Teaching Neuro*Images*: Long-term outcome of untreated Rasmussen syndrome

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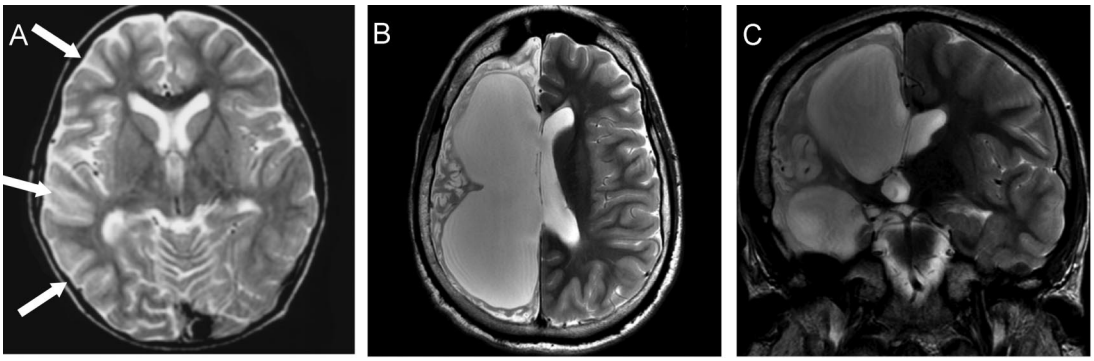
Figure **Serial T2-weighted MRI brain showing changes over time in untreated Rasmussen syndrome**



Earliest abnormal MRI at 3 years 2 months of age (A) during the acute phase shows increased cortical signal (arrows). MRI at 13 years 9 months (B, C) in the residual phase shows severe unilateral atrophy of the entire right hemisphere with normal signal intensity. Signal intensity correlates with level of inflammation (T cells and reactive astrocytes).[2]

A 13-year-old boy diagnosed with Rasmussen syndrome as a toddler presented with worsening seizures. History revealed onset of left-sided focal seizures in a healthy 18-month-old child. Epilepsia partialis continua was refractory to anticonvulsants and immunotherapy. MRI showed inflammatory changes (figure, A). Hemispherectomy and neurology follow-up were declined. Off anticonvulsants, seizures plateaued and development regressed to a nonverbal encephalopathy with hemiparesis. The latest MRI shows end-stage atrophy (figure, B and C). Rasmussen syndrome is a progressive, focal autoimmune encephalitis of unknown etiology that leads to intractable epilepsy, cognitive decline, and hemiparesis.[1,2] Immunotherapy or hemispherectomy is recommended,[2] so long-term neuroimaging of untreated patients is rare.

REFERENCES

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