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LETTER TO THE EDITOR

Brush sign in Sturge-Weber syndrome

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Dear Editors,

We read with great interest the pictorial review by Linscott et al [1], "Imaging patterns of venous-related brain injury in children," which offers a clear and valuable summary of these patterns. Furthermore, we would like to share with the readers of Pediatric Radiology an imaging pattern related to Sturge-Weber syndrome – the brush sign.

The brush sign was first described by Morita et al. [2] in 2008 in a T2*-weighted sequence in the context of acute stroke and was defined as asymmetrical visualization of the subependymal and medullary veins in the deep white matter (Fig. 1). This phenomenon was attributed to BOLD (blood-oxygen-level-dependent) effect due to an increase in intravenous deoxyhemoglobin in cases with major vessel occlusion, reflecting an increased oxygen extraction fraction. Later, Horie et al. [3] identified the brush sign in susceptibility-weighted imaging (SWI) of patients with Moyamoya disease and used this sign to assess disease severity by determining the number of conspicuous deep medullary veins. Again, this phenomenon was attributed to increased oxygen extraction as well as to the chronic state of cerebral vasodilation in these patients.

Sturge-Weber syndrome is a sporadic condition associated with vascular malformation of the skin, eye and brain that classically includes facial port-wine stain and ipsilateral pial venular angioma. Abnormal development of the cortical veins is a typical feature, and disease progression has been linked to progressive venous occlusion and resulting venous stasis, for which aspirin is recommended by some clinicians [4]. Engorgement of the medullary veins is commonly seen and was previously described in conventional angiography and MRI [5, 6]. This enlargement resembles the brush sign (Fig. 1) and can be detected in T1-W contrast-enhanced imaging (as in girl with known in Linscott et al. [1]) and is also delineated clearly in SWI sequences, which enhance the abnormal neurovasculature and therefore improve the detection of the prominent deep veins [7]. These collateral channels develop early in the disease and, to a certain extent, are efficient in preventing damage to the adjacent cortex [8], as they can be appreciated in imaging before cortical hypometabolism develops [8]. Furthermore, the development of collaterals may also increase as the disease progresses, as seen in a case presented by Mentzel et al. [9], who demonstrated an increase in medullary veins in a follow-up MRI in a neonate in comparison with the initial exam.

In conclusion, Sturge-Weber syndrome should be added to the list of diseases causing the brush sign. It develops due to engorgement of the medullary and subependymal veins serving as collateral channels from the cortex to the deep venous system resulting from the progressive occlusion of the abnormal cortical veins, and an increase in local deoxyhemoglobin concentration caused by hypoxia due to venous stasis.

To determine whether this sign can facilitate assessment of disease severity, according to the number or the degree of engorgement of conspicuous deep medullary veins, or whether it can be used to monitor therapy (e.g., effect of aspirin), further studies with serial MRI and clinical correlation are required.

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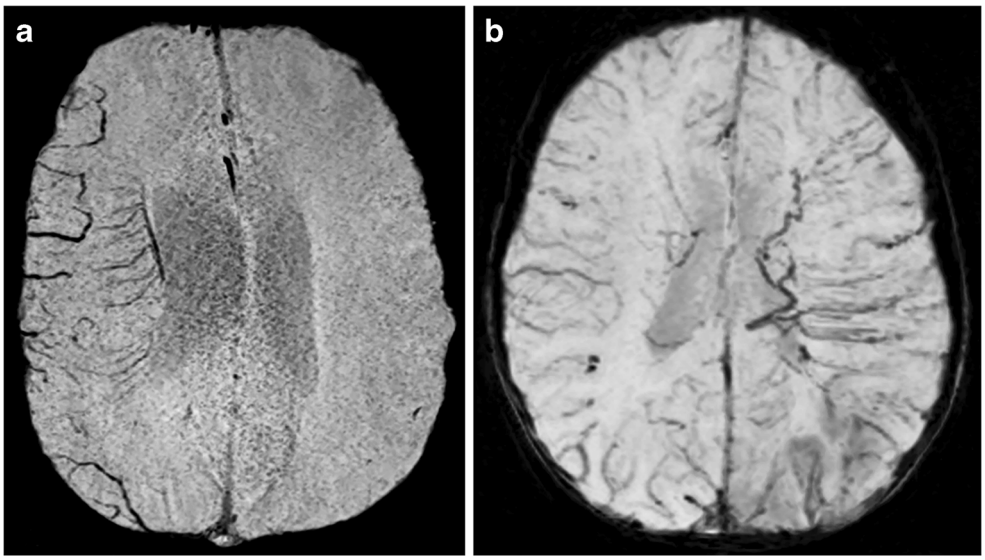

Fig. 1 Minimum intensity projection reconstructed susceptibility-weighted images from two patients. Both images show engorged medullary veins resembling brush sign. **a** A 15-year-old girl with acute occlusion of the right middle cerebral artery. **b** A 2-year-old girl with a known Sturge-Weber syndrome Roach type 1

Compliance with ethical standards

Conflicts of interest None

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