HEREDITARY HEMORRHAGIC TELANGIECTASIA (HHT) or the Rendu-Osler-Weber syndrome, is an autosomal dominant disorder characterized by the presence of multiple dermal, mucosal, and visceral telangiectasias associated with intermittent bleeding episodes. Epistaxis is the most common presenting manifestation of this bleeding tendency, but gastrointestinal hemorrhage, hemoptysis, or hematuria may also occur. Neurological complications of HHT as a consequence of vascular malformations have become more commonly recognized in recent years, but are still not appreciated widely. One type of vascular malformation, intracerebral saccular aneurysm has been recognized recently as a potential source of thromboembolic material leading to transient ischemic attacks (TIA) or completed cerebral infarction in patients without HHT. We report the first HHT patient with ischemic cerebrovascular symptoms as the presenting manifestation of an intracerebral aneurysm.

Case Report
A 55-year-old woman with a history of recurrent epistaxis, multiple dermal telangiectasias and a family history of telangiectasias, but an otherwise unremarkable past medical history was evaluated for right-sided weakness. The first episode occurred four weeks prior to admission and was characterized by the abrupt onset of right arm and leg weakness and numbness with associated slurred speech. The symptoms resolved entirely over two days. On the day of admission the patient noted the abrupt onset of right leg weakness causing her to fall to the floor. The right arm was also weak but to a lesser degree. The episode gradually resolved over 30 minutes. Later that evening the patient again noted the abrupt onset of right leg weakness and numbness without any associated language or visual disturbance.

General physical examination was remarkable only for the presence of multiple dermal telangiectasias. Vital signs were normal. Neurologic examination disclosed a mild right central facial weakness and a moderate right hemiparesis with associated decreased tone. Pain, temperature and vibratory sensations were diminished over the right arm and leg. Right sided deep tendon reflexes were suppressed but the right plantar response was extensor. The rest of the examination, including detailed language function testing was unremarkable.

Laboratory studies included a normal CBC, PT, PTT, platelet count, bleeding time, serum electrolytes, arterial blood gases, serum protein electrophoresis, ESR and ANA. Chest X-ray and full lung tomograms were unremarkable as were the ECG, echocardiogram and 24 hour Holter monitor. EEG, L.P. and initial CT scan were also normal. Bilateral carotid and vertebral angiography disclosed bilateral carotid aneurysms at the level of the carotid-ophthalmic artery junction, and otherwise normal extracranial and intracranial vasculature (fig. 1).

On the morning following admission, the patient's right sided weakness had progressed to a flaccid hemiparesis. A CT scan was repeated 1 week after admission and revealed an area of low density in the region of the left corona radiata and posterior limb of the internal capsule compatible with a subacute ischemic infarction (fig. 2). Over the ensuing course of her hospitalization the patient's right sided weakness progressively resolved to a mild paresis. The patient was discharged on 300 mg of aspirin per day and has remained asymptomatic. Repeat angiography two years after the initial presentation demonstrated no significant change in the size of the aneurysms.

Discussion
Our patient was carefully screened for the presence of commonly recognized mechanisms which predispose to the development of stroke such as: extracranial arteriosclerotic and non-arteriosclerotic vascular disease, a source for cardiac emboli, episodic arrhythmia, orthostasis, arteritis, hypercoagulability, polycythemia, thrombocytosis, and most importantly in HHT a pulmonary arteriovenous fistula (PAVF). Investigations for these entities were all unrevealing. There was also no evidence of a small intracerebral hemorrhage as might occur with the rupture of a telangiectasia.

Unruptured intracerebral saccular aneurysms can cause transient or permanent ischemic neurological symptoms on the basis of aneurysm to artery emboli. Documentation requires arteriographic evidence of thrombus in or extruding from the aneurysmal sac and distal emboli in a patient without any other recogniz-
Figure 1. Top row: Right common carotid injection shows carotid-ophthalmic artery aneurysm. Bottom row: Left common carotid injection shows smaller carotid-ophthalmic artery aneurysm.

Figure 2. Top row: Normal unenhanced CT scan on day of admission. Bottom row: Infarction in the region of left posterior limb of internal capsule and corona radiata, one week later.
Heparin for Lacunar Stroke in Progression

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SUMMARY Four patients with mild hemiparesis were treated with heparin for presumed progressing stroke. All worsened to hemiplegia with pure motor deficits and lacunar infarcts despite this medical intervention. Clinical distinction between large artery and small, penetrating vessel thrombotic disease is needed to best evaluate any benefit of anticoagulation for stroke in progression.

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ANY EFFICACY OF HEPARIN to halt or reverse the neurologic signs of acute progressing stroke could be specific for stroke subtypes with particular clinical and vascular pathological profiles. Anecdotal and controlled studies of heparin use in patients with serially evaluated and progressing hemiparesis, generally within 36 hours of onset, do not clearly distinguish whether the causative pathology is in a carotid, vertebrobasilar, major branch, or penetrating artery.

From 20 to 60 percent of lacunar infarcts producing a pure motor deficit progress by history or serial examination in a steady, step-wise, or fluctuating pattern. We anticoagulated four patients with this lesion in an attempt to halt progression, but they continued to deteriorate until hemiplegic.

Methods From a group of 120 consecutive patients with stroke admitted to the Daniel Freeman Hospital Medical and Rehabilitation Center, 29 suffered presumed lacunar infarcts with just motor deficits. Cerebral localization and vascular pathology for all 120 stroke patients were based on criteria established by the neurologists participating in the NINCDS Pilot Stroke Data Bank. Nine patients who progressed in the hospital according to physicians’ notes and patient confirmation were not seen by us until their deficits appeared stable. None of these patients had been anticoagulated. We did examine 5 patients serially and appreciated progression of their pure motor deficit. One had a

References

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