Neuroimaging Highlight

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Asphyxiation Causing Distinctive Basal Ganglia Injury and Generalized Dystonia

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A previously healthy five year-old boy was seen after intermittent strangulation attempts over the course of two days. Details of the event are vague due to lack of reliable adult witnesses. There was no indication of toxic exposure. General examination on arrival to the emergency department revealed an alert boy with bruising of the neck consistent with multiple strangulation attempts. There was no evidence of heart, kidney, or liver involvement to suggest systemic hypoperfusion or organ failure. Investigations done on arrival showed no acidosis, and a capillary blood gas 48 hours later revealed a normal carboxyhemoglobin. Although the initial neurologic exam was near normal, severe generalized dystonia developed over the next four weeks, requiring intrathecal baclofen treatment. No akinetic or rigid symptoms developed, however speech gradually

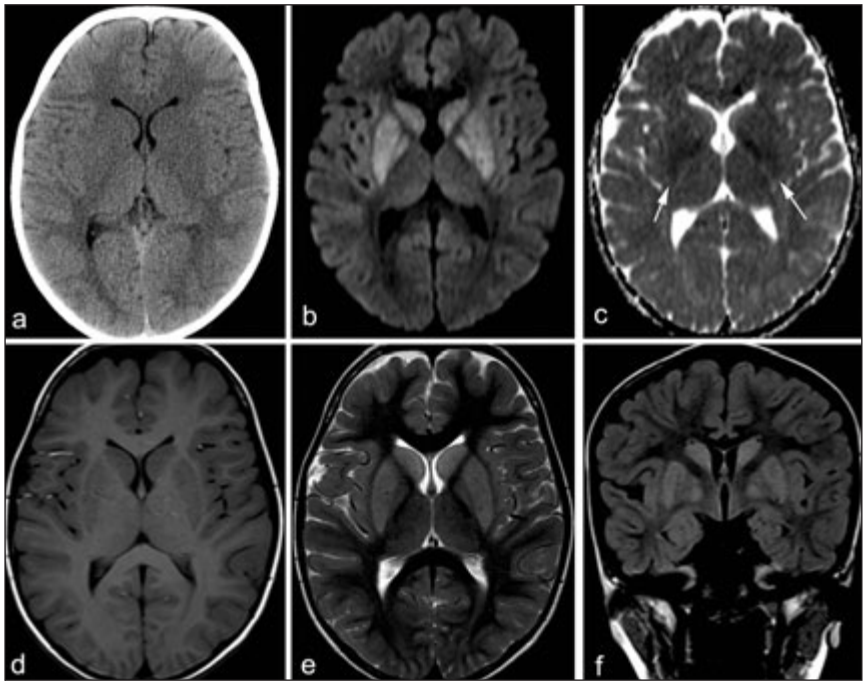

Figure 1: *Non-contrast CT scan at presentation (a) shows hypodensity of bilateral caudate nuclei, putamina, and globi pallidi. Diffusion-weighted image (DWI) (b) one day after initial CT shows hyperintense signal in bilateral globi pallidi, putamina, and caudate nuclei. Apparent diffusion coefficient (ADC) map (c) shows slightly decreased signal in bilateral globi pallidi and putamina (arrows). T1-weighted MRI (T1-MRI) (d) does not show obvious abnormality. Slightly increased signal is seen in bilateral globi pallidi, putamina, and caudate nuclei on both T2-weighted (T2-MRI) (e) and fluid attenuated inversion recovery (FLAIR) (f) images.*

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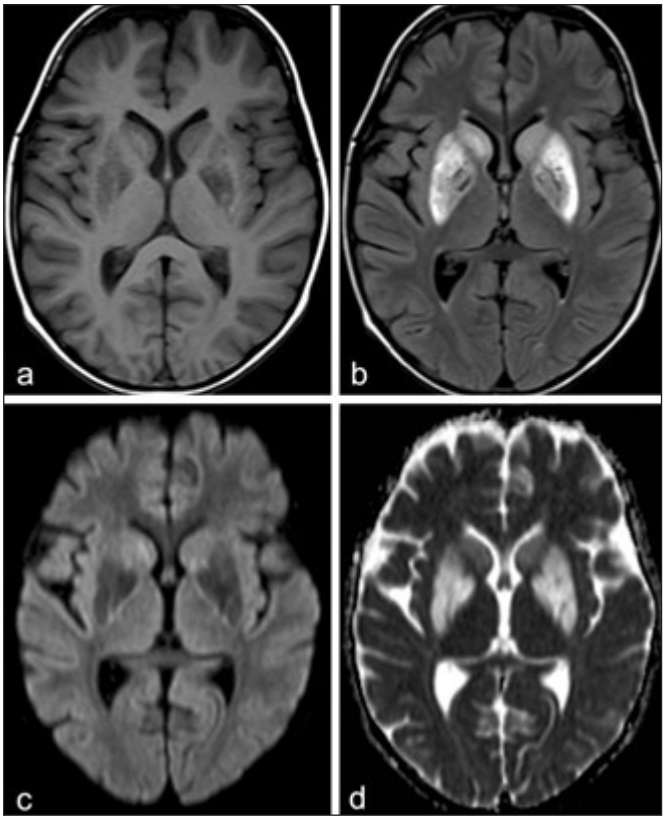

Figure 2: *MRI six weeks after initial CT shows persistent signal changes in bilateral basal ganglia. The putamina and globi pallidi are (a) hypointense on T1-MRI (b) predominantly hyperintense on FLAIR images (c) hypointense on DWI (d) and hyperintense on ADC map. The signal changes of the caudate nuclei are much milder, and only appreciated as mild hyperintensity on FLAIR and ADC map.*

diminished until there was no verbal output. Despite this, he remained able to perform poorly controlled voluntary movements and to communicate through language boards using pointing. Profound axial hypotonia developed over the initial two weeks and did not improve with treatment. Six months after the injury neurological disability remained significant with minimal signs of further improvement.

Initial computed tomography (CT) scan showed bilateral hypodensities in the caudate nuclei, putamina, and globi pallidi with sparing of the thalamus and cortex (Figure 1). Magnetic resonance imaging (MRI) completed soon after showed diffusion weighted (DWI) hyperintensities and apparent diffusion coefficient (ADC) hypointensities in the corresponding basal ganglia structures indicating acute infarction (Figure 1). Follow up MRI showed persistent abnormalities within the basal ganglia (Figure 2). Given the history of strangulation, these changes almost certainly resulted from hypoperfusion injury. Other etiologies for acute basal ganglia changes are shown in the Table.[1]

The selective vulnerability of the basal ganglia to hypoperfusion injury has been well documented, as has the wide range of movement disorders that may develop[2]. Dystonia is more common in younger patients, while older patients are more prone to akinetic-rigid syndromes[3]. This case is remarkable in that the striatum and globi pallidi were both severely affected, with sparing of remaining brain structures, including the thalamus and cortex.

The vascular supply of the striatum differs from that of the globi pallidi. The intrinsic properties of the neurons contained within these structures may therefore account for the pattern of damage in this case, as opposed to watershed lesions. Glutamate has been suggested as a key player in ischemic brain damage[2], however the numbers of excitatory amino acid receptors varies significantly between the striatum and the globi pallidi[4], suggesting the likelihood of an alternate mechanism of our patient's basal ganglia lesions. It is possible that repeated attempts at asphyxiation led to sensitization of neurons to injury in some basal ganglia structures, with neuroprotection of other structures through "ischemic tolerance"[5]. Alternatively, the severity and duration of the repeated attempts at asphyxiation likely differed between events, resulting in separate injuries to the striatum compared with the globi pallidi[6]. Given the severity of the damage seen on imaging and the persistence of neurological deficit, the likelihood of full recovery or significant improvement is low for this patient.

Table: Differential diagnosis of acute basal ganglia disease in childhood. Adapted from Barkovich 2005[1].

Hypoxia
Hypoglycemia
Osmotic myelinolysis
Encephalitis
Parainfectious encephalomyelitis
Tegretol toxicity

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