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Acquired aphasia without deafness in childhood – the Landau-Kleffner syndrome

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Abstract
A young boy presented with loss of speech and behaviour disturbance and was thought to be deaf. He was subsequently found to have the Landau-Kleffner syndrome (LKS), or acquired aphasia with epilepsy. Children with this disorder commonly present to an audiology or ENT clinic. Early recognition is important to initiate supportive, speech and educational care.

Key words: Aphasia, acquired; Epilepsy

Introduction

Acquired epileptic aphasia (verbal auditory agnosia with convulsive disorder or the Landau-Kleffner syndrome) is characterized by acquired receptive and expressive aphasia and epileptic seizures. This rare disorder presents during childhood, with a peak at four to seven years of age (Beaumanoir, 1985; Paquier *et al.*, 1992). The aphasia usually occurs over days, but may develop over weeks. Epileptic seizures present at the same time as the aphasia or develop subsequently and may be controlled with antiepileptic medication or corticosteroids and usually remit with time, in contrast to the comprehension and language difficulties which frequently persist into adult life (Deonna *et al.*, 1989). The pathogenesis is unknown. The diagnosis of Landau-Kleffner syndrome depends upon an awareness of the condition. Children are frequently referred initially to an audiology or otolaryngology clinic with presumed deafness as being responsible for the loss of comprehension or speech. The following case illustrates the characteristic presentation and course of a child with LKS and emphasizes the need to be aware of this condition.

Case report

This four and a half-year-old boy was seen by his general practitioner with a three-day history of almost total loss of speech, clumsiness and marked physical aggression towards other children and his school teachers. His behaviour continued to deteriorate and five weeks later the school doctor referred the child for a combined ENT/audiological assessment. Pure tone audiometry, performed with difficulty, revealed a 20 decibel conductive loss bilaterally.

Further electrophysiological assessment, including brain stem auditory evoked responses, was not possible because of the child's marked agitation; it was considered inappropriate to undertake these investigations under sedation or general anaesthesia. Examination of the ear, nose and throat, tympanometry and stapedal reflexes were all normal and the child was referred to a paediatric neurologist (R.E.A.). The patient's birth, perinatal period and early development (including speech and language) had been normal. The patient had not yet learned to read or write. There was no relevant family or past medical history, and no recent illness. General physical examination was normal. The child was restless and agitated, did not respond to simple questions or commands and neither talked nor vocalized in any way. Neurological examination was normal; the patient was right hand dominant. A diagnosis of probable Landau-Kleffner syndrome was made. A magnetic resonance imaging (MRI) head scan was normal and an awake electroencephalogram (EEG) showed a persistent high amplitude, right temporo-parietal spike and slow-wave discharge which became almost continuous and generalized during slow-wave sleep, consistent with a diagnosis of Landau-Kleffner syndrome (Beaumanoir, 1985; Paquier *et al.*, 1992). The patient was referred for urgent speech therapy and educational psychology assessments. Three months after presentation the patient developed partial and generalized epileptic seizures which were initially unresponsive to a number of antiepileptic drugs. Simultaneously the patient's behaviour deteriorated further and he was excluded from school.

Currently, aged six years, the patient has remained seizure-free for eight months, on a combination of carbamazepine and vigabatrin. Comprehension and speech have improved considerably (but are not yet normal) and he is awaiting placement in a mainstream school with a speech and language unit.

Discussion

This patient presented with sudden loss of speech, inattentiveness to auditory stimuli and difficult, aggressive behaviour. He was initially considered to be deaf and was referred for a hearing test and subsequently for an otolaryngology assessment. This is a common presentation for children with the Landau-Kleffner syndrome (Paquier *et al.*, 1992). Over 50 per cent of children with this disorder present with loss of understanding and speech and deafness is frequently the initial considered diagnosis. A delay in establishing the correct diagnosis may lead to significant behavioural difficulties, with possible exclusion from school (as demonstrated in this case) and also marked parental anxiety. It is clearly important to initiate early supportive therapy including speech therapy and educational assistance. Although the condition is uncommon, there is a need for an increased awareness of this disorder, particularly amongst those professionals to whom are commonly referred children with acute or subacute loss of speech/language (Thorpe *et al.*, 1991). Following a normal audiological and ENT assessment, urgent referral to a paediatric neurologist or Child Development Centre is recom-

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mended, particularly if epileptic seizures already co-exist at presentation, or develop later.

Conclusion

The Landau-Kleffner syndrome must be considered in a child who presents with acquired receptive and expressive aphasia which is unassociated with deafness.

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