A DIFFICULT CASE

not just a child with simple learning disability beware the opercular syndrome

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An 8-year-old girl with a speech disorder was admitted to our Child Neurology Department. She had been born at term after an uneventful pregnancy, but on delivery she did not start to cry and she was cyanotic. Her APGAR score was 4 within the first minute and rose to 7 in 10 minutes. Head circumference, weight and height were normal. There was no hypotonicity and she had no difficulty in sucking in the Neonatal Intensive Care Unit . There were no convulsions or episodes of apnoea.

Developmental milestones such as holding her head upright, sitting and walking unsupported were all passed on time. She started saying single words at the age of two years, but she did not get beyond this. At the age of five, she still had the speech difficulty but on examination the child was not considered to have any additional clinical problems other than being a little hyperactive - the paediatrician explained to the family that her mental ability had been affected by the difficult delivery, and that her condition was untreatable.

By the age of seven, her speech was barely intelligable, and she was unable to go to school. Her younger sister was completely healthy, and her parents were not consanguineous. There was no history of any neurological or psychiatric disorders in the familiy. Systemic examination of the child did not reveal any abnormalities in the cardiovascular or respiratory systems - neither dysmorphism nor cutaneous lesions were detected. She was right-handed, awake and fully oriented but her speech was severely dysarthric, and she was drooling. She was able to comprehend the conversation and her mental status was judged to be normal. She had a left facial weakness. Gag reflex was weak on both sides. She could not move her tongue to the right or left, and she was unable to protrude it. She was able to rapidly repeat labial consonants (me-me), but not guttural consonants (ga-ga). The pyramidal, extrapyramidal and cerebellar systems were all normal. Except for her timidity, she had no affective inappropriateness and displayed no signs of hallucinations or agitation. She was able to comprehend conversation, read aloud, and write. Her hearing seemed sufficient, a tympanogram was normal and also brainstem auditory evoked responses. Following a detailed interview, we felt that the child was quite cooperative and had a performance IQ (Weschler Intelligence Scale for Children) of 91. Routine blood and urine tests, and screening tests for congenital metabolic diseases were all normal. She had no seizure disorder, and her EEG was normal.

Brain MRI revealed bilateral perisylvian and insular hypointensities, more prominent in the right frontal area on T1-weighted axial images

(Figure 1), and hyperintensity in the perisylvian regions on both sides and fronto-parietal operculum on T2- weighted and FLAIR images (Figure 2). The lesions were interpreted as encephalomalacia and gliosis due to her difficult birth.

As she was a year behind her peers in starting elementary school, we decided she should get special education to improve her verbal skills. After two years, thanks to speech therapy, she is now able to talk more clearly, although still not very fluently. However, her teachers have reported that she is very good at conveying her knowledge in written form. Her formal performance measures were quite satisfactory.

DISCUSSION

Also known as the Foix-Chavany-Marie syndrome, the opercular syndrome in adults is characterized by restricted movements of the tongue, orofacial paresis and dsyarthria, which develops acutely due to bilateral damage of the anterior operculum (1). The symptoms are directly related to the involvement of the insular and opercular areas and are caused by stroke, viral encephalitis (particulalarly herpes simplex) and status epilepticus. But aetiological factors in children are more varied - congenital bilateral perisylvian polymicrogyria, bilateral perisylvian pachigyria, and schizencephalia are some of the most common (1,2). Gliosis due to perinatal hypoxia or encephalomalacia may also lead to the opercular syndrome where, as in our case, the condition is static, as in other cerebral palsies and the dysarthria can be rehabilitated and the child's eduction restored. An isolated opercular syndrome can be mistakenly diagnosed as learning disability, again as in our case. However, appropriate identification of the pseudobulbar signs and the characteristic pattern of lesions on brain MRI help make the diagnosis.

The formation of the insula takes place in the 18th week of gestation and it is covered by the operculum in the 30th week. If MRI shows that the insula is partially covered or not at all covered by the operculum, this suggests that the development stopped due to some condition which had occurred before the 30th week. The detection of polymicrogyria or heterotopia is evidence that the anomaly developed some time between the 12th-16th weeks, when neuronal migration and organization take place. Encephalomalacic and gliotic lesions in this area are more suggestive of perinatal hypoxia, as in our case.

The severity of oromotor dysfunction is related to how symmetric the involvement of the perisylvian region is, rather than the extent of the lesions (1). Asymmetrical anomalies cause just mild dysarthria and dysphagia. More symmetric lesions cause an opercular syndrome with severe dysarthria, drooling, severe dysphagia/dysphonia and bilateral/unilateral central facial paralysis.

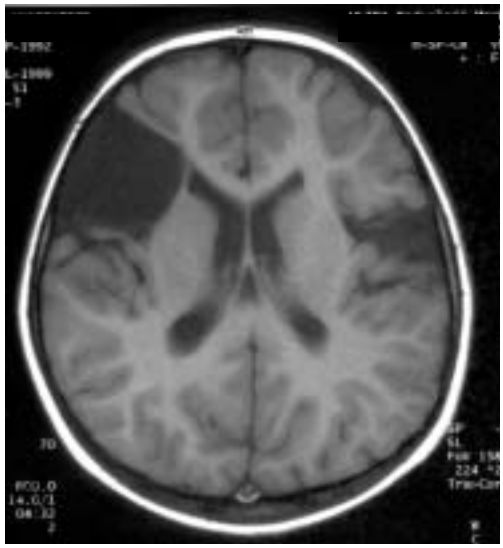

Figure 1 (T1 axial): Bilateral perisylvian and right frontal encephalomalacic areas

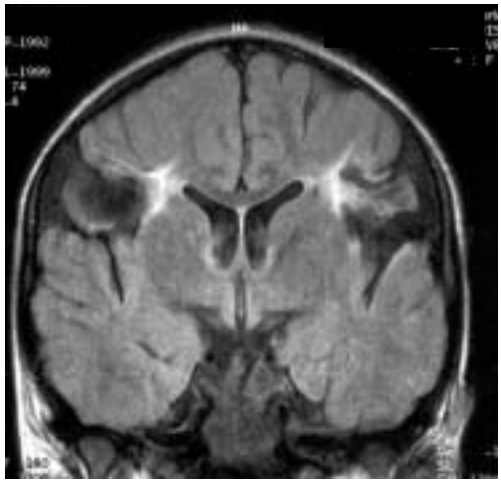

Figure 2 (Flair coronal): Bilateral hyperintense gliotic lesions with hypointense encephalomalasic areas

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