcinoma of the lung.

The patient was treated with cisplatin 60 mg/m² for 1 day and etoposide (VP-P) 60 mg/m² on days 1-3, repeated every 3 weeks for a total of 6 cycles. There was complete response to chemotherapy (Fig. 2) associated with an improvement in functional specialization.

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