

Short communication

# Spontaneous obliteration of MRI-silent cerebral angiomatosis revealed by CT angiography in a patient with Sturge–Weber syndrome

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Received 19 May 2007; accepted 23 July 2007

Available online 27 August 2007

## Abstract

Type I Sturge–Weber syndrome (SWS) is a neurocutaneous syndrome characterized by facial port wine stain and ipsilateral leptomeningeal angiomatosis. It is commonly complicated by glaucoma and neurological disturbances including seizures, hemiparesis, transient stroke-like deficits, and behavioral problems. Gadolinium-enhanced MRI is the study of choice for demonstrating the leptomeningeal angiomatosis, and is particularly useful for making the diagnosis. Cerebral hematoma and spontaneous obliteration of cerebral angiomatosis have never been reported in SWS. We describe a patient with type I SWS in which a hemorrhage in the left temporal lobe was caused by an underlying angiomatosis. No arteriovenous malformations were detected by brain CT or gadolinium-enhanced MRI, whereas a small angiomatous nidus draining into an ectasic venous collector near the hematoma was disclosed by brain CT angiography. Carotid angiography, performed 3 years after the hemorrhage, demonstrated the spontaneous obliteration of the angioma. This description expands the phenotypic spectrum of type I SWS in which the occurrence of cerebral hemorrhage and MRI-silent cerebral angiomatosis should be included, and emphasizes the need of performing a brain CT angiography in SWS patients whose routine neuroimaging studies fail to detect potentially harmful vessel malformation. © 2007 Elsevier B.V. All rights reserved.

**Keywords:** Arteriovenous malformation; Brain CT angiography; Brain MRI angiography; Cerebral hemorrhage; Epilepsy; Sturge–Weber syndrome

## 1. Introduction

Type I Sturge–Weber syndrome (SWS) is a rare neurocutaneous syndrome characterized by facial port-wine nevus and histological or radiographic features of ipsilateral intracranial angioma, and is commonly complicated by ipsilateral glaucoma (“classic” SWS) [1]. Neurological disturbances include seizures, headache, hemiparesis, transient stroke-like deficit, and behavioral problems [2]. Gadolinium-enhanced MRI is the study of choice for demonstrating the typical leptomeningeal angiomatosis,

and is particularly useful for making the diagnosis [3,4]. Intracerebral hematoma and spontaneous resolution of the cerebral angiomatosis have never been reported in SWS. Here we describe a patient with type I SWS in which spontaneous resolution of an MRI-silent cerebral angiomatosis was demonstrated by brain CT angiography.

## 2. Case report

A 37-year-old woman who was born after a dystocic labor showed at birth paresis of the right arm and typical port-wine stain on the left hemiface involving the distribution of the I and II trigeminal branches. Apart from slight paresis with brisk tendon reflexes and hypotrophy of the right arm, she had normal neurological and cognitive development. She successfully completed secondary school. At the age of 13

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she developed left-sided severe glaucoma which was successfully operated. She married at the age of 20 and had 3 normal children. Since the age of 22 she has worked as a civil servant. At the age of 25, brain CT and gadolinium-enhanced cerebral MRI, performed because of recurrent headaches, gave normal results and failed to show any feature of SWS (Fig. 1). At the age of 37 she had 2 brief episodes characterized by brightness in the right visual field, dysarthria, nausea, and a strange feel in her right arm, followed by loss of consciousness. No antiepileptic treatment was initiated. A few days after the second seizure she was admitted to the emergency department because of three serial seizures characterized by right-sided facial-brachial hemiclonic jerks and motor aphasia followed by secondary generalized tonic fits and transient (30') right hemiparesis. Laboratory data were unremarkable and she had no risk

factors for stroke, including cardiac diseases (normal electrocardiograms and transthoracic echocardiography), hypertension, diabetes and smoking. Brain CT at admission revealed a 3.5-cm-diameter hematoma in the temporal lobe of the left hemisphere (Fig. 2 A, B). Brain CT angiography, performed 7 days after admission, revealed a small angiomasia (a cortical nidus of about 2 cm diameter, draining into an ectasic venous collector) near the temporal hematoma (Fig. 3A–C). Ten days after admission, an MRI performed with and without contrast confirmed the presence of the hematoma and failed to show leptomeningeal enhancement (LH) in the left temporal lobe (Fig. 2 C, D). On the same day, MRI angiography did not show arteriovenous malformations (Fig. 3D–E). Fifteen days after admission the patient had no additional deficits and was discharged on treatment with levetiracetam 1.5 g/day.

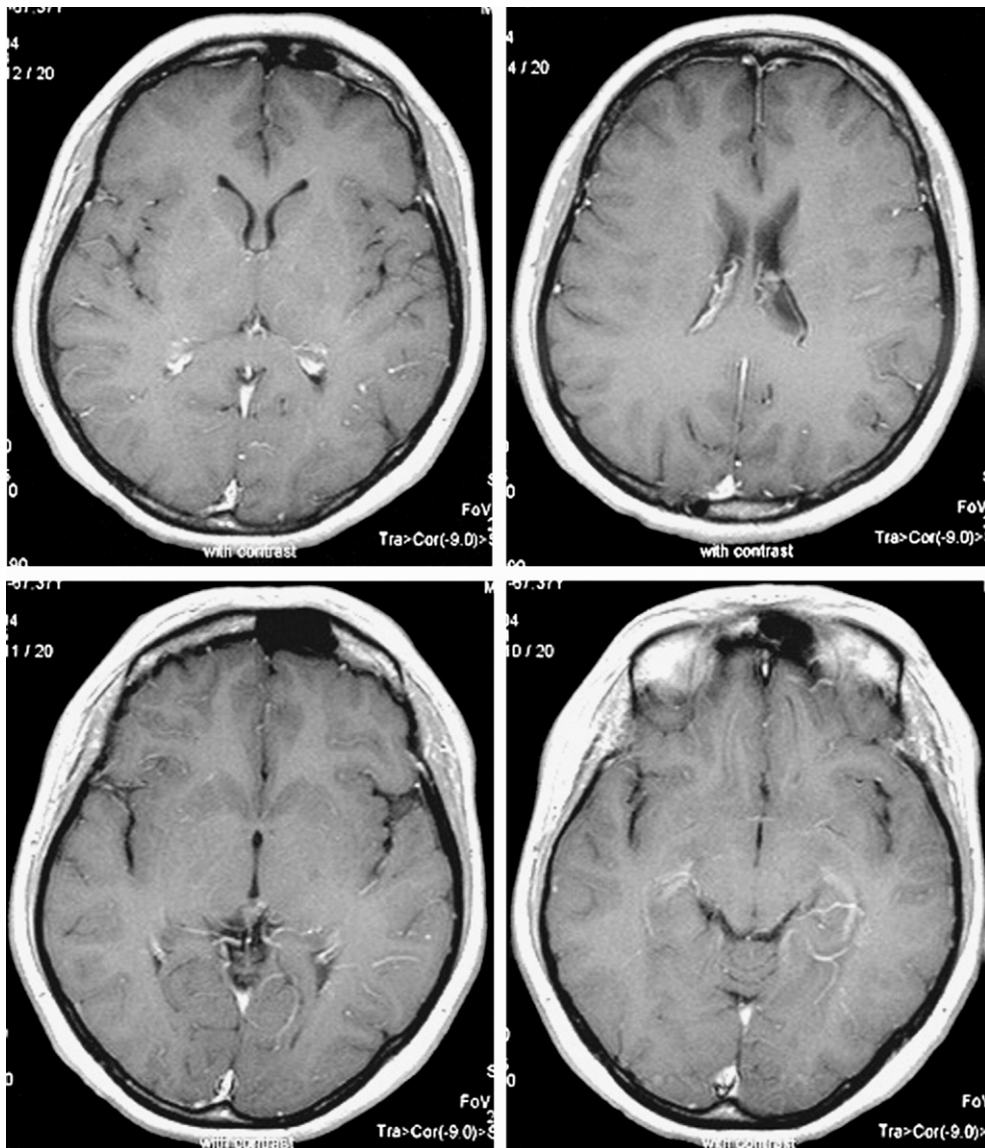


Fig. 1. Gadolinium-enhanced MRI at the age of 25 shows no leptomeningeal enhancement. Note absence of choroid plexus thickening and enhancement on the left side. There are no developmental abnormalities of the brain.

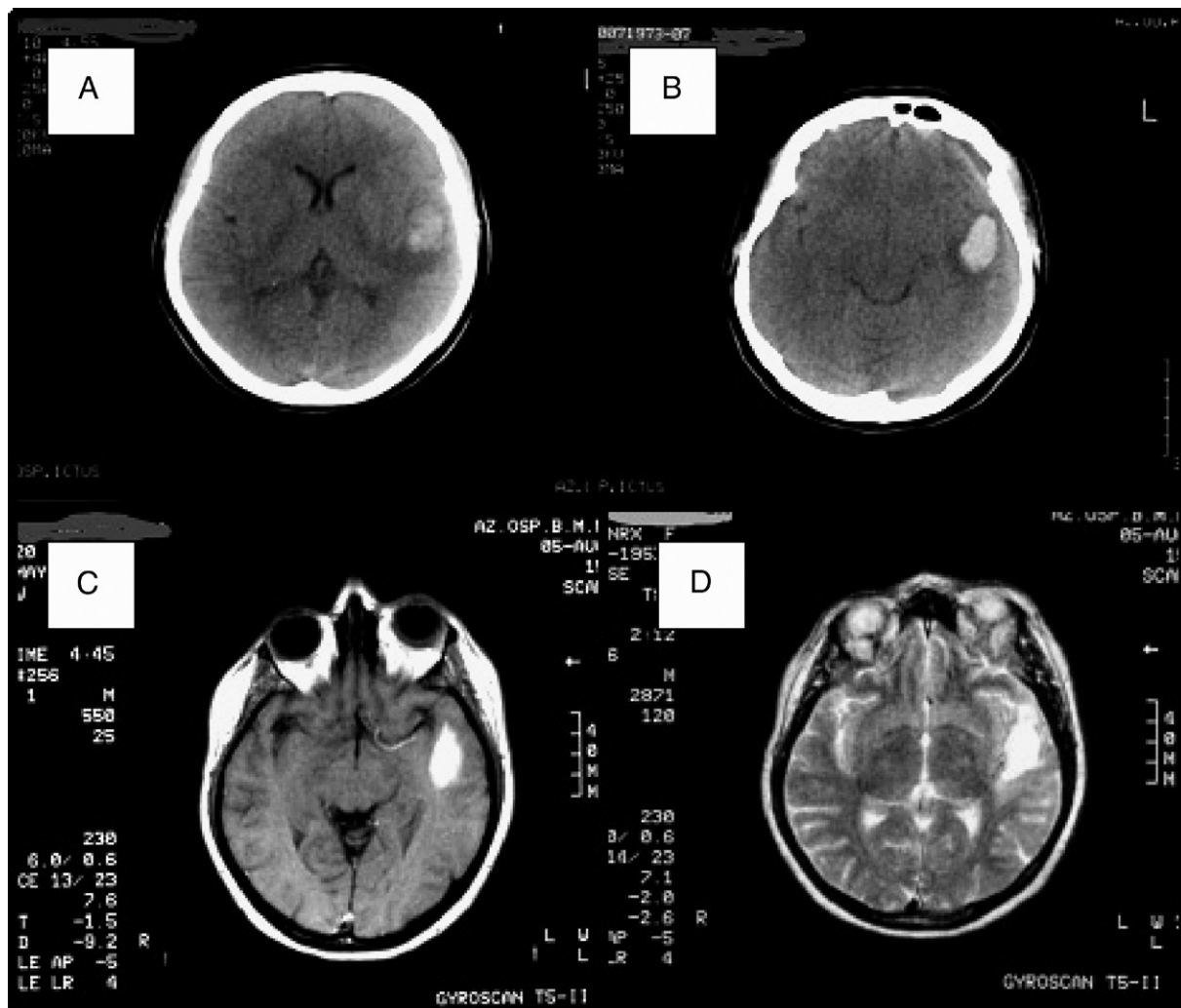


Fig. 2. A, B: Axial brain CT in the emergency department: hematoma of 3.5 cm diameter in the left temporal lobe. Note the perilesional edema. C, D: MRI 10 days after admission (C: axial T1-weighted MRI with contrast enhancement; D: axial T2-weighted MRI): persistence of the hematoma (C, D). There is no leptomeningeal enhancement (C).

During the following 2 years she had monthly seizures characterized by nausea and feelings of strangeness, and sporadic secondary generalized tonic–clonic fits. Replacing levetiracetam with carbamazepine (600 mg/day) completely prevented seizures during the following year. Three years after the hemorrhage brain CT angiography failed to show the previously documented angiomatous nidus, and carotid arteriography confirmed its obliteration (Fig. 4).

### 3. Discussion

The young woman described here has all the clinical characteristics of type I SWS [1,2] including typical facial portwine stain, glaucoma and neurological abnormalities (paresis with hypotrophy of the right arm and seizures). However, the patient lacked both cranial CT scan (cerebral atrophy, cortical calcification, ipsilateral bone thickening and sinus hypertrophy) and brain MRI (LH or gyral contrast enhancement and choroid plexus thickening and enhancement) findings of SWS

[3,4]. Minimal or absent LH has been described in a few SWS-patients whose cranial CT scan and brain MRI showed different stigmata of the disease [3,5]. No LH and presence of focal brain transmedullary enhancement on routine gadolinium-enhanced MRI have recently been described in one patient with type I SWS in which three dimensional time-of-flight MR venography showed left parietal LH [6]. We did not observe focal brain transmedullary enhancements, but we failed to perform venography, so we were unable to confirm the value of this method in increasing the chances of detecting LH. It has been hypothesized that the absence of LH in these patients is the result of obliteration of the leptomeningeal vessels as a result of chronic progressive thrombosis [5]. LH is thought to represent either leakage of contrast media or slow flow through anomalous leptomeningeal microvasculature. Lack of involvement of the leptomeningeal vessels may explain the absence of LH in the patient described here. The first cerebral lesion diagnosed in this patient was a small left temporal hemorrhage. Stroke-like episodes [7], deep cerebral venous occlusion [8]

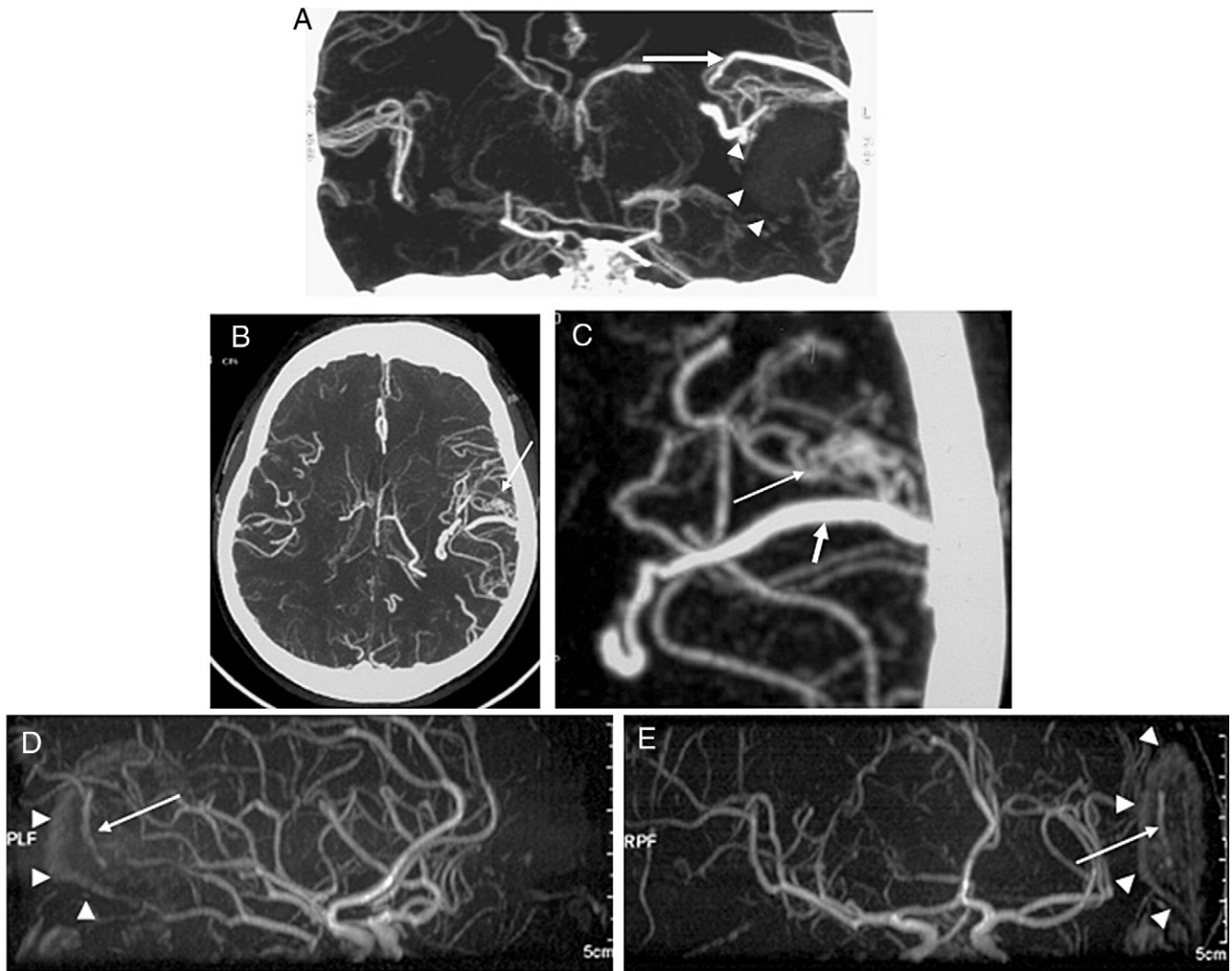


Fig. 3. A, B, C=Brain CT angiography performed 7 days after admission. Coronal (A) and axial (B and C) planes: angiomatous malformation (arrows) near the hemorrhage (arrowheads) in the left temporal lobe. Note a cortical angiomatous nidus (thin arrows) draining into an ectatic venous collector (thick arrows). C, D=MRI-angiography performed 10 days after admission. Lateral (C) and oblique coronal (D) planes. There is no evidence of arteriovenous malformations in the left hemisphere. Only an ectatic venous collector (arrows) is evident near the hematoma (arrowheads).

and hemiplegic migraine [9] may occur in SWS. Although the most common presentation of arteriovenous malformations of the brain is cerebral hemorrhage [10], intracranial bleeding is not a “major problem” in SWS [1]. To our knowledge, ours is the first documentation of the occurrence of a cerebral hemorrhage in an SWS patient. This patient had two major risk factors for hemorrhagic arteriovenous malformation presentation, including adult age and exclusive deep venous drainage [11]. Of interest, the small angioma was not revealed by brain MRI or MRI-angiography, and was well documented by brain CT angiography. This was probably due to the low sensitivity of MRI to detect tiny arteriovenous malformations having high flow velocity. Finally, we documented spontaneous resolution of the cerebral angioma after the bleeding. Spontaneous regression or disappearance of the LH over time has been described in a few SWS-patients [12–14]. Resolution of a temporary state of increased venous pressure due to seizure

activity [13] has been hypothesized to explain this unusual time evolution of the LH in SWS, but it seems not to be relevant in this patient. Small dimension, superficial localization, feeding by a single artery and draining by a single vein make spontaneous obliteration of the arteriovenous malformation likely in this patient [15]. It has been hypothesized that progressive venous restriction leading to occlusion in the presence of protective factors may explain the spontaneous obliteration of arteriovenous malformations [16]. Such protective factors may include feeding vessels with a large pressure drop along their length, or spasm of the feeding vessels in the presence of hemorrhage. These factors would prevent an increase in intranidal pressure during venous occlusion, thereby limiting the size of hemorrhage from the nidus, and may explain the small size of the left temporal hematoma in this patient.

In conclusion, this description expands the phenotypic spectrum of type I SWS in which the occurrence of cerebral

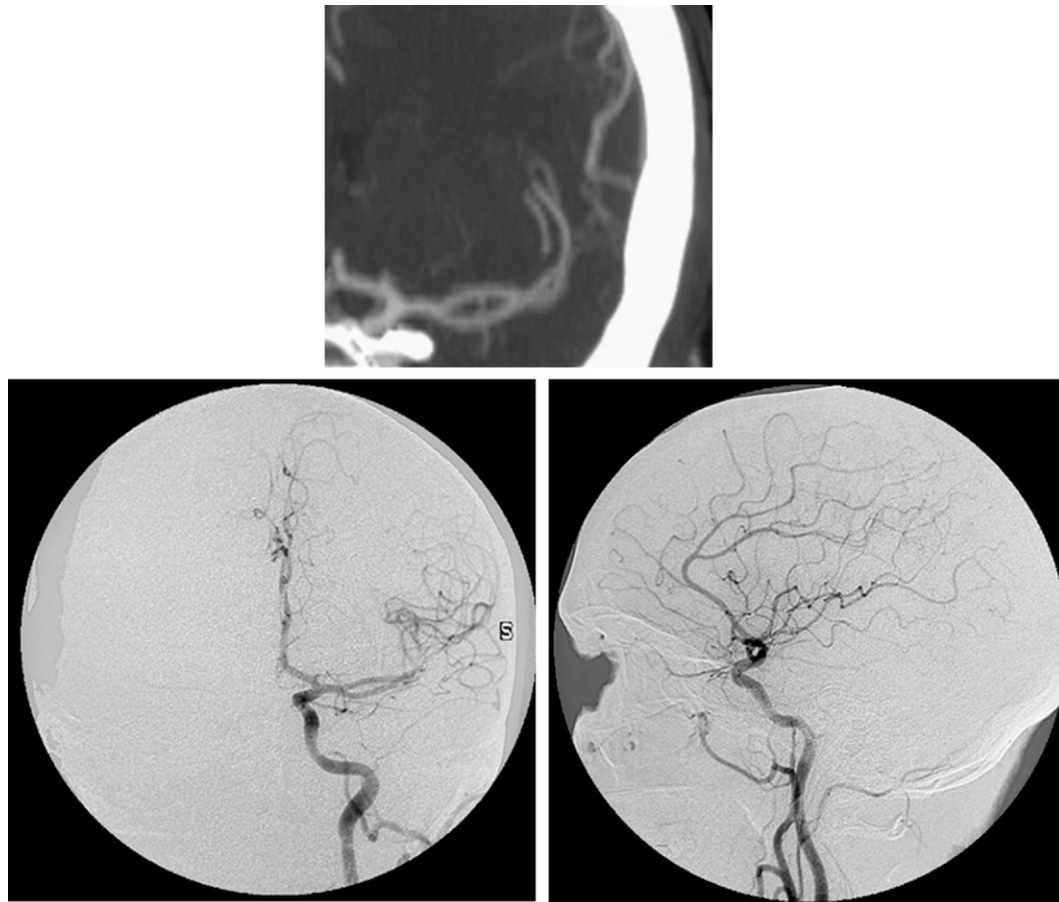


Fig. 4. Neuroimaging study performed 3 years after the hemorrhage. Brain CT angiography, coronal plane (top): disappearance of the angiomatous nidus. The ectasic venous collector remains evident. Left carotid arteriography (bottom) with no evidence of angiomatosis.

hemorrhage and MRI-silent cerebral angiomatosis should be included, and emphasizes the need of performing brain CT angiography in SWS patients whose routine neuroimaging studies fail to detect potentially harmful vessel malformation.

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