Neuroimaging Highlight

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The Cutaneous Angioma of Sturge-Weber Syndrome

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A 16-month-old male presented with focal motor seizures with secondary generalization. On physical exam, a bilateral facial cutaneous angioma, sharply demarcated by the distribution of the ophthalmic and maxillary branches of the trigeminal nerve, was noted (Figure 1). Magnetic resonance imaging (MRI) demonstrated calcification and atrophy of the right frontal lobe as well as an associated pial angioma, consistent with the clinical suspicion of Surge-Weber syndrome (SWS) (Figure 2). However, no abnormality was seen in the right hemisphere, or in the parietal or occipital lobes. Further evaluation also revealed unilateral glaucoma on the right, but with bilateral diffuse choroidal hemangiomas.

Surge-Weber syndrome is a non-hereditary neurocutaneous disorder characterized by leptomeningeal angiomatosis with an associated facial cutaneous angioma, also called a port-wine nevus. The abnormality is thought to be due to a malformation of the embryonic vascular plexus within the cephalic mesenchyme at five to eight weeks gestation, interfering with the development of vascular drainage.[1] Due to poor drainage of the eye, face, leptomeninges and brain, angiomatosis develops as an alternate pathway for venous drainage,[1] leading to multiple neurological and ophthalmologic complications, including seizures, headaches, stroke like episodes and glaucoma.

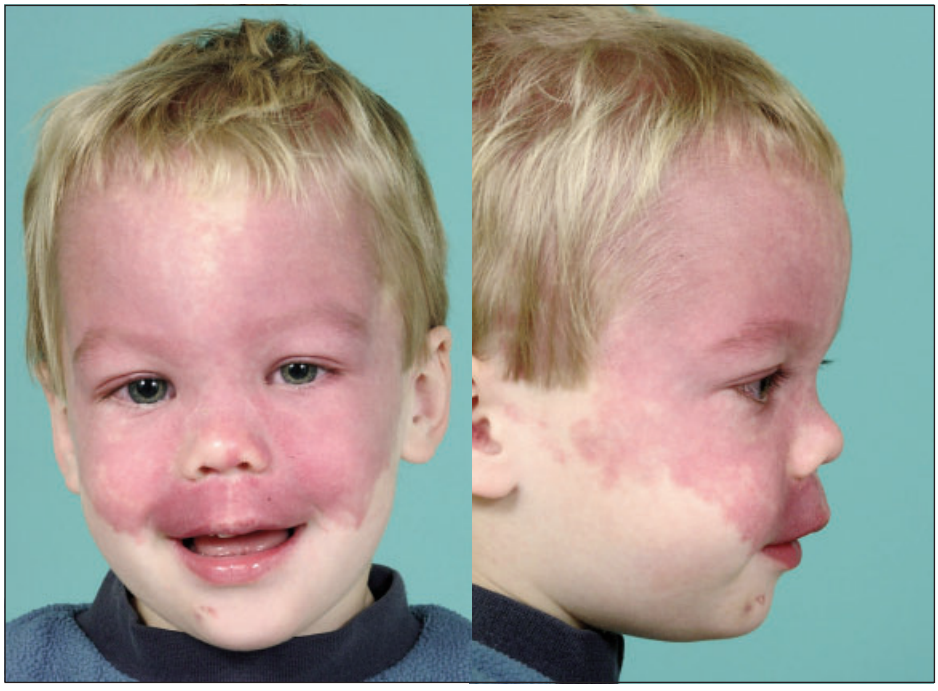

Figure 1: *The facial angioma, shown in both "face-on" and lateral views, can be seen to follow exactly the distribution of the first two divisions of the trigeminal nerve, involving the forehead, eyelids and nose, while sparing the pre-auricular area.*

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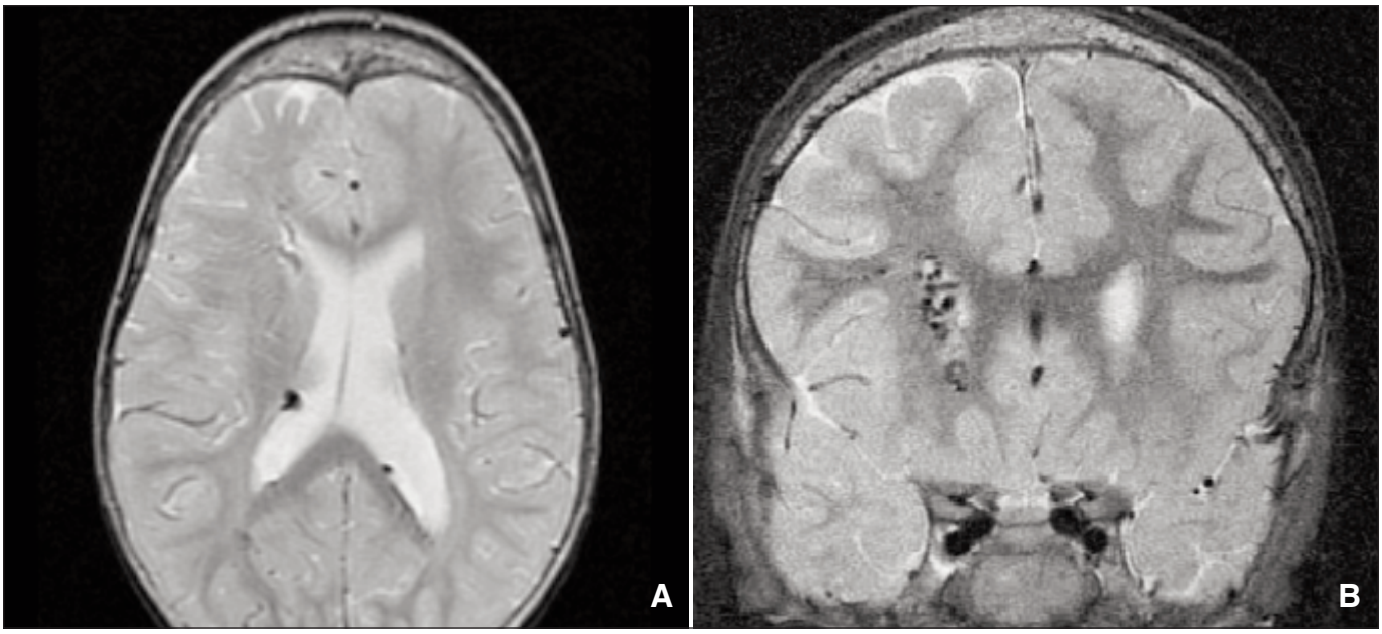

Figure 2: *Axial(2a) and coronal (2b) MRI images demonstrating early atrophy of the right frontal lobe with an associated anomalous venous structure and increased signal within the right frontal sulci, suggestive of an underlying pial angioma.*

In 1860, Schirmer was the first to characterize the anatomical correlation between cutaneous angioma and the trigeminal nerve.[2] Sturge, however, gave the initial clinical account of the syndrome in 1879, and called the nevus a "port-wine" stain.[3] In 1900, Cushing noted that the port-wine stain followed the distribution of the trigeminal nerve branches,[4] while Weber is credited with the first radiologic report of the intracranial calcifications seen on skull radiographs in 1922.[5]

Since the venous abnormality accounts for the principal clinical manifestations, SWS can also be called Cerebrofacial Venous Metameric Syndrome (CVMS). This classification is based on the embryological abnormality, specifically the involvement the rostral mesoderm and neural crest cells. Involvement of both the medial and lateral prosencephalic group derived from the neural crest cells leads to the involvement of the forehead and nose (CVMS 1), as well as the occipital lobe eye, cheek and maxilla (CVMS 2). Thus, our patient would be classified as CVMS 1+2[4].

In our patient, despite bilateral congruent cutaneous angiomas, as well as bilateral choroidal involvement, leptomeningeal angiomatosis and glaucoma were only present on the right side. Only 10-20% of children with a port-wine nevus of the forehead have a leptomeningeal angioma, which is typically ipsilateral to the facial nevus.[6] Tallman et al followed 310 patients with Sturge-Weber syndrome and found that 85% of patients had a unilateral lesion, while 15% had a bilateral lesion, and 68% involved more than one dermatome of the trigeminal nerve. However, only those patients with involvement of the V1 branch had ocular involvement and only if the eyelid was involved.[7] Furthermore, CVMS-2 usually involves the occipital lobe as well as the eye, cheek and maxilla as they all originate from the lateral neural crest cells;[4] however, in our patient, involvement of the occipital lobe is notably absent.

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