Olfactory nerve hypertrophy: a clue to the presence of ipsilateral megalencephaly

Rajesh Shankar Iyer,[1] Bejoy Thomas,[2] Saleem Akhtar,[1] Praveen Kumar[3]

[1]Department of Neurology, KG Hospital, Coimbatore, Tamil Nadu, India
[2]Department of Imaging Sciences and Interventional Radiology, Sree Chitra Tirunal Institute for Medical Sciences and Technology, Trivandrum, Kerala, India
[3]Department of Radiology, KG Hospitals, Coimbatore, Tamil Nadu, India

Correspondence to
Dr Rajesh Shankar Iyer,
dr_rsh@hotmail.com

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DESCRIPTION

A boy aged 4 years presented with refractory focal seizures since 9 months of age. The seizures involved the left side of the body and consisted of clonic movements with frequent generalisation. MRI brain showed abnormal area with poor grey-white differentiation involving the right frontal lobe, especially the basifrontal region and the temporal lobe (figure 1A,B) suggesting the presence of a frontotemporal developmental malformation. Interestingly olfactory nerve hypertrophy was also noted on the same side (figure 1C,D). Video-electroencephalogram study demonstrated concurrence of the interictal (figure 2A) and ictal data (figure 2B) with the MRI defined abnormality. An 18-fluoro-2-deoxy-D-glucose positron emission tomography/CT of the brain showed severe right frontotemporal hypometabolism. The boy underwent a disconnection procedure involving the right frontal lobe sparing the motor cortex and the temporal lobe. Histopathology of the abnormal brain obtained during the surgery showed large atypical neurons, balloon cells, astrocytosis and neuronal heterotopia suggesting localised megalencephaly. The child is seizure-free at 6 months follow-up.

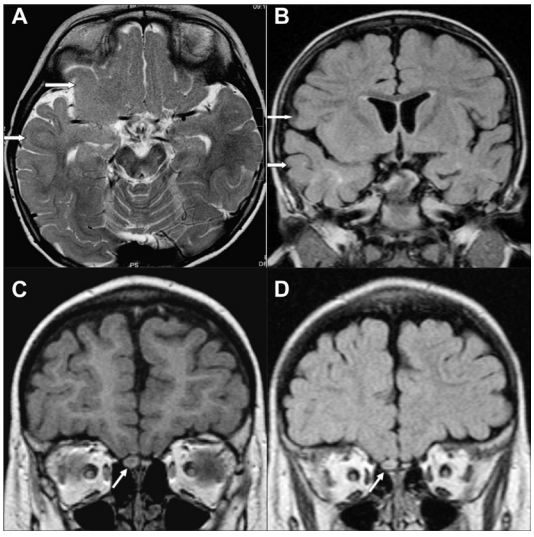

Figure 1 Brain MRI axial T2 weighted TSE(A) and coronal T2 FLAIR (B) showing the malformation of cortical development involving the right frontotemporal region (arrows). Coronal T1 weighted FLASH (C) and Coronal T2 FLAIR (D) showing the hypertrophy of the olfactory tract on the right side (arrow). FLAIR, fluid attenuation and inversion recovery; FLASH, fast low angle shot; TSE, turbo spin echo.

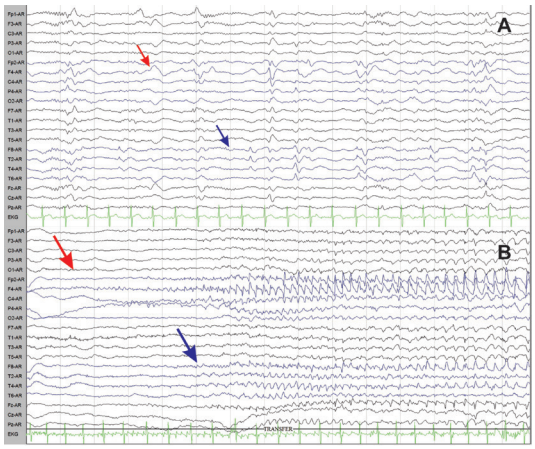

Figure 2 Electroencephalogram (EEG) showing interictal discharges (A) over the right frontal (red arrow) and right temporal regions (blue arrow). Ictal record (B) demonstrating the seizure onset from the right frontal region (red arrow) spreading further to the right temporal region (blue arrow).

Olfactory nerve hypertrophy is a morphological abnormality detected on MRI, outside the involved hemisphere. This was noted in 8 out of 30 patients (26.7%) with hemimegalencephaly in one study.[1] The other such abnormalities occurring outside the involved hemisphere are cerebral vascular dilations and cerebellar enlargement. These abnormalities are seen more in association with hemimegalencephaly or localised megalencephaly. They are generally not seen in association with multilobar cortical dysplasia.[2] It is sometimes difficult to differentiate between localised megalencephaly and multilobar cortical dysplasia from the MRI

Learning points

- Olfactory nerve hypertrophy can sometimes be seen in the MRI ipsilateral to a cerebral developmental malformation.
- It is usually seen in association with localised megalencephaly involving the anterior cerebral quadrant and also with hemimegalencephaly and not seen in association with focal cortical dysplasia.
- The detection of this finding in the MRI may be a clue to the underlying ipsilateral megalencephaly.

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picture, as happened in our case. However the presence of olfactory nerve hypertrophy was the clue which can help identify ipsilateral diffuse as well as anterior quadrantic type of localised megalencephaly.[2]

The magnocellular cholinergic neurons of the basal forebrain constitute the major cholinergic output of the central nervous system. They innervate the neocortex, hippocampus and the olfactory bulb where there is maximum expression of the choline acetyl transferase activity. Nerve growth factor (NGF) produced by the neurons regulate choline acetyl transferase activity. In hemimegalencephaly, abnormal brain expresses increased levels of NGF and NGF-receptor positive cells whose interaction is responsible for neuronal growth and differentiation.[3] Olfactory nerve fibres on the same side as the hemimegalencephaly also express increased amounts of NGF and NGF-receptor positive cells thereby explaining the hypertrophy.

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