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Clinico-pathological Conference

Nine-Year-Old Girl With Blank Stares and Recent-Onset Diabetes

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Patient Presentation

A 9-year-old previously healthy girl presented with the new onset of multiple blank starring episodes lasting 30-60 seconds and being "slow to respond." These episodes were preceded by two to three weeks of progressive polydipsia, polyuria, and decreased appetite. Her past medical history was unremarkable, and her growth and development had been normal. There was no family history of seizures or neurological disorders. Her initial examination was normal aside from altered mental status. Upon evaluation her blood glucose was found to be 533 mg/dL, pH 7.34, and bicarbonate 24 mmol/L. She also had 3+ urine ketones and 3+ glucose. Complete blood count, electrolytes, liver function tests, and renal studies were unremarkable.

Upon hospitalization and treatment with intravenous fluids and insulin, her glucose rapidly corrected, yet she continued to have altered mental status with frequent staring spells accompanied by speech arrest. There was no postictal confusion associated with these episodes, and her interictal speech and mentation remained normal.

Her initial electroencephalographic (EEG) recording captured a typical episode of speech arrest. The electrographic correlate consisted of rhythmic sharply contoured theta activity over the left temporal-parietal region. Interictal epileptiform activity was also present over the left temporal-parietal region. Prominent mixed theta and delta frequency slowing of moderate to high amplitude was noted over this region. She was given a loading dose of intravenous levetiracetam and then placed on maintenance therapy, which decreased the frequency of the episodes without total control.

Her initial computed tomography (CT) scan without contrast (Fig 1) demonstrated a small focal hyperdensity along the left anterior frontal cortex (arrow), with associated decreased attenuation of the overlying extra-axial space. These findings were suggestive of hemorrhage or calcification with overlying fat (based on Hounsfield units).

Her cerebrospinal fluid (CSF) studies were normal except for elevated glucose, which corresponded with her elevated serum glucose. CSF herpes simplex virus type 1 and 2 DNA replication and bacterial and fungal cultures were negative. The CSF IgG:albumin ratio, IgG index, and olgioclonal bands were normal. Tuberculosis skin testing was negative. Serum Epstein-Barr virus antibody panel and cryptococcus antigen

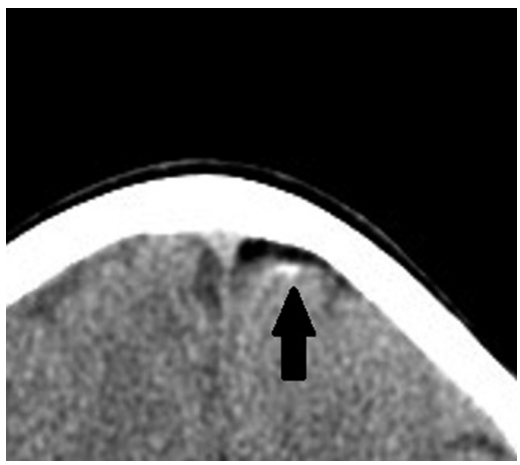

FIGURE 1.
Computed tomography without contrast reveals a small focal hyperdensity along the left anterior frontal cortex (arrow), with associated decreased attenuation involving the overlying extra-axial space.

The authors have no conflicts of interest to disclose.

Editor's note: This clinicopathological correlation represents a new feature for Pediatric Neurology. Submission requirements for these papers can be found in our online instructions for authors.

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TABLE.
Laboratory Evaluation Results

Laboratory Test	Patient's Value	Normal Range
CSF color	Clear, colorless, no xanthochromia	-
CSF glucose	141 mg/dL*	45-75 mg/dL, serum glucose was 355 mg/dL*
CSF white blood cells	2	0-10
Differential	11% neutrophils, 73% lymphocytes, 16% monocytes	-
CSF RBCs	<1000	<1000
CSF protein	17 mg/dL	15-45 mg/dL
CSF culture	No growth, no organisms on Gram stain	-
CSF HSV 1&2 DNA amplification	Negative	-
CSF fungal culture	No fungus at 4 weeks	-
CSF acid fast culture	Negative	-
CSF IgG	1.1 mg/dL	0-6.0 mg/dL
CSF IgG:albumin ratio	0.12	0.09-0.25
CSF IgG index	0.66	0.28-0.66
CSF IgG synthesis rate	<0.0 mg/day	≤8.0 mg/day
CSF oligoclonal bands	Negative	Negative
CSF myelin basic protein	0.18 ng/mL	0-5.50 ng/mL
CSF angiotensin converting enzyme	0.8 U/L	0.0-2.5 U/L
Tuberculosis PPD	0 mm at 72 hours	<15 mm
Cryptococcal Ag	Negative	-
Mycoplasma pneumoniae Ab		
IgG	0.00 U/L	<0.09 U/L
IgM	0.04 U/L	≤0.76 U/L
EBV antibody panel	Negative	Negative
Thyroid peroxidase antibody	<1.0 IU/mL	<9.0 IU/mL
Thyroglobulin antibody	<1.0 IU/mL	<1.0 IU/mL-negative
TSH	1.97 mlU/L	0.50-4.50 mlU/L
ANA IgG	None detected	None detected
Tissue transglutaminase antibodies IgA	3 Units	0-19 Units
Serum IgA	144 mg/dL	45-234 mg/dL
Human insulin antibody	<5.0 μU/mL	<5.0 μU/mL-negative
Anti-GAD 65 antibody	4.1 U/mL*	>0.5 U/mL-positive
ICA 512 antibody	36 U/mL*	>1.0 U/mL-positive
B2 glycoprotein 1		
Antibody IgG	0 SGU	0-20 SGU
Antibody IgA	2 SGU	0-20 SGU
Antibody IgM	5 SGU	0-20 SGU
ANCA	<1:20	<1:20
Lupus anticoagulant panel		
PT	13.8 seconds	12-15.5 seconds
PTT	34 seconds	32-48 seconds
dRVVT 1:1	32 seconds	33-44 seconds
Cardiolipin antibodies		
IgG	13 MPL	0-14 MPL
IgM	6 MPL	0-12 MPL
ESR	7 mm/hour	0-20 mm/hour
Serum quantitative amino acids	normal	-
Angiotensin converting enzyme	30 U/L	24-121 U/L
Pyruvic acid	0.099 mmol/L	0.030-0.107 mmol/L
Lactate	0.9 mmol/L	0.7-2.1 mmol/L
Urine organic acids	Slight generalized elevation of acylglycines	Nonspecific, may indicate gastrointestinal problem

Abbreviations:
ANA = Antinuclear antibody
ANCA = Anti–neutrophil cytoplasmic antibody
CSF = Cerebrospinal fluid
dRVVT = Dilute Russell viper venom time
EBV = Epstein-Barr virus
ESR = Erythrocyte sedimentation rate
HSV = Herpes simplex virus
PPD = Purified protein derivative
PT = Prothrombin time
PTT = Partial thromboplastin time
RBC = Red blood cell
TSH = Thyroid-stimulating hormone
* Denotes abnormal result.