

Dysmorphia of Broca's Area with Language Delay and Hearing Loss: A Case Report

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Abstract

Case Report

We report the case of a 10-year-old girl, with no personal or family history that could account for a language or hearing disorder, referred for language delay with otherwise normal psychomotor development. Otologic and ENT examination were strictly normal. Auditory brainstem response (ABR) testing showed a bilateral hearing threshold of 50 decibels. Temporal bone CT was unremarkable, whereas brain MRI revealed dysmorphia of the foot of the third frontal gyrus (F3), bilateral and more pronounced on the left, corresponding to Broca's area. Speech-language assessment confirmed a predominantly expressive language delay. Speech therapy combined with conventional hearing aid fitting was started, with slow progress and persistently poor vocabulary and short sentences. This case highlights the value of brain morphological imaging in children presenting with language delay associated with hearing loss, and raises the question of a link between frontal gyration anomalies and language development disorders.

Keywords: language delay; Broca's area; foot of F3; childhood hearing loss; brain MRI; auditory brainstem response.

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INTRODUCTION

Language delay is a common presenting complaint in pediatrics and otorhinolaryngology, with multiple possible etiologies: hearing loss, neurodevelopmental disorder, environmental causes, or, more rarely, structural brain abnormality. Broca's area, the seat of expressive language, is classically described at the foot of the third frontal gyrus (F3), most often within the dominant, usually left, hemisphere. Dysmorphia of this region, particularly when bilateral, is an uncommon finding in the work-up of isolated language delay and raises the question of its causal, associative, or incidental role in the clinical picture.

We report the case of a 10-year-old girl presenting with expressive language delay associated with moderate bilateral hearing loss, in whom brain MRI revealed bilateral dysmorphia of the foot of F3, through which we discuss the diagnostic hypotheses and the value of a multidisciplinary work-up.

CASE PRESENTATION

A 10-year-old girl, with no notable personal history and no family or perinatal history that could account for a language disorder or hearing loss, presented with language delay. Overall psychomotor development was otherwise normal.

Otologic examination was normal, with no abnormality of the external auditory canal or tympanic membrane. The remainder of the ENT examination, as well as the rest of the clinical examination, was normal. Speech-language assessment confirmed a predominantly expressive language delay; no standardized scores were reported.

On audiological work-up, auditory brainstem response (ABR) testing showed an estimated hearing threshold of 50 decibels bilaterally. Behavioral and pure-tone audiometry were not performed.

Temporal bone CT was unremarkable.

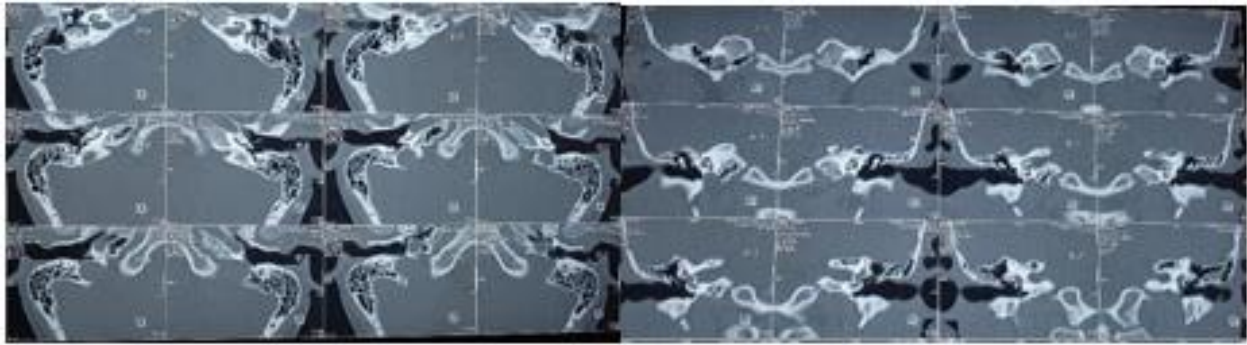


Figure 1: Image of Temporal bone CT

Brain MRI revealed dysmorphia of the foot of the third frontal gyrus (F3), bilateral and more

pronounced on the left, corresponding to Broca's area, with no other associated abnormality.

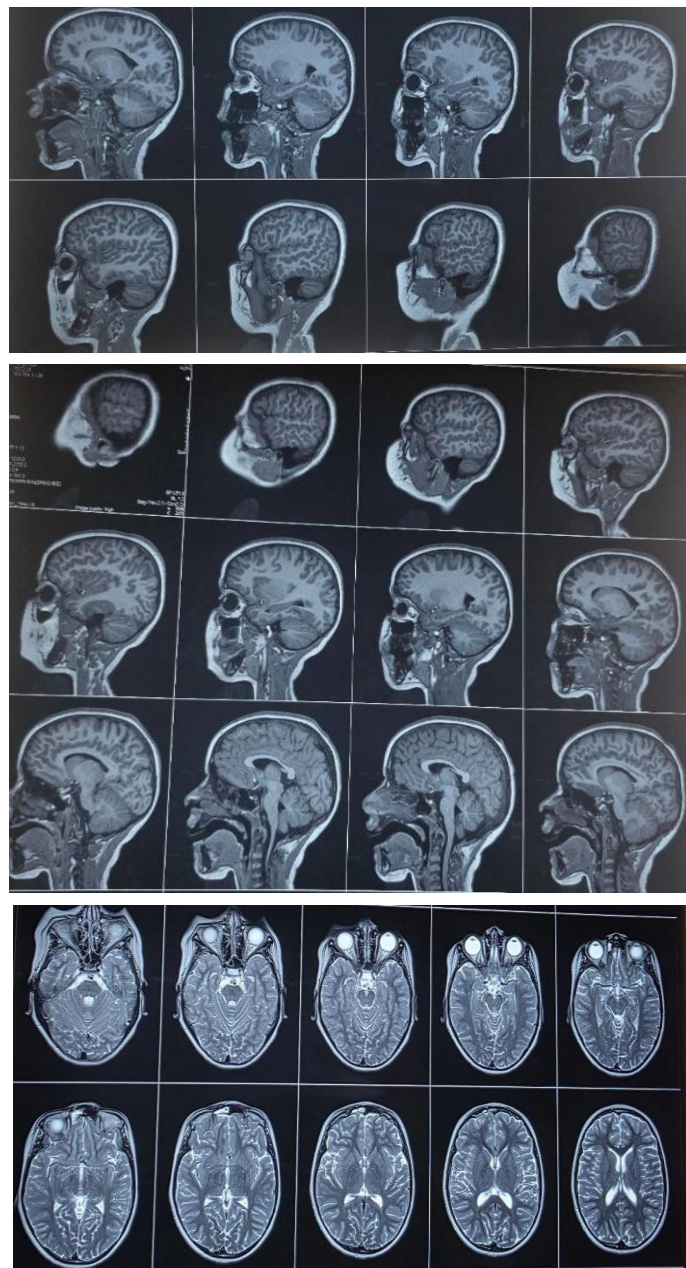


Figure 2: Brain MRI revealing dysmorphia of the foot of the third frontal gyrus (F3), bilateral and more pronounced on the left (Broca's area) with no other associated abnormality

Management combined speech therapy with bilateral conventional hearing aid fitting. Progress was slow, with persistently poor vocabulary and short sentences despite management.

DISCUSSION

Broca's area, described at the foot of F3 by Paul Broca in 1861 [1], lies immediately anterior to the rolandic operculum, which controls the phono-articulatory apparatus; it constitutes the expressive pole of language, classically lateralized to the dominant, usually left, hemisphere. The dysmorphia observed in our patient, although more pronounced on the left, is notable for its bilateral character, which is atypical given the usual lateralization of language and is more suggestive of a malformation of cortical development, such as polymicrogyria, than of an isolated anatomical variant.

The literature on bilateral perisylvian polymicrogyria is informative in this regard. Large descriptive series have shown that the topography of the cortical abnormality largely determines the severity of the clinical picture: diffuse forms involving the entire perisylvian region are associated with severe language delay or complete absence of speech, whereas more focal, particularly posterior, forms present with milder, predominantly phonological or expressive impairment [2]. A study by Guerreiro and colleagues in children with developmental language disorder confirmed that clinical severity varied with the extent of the cortical abnormality, with limited polymicrogyria corresponding to an attenuated form of the congenital perisylvian syndrome [3]. A recent systematic review further showed that expressive impairment is frequently more pronounced than receptive impairment in bilateral perisylvian polymicrogyria, consistent with the pattern observed in our patient [4], and a larger cohort study confirmed the language delay, with the cortical dysmorphia being an associated rather than strictly causal finding. frequency of speech, language, and oromotor dysfunction in these children [5].

Regarding connectivity, absence or hypoplasia of the left arcuate fasciculus, which connects Broca's and Wernicke's areas, has been identified as a specific marker of language dysfunction in children with cortical malformations involving these regions, which could represent a useful further avenue of investigation via tractography imaging in our patient, where available [6].

Several hypotheses may therefore be proposed to explain this clinical picture. The first is a direct causal link between the cortical morphological abnormality and the expressive language delay, consistent with the data above on perisylvian polymicrogyria. The second is a contributory role of the bilateral hearing loss (threshold of 50 dB, consistent with moderate hearing impairment) in the genesis of the third is that of a broader underlying

neurodevelopmental disorder, of which the frontal dysmorphia and the hearing impairment would be two expressions among others, pointing toward a genetic or malformative origin not yet characterized in this patient. These hypotheses are not mutually exclusive.

The absence of perinatal or family history, and of any associated psychomotor delay, argues against classic acquired causes (perinatal hypoxic-ischemic injury, congenital infection) and strengthens the case for further etiological work-up, particularly genetic testing, which was not performed in our patient.

On the audiological side, the 50 dB threshold obtained on ABR remains an electrophysiological estimate of hearing threshold; international guidelines emphasize the need to confirm it with behavioral audiometry once the child's age allows, as behavioral thresholds systematically differ from electrophysiological thresholds, particularly as the degree of hearing loss increases [7].

On the cerebral electrophysiological side, EEG abnormalities, including subclinical ones, have been reported in children with specific language impairment, leading to a proposed neurodevelopmental model linking cortical electrical dysfunction to language disorder [8]. This finding supports the systematic use of EEG in children with cortical dysmorphia associated with a language disorder, even in the absence of overt clinical epileptic manifestations.

Guidelines from the Haute Autorité de Santé on the identification of neurodevelopmental disorders and on speech therapy management of language disorders emphasize the need for a graduated multidisciplinary evaluation whenever warning signs of language development appear, combining hearing, speech-language, neurological, and, if needed, genetic assessment [9,10]. This case illustrates the value of systematically combining brain morphological imaging with audiological work-up in children with unexplained language delay, particularly when hearing loss alone does not appear to account for the severity of the clinical picture. A multidisciplinary approach involving otorhinolaryngology, neuropsychiatry, genetics, and speech therapy appears warranted to refine the etiological diagnosis and inform prognosis.

CONCLUSION

The association of expressive language delay, moderate bilateral hearing loss, and bilateral dysmorphia of the foot of F3 in this child illustrates the complexity of the etiological diagnosis of language delay in children. It underscores the importance of a comprehensive multidisciplinary evaluation (audiological, speech-language, neuroradiological, and genetic) before concluding on a simple causal link between a cerebral

morphological abnormality and a language development disorder.

Declarations

Conflicts of interest: the authors declare no conflict of interest related to this work.

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