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Verbal auditory agnosia with focal EEG abnormality: an unusual case of a child presenting to an ENT surgeon with "deafness"

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Abstract
Verbal auditory agnosia implies the failure to recognize sounds in a patient who is nevertheless not deaf. We present a child referred to the ENT outpatients for suspected hearing loss investigation. At one point she had grommets inserted on the basis of a flat tympanogram but with no effect. All hearing tests, including electrocochleography and distraction tests, revealed normal hearing thresholds. At the age of three years and three months, and on direct questioning of the parents, we discovered that the child responded remarkably well to music. Paediatric neurologists made the diagnosis of verbal auditory agnosia. The condition is very similar to another acquired language disorder called Landau-Kleffner syndrome. It is unusual for such a case to present primarily to an ENT surgeon.

Key words: Agnosia, verbal, auditory

Case report

A right-handed girl was referred by her GP to the ENT outpatients at the age of 17 months because of her inability to respond to sounds and to utter any word. She never babbled and her only vocalization was limited to screaming. The product of a normal pregnancy and delivery, she had had mild physiological jaundice at birth which had settled spontaneously. There was no history to suggest a previous infection episode or seizure disturbance. On preliminary assessment the child appeared bright, active and co-ordinated. We were unable to examine her tympanic membranes because of impacted wax. Tempted by a flat tympanogram the child was listed for examination, under anaesthesia, of the ears and possible insertion of grommets.

At the age of one year and nine months grommets were inserted despite the absence of middle ear effusion. The operation however did not have any impact on the child's behaviour and was judged as totally ineffective during the following months. Electrocochleography was therefore arranged and showed a threshold of 40 dB on the right and 15 dB on the left. In our centre we consider these values as normal. She was therefore put on regular follow-up by our paediatric audiology clinic.

Distraction tests of hearing done at the age of two years and seven months revealed again normal levels of hearing on both sides. However the child was not improving in as far as the parents could tell. It was not until the age of three year and three months that, on direct questioning, we discovered that the child responded to music remarkably well. The parents noted that the child used to move her body in a rhythmical way on hearing music and was known to turn up the volume of the television in the case of any pleasant music. It was then that we decided to refer the patient to a paediatric neurologist.

Neurological examination did not reveal any localizing signs. Non-verbal skills were found to be age appropriate. She showed little eye contact but gave a good smile to a cuddle. There was no evidence to suggest autism and the child showed normal interpersonal behaviour. It was concluded by the neurologist that the disturbance was one of language or auditory processing - a verbal auditory agnosia. EEG revealed a focal disturbance in the left hemisphere. She was started on anticonvulsants. A CT scan of the brain was normal. The Makaton signing system was introduced and the child has shown some improvement in her communication skills.

Discussion

Landau-Kleffner syndrome (LKS) is characterized by electroencephalographic spike discharges and verbal auditory agnosia in previously healthy children (Paeto *et al.*, 1991). An acquired disorder, it normally affects children between the ages of two and seven years (Hirsch *et al.*, 1990; Von Suchodoletz, 1992). It has been suggested by Paeto *et al.* (1991) that unilateral discharges at or near the auditory cortex disrupt auditory discrimination in the affected hemisphere, and lead to suppression of auditory information from the opposite hemisphere, thereby accounting for the two main criteria of LKS. Anti-epileptic drugs , sleep-modifying drugs and corticosteroids are among the suggested medications for this type of disorder (Marescaux *et al.*, 1990).

In our case there was no evidence to suggest that this child had an acquired disorder. However this possibility is

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not rejected altogether in case the problem was acquired earlier than it could easily be diagnosed on a clinical basis. The presence of a focal EEG abnormality with verbal auditory agnosia satisfies the two main criteria for LKS. Several cases of unusual presentations for this very interesting syndrome have been reported in the literature. These include cases with non-verbal auditory agnosia (Koeda and Kohno, 1992) and others with global regression of cortical functions (Hirsch *et al.*, 1990). It is therefore possible that our case could be a form of LKS.

The three major language disabling conditions are autism, profound deafness and mental handicap. Our child was very sociable and interactive. Both behavioural and objective tests of hearing were normal. Furthermore, apart from language development the child's milestones did not suggest any global retardation. The child had no physical or sensory disability. There was no evidence to suggest any emotional deprivation or adverse environmental factors, both of which are considered among the lesser handicapping conditions which can affect language. Our only concern which eventually caused a delay in diagnosis was the possibility of this child having intermittent middle ear effusion, with a resulting conductive hearing loss which was not detected at the time of her hearing assessment. This possibility was however excluded after grommet insertion.

It is very important that a full investigation of possible physical, mental and emotional disabilities is performed as a first course of action with any child showing language difficulty.

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