A previously-healthy 18-month-old girl had a right hemi-body seizure followed by prolonged hemiparesis. Blood pressure was above the 99th percentile for her age, and it was higher in the upper extremities (UE) than in the lower extremities (LE). Femoral pulses were decreased.

Figure 1 shows the brain MRI. A transthoracic echocardiogram revealed an ejection fraction of 22%, without intracardiac shunting. An abdominopelvic CT angiogram demonstrated a midaortic syndrome (Figure 2). Hypercoagulability and rheumatological studies were normal.

Midaortic syndrome is a rare disorder with stenosis of the distal thoracoabdominal aorta. Its classic triad includes abdominal bruit, elevated UE/LE blood pressure ratio, and decreased LE pulses. Most cases (~60%) are idiopathic. Secondary causes/associations include aortitis, atherosclerosis, neurocutaneous syndromes, and Williams syndrome. Midaortic syndrome patients can develop subarachnoid or intraparenchymal hemorrhages or, rarely, ischemic strokes\textsuperscript{1,2,3}.

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{brainMRI.png}
\caption{Neuroimaging findings: Non-contrast brain MRI shows subacute left parietal infarct (arrows) on diffusion-weighted (1A), apparent diffusion coefficient (1B), and T2-FLAIR sequences (1C). Susceptibility-weighted images reveal scattered foci of microhemorrhages (1D, asterisks). Given imaging findings consistent with embolic stroke, severely decreased left ventricle ejection fraction, lack of intracardiac shunting, normal hypercoagulability and rheumatological studies, and no other sources of emboli, the stroke mechanism is likely cardioembolic from severe systolic dysfunction.}
\end{figure}

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Figure 2. Midaortic syndrome: Abdominal CT angiogram shows tapering of the abdominal aorta just below the renal arteries (arrowhead), and stenosis at the origin of the celiac trunk and superior mesenteric artery (arrows).

References

