PAINFUL TONIC SPASMS are a distinct, descriptive clinical entity. We believe this is the first case of known association with a contralateral putaminal infarct. These movements consist of a sudden, vigorous muscle spasm, preceded or accompanied by pain in the same limb(s). Usually unilateral, the arm is affected oftener than the leg, although both may simultaneously be involved. Facial grimacing can occur directly from facial spasm, or be reactionary to severe limb pain. Spasms last seconds to 1-2 minutes, and recur in stereotyped fashion daily, or even several times per hour. Temporary remissions may occur. Attacks can be triggered by hyperventilation, anxiety, physical activity or sensory stimuli, but usually are spontaneous. A classic "tetanic" posture can be assumed during a spasm, with arm adduction at the shoulder; elbow, wrist, and metacarpophalangeal flexion; and, interphalangeal extension. Variant postures with extension, fisting or clawing also exist. Flexion or extension occurs in the lower limb, usually with inversion of the foot.2,4

Case Report

This 33-year-old woman was healthy until ten years ago when a prolonged bleeding time was discovered during evaluation for tonsillectomy. Neither patient nor family had a clinical bleeding disorder. The patient had recurrent bouts of pyelonephritis and keratoconjunctivitis. Progressive thrombocytopenia, resistant to steroid therapy, improved after splenectomy seven years ago.

Soon thereafter, retrobulbar neuritis of the right eye occurred, followed by five recurrences of retrobulbar neuritis, in either eye, over the ensuing seven years. Typically, her vision would painlessly diminish to "finger counting" at five feet, with quadrantic or altitudinal visual deficits, central scotomas, and afferent pupillary defect. The optic disc would pale without papilledema or vascular abnormalities. Her vision repeatedly responded to steroids, so she was kept on a chronic daily dose of 15 mg. prednisone.

During the second episode of retrobulbar neuritis, further evaluation included a normal EEG, CT Scan, and temporal artery biopsy. Lumbar puncture revealed 1 RBC, 7 WBC (all lymphocytes), glucose = 68 mg/dl, protein = 48 mg/dl (normal electrophoresis, 9% gamma globulin), with cultures and VDRL negative.

The patient was felt to have systemic lupus erythematosus with lupus anticoagulant, leading to prolonged partial thromboplastin time (patient PTT = 31.6-48.0 sec., control = 29.0 sec.). Elevated ANA titers ranged to 1:640 and hypocomplementemia was present. Venous and arterial thromboses later occurred in the left leg and a small intestinal obstruction was surgically relieved. There was never any arthralgia, myalgia, rash, alopecia, or Raynaud's phenomenon.

Four months ago, the patient began to have acute episodes of painful flexor stiffening of her right arm and hand. Flexion occurred at the elbow, wrist, metacarpophalangeal, and interphalangeal joints. Occasionally the right leg was involved also, but the face was spared. These spells lasted 30-60 seconds, occurring 12-15 times daily, usually spontaneously. At times, they happened when she stretched her arm and neck, as when bathing in the shower. Right hand numbness and clumsiness remained between episodes. There was no associated clonic movement, march, or impairment of consciousness.

On examination, there was a left monocular temporal deficit and mild bilateral optic disc pallor. Acuity
was 20/20 without afferent pupillary defect. Mild pronator drift of the right arm with pseudoathetotic movements was seen. Reflexes were brisker on the right with a right Babinski sign. Position sense was slightly diminished in the right fingers with other modalities intact.

No metabolic abnormalities were found, including normal serum calcium, phosphate, and liver enzymes. EEG was normal, even during two episodes of tonic spasm. CT (fig. 1) showed a 4–5 mm. lucency in the outer left putamen without enhancement, which was not detected by MRI Scan. Furthermore, the multiple periventricular demyelinating lesions typical of multiple sclerosis were not seen on MRI Scan.

The painful tonic spasms were fully controlled with carbamazepine, 600 mg. daily.

Discussion

Various movement disorders are related to lacunar infarcts of the basal ganglia. Hemiballismus from a lesion in the contralateral subthalamic nucleus of Luys is well known. Acute hemichorea and hemiballismus have been reported in contralateral infarcts of the caudate and internal capsule, with or without putaminal involvement. Virtually constant distal chorea of the limbs with occasional proximal ballistic movements were described. Continuous proximal chorea with orofacial dyskinesia has been reported with bilateral caudate and putaminal infarcts. Chronic dystonic posturing of the hand described as “slow, sustained and twisting” was associated with a contralateral “lenticular” infarct. None of these movements however, were paroxysmal or painful, and they responded to haloperidol.

Painful tonic spasms are frequently associated with multiple sclerosis (MS), often from brainstem and spinal cord lesions. Watson and Chiu described painful tonic spasms in the hemiparetic limbs of a young woman with MS, who had a presumed plaque, on CT, in her internal capsule. Possibly basal ganglia involvement would be evident at autopsy. Lance has noted that cases of painful “tonic seizures” are associated with basal ganglia lesions such as hepatolenticular degeneration, encephalitis lethargica and choreoathetotic disorders (familiar or “acquired”). Unfortunately, many of such cases were in the pre-CT era or lacked autopsy data.

Cerebral micro-infarcts are known to occur in systemic lupus erythematosus (SLE), with or without lupus anticoagulant. We believe this is the etiology of the putaminal infarct in our case, and feel there is little evidence here for MS. Painful tonic spasms have been described previously in a patient with SLE, but with spinal cord and brainstem lesions. As a whole, painful tonic spasms respond remarkably to anti-convulsants (carbamazepine, phenytoin, phenobarbital) despite the failure to demonstrate an epileptogenic mechanism. Our patient likewise has her spasms controlled on carbamazepine.

The mechanism of painful tonic spasms may be either one of enhanced neuronal excitability or disinhibition. Hypocalcemia and hyperventilation-induced alkalosis could lead to the former. Regarding our case, perhaps a putaminal infarct “disinhibits” the pallidogranular part of the basal ganglia, allowing painful tonic spasms to occur. Such a disinhibition theory has been proposed to account for the hemichorea, hemiballismus and focal dystonia related to infarcts of the caudate, putamen and subthalamic nucleus. Evidence favoring this theory exists in the neurosurgical experience with ablative therapy for hemiballismus. Ballistic movements often were controlled by lesions placed in the globus pallidus or substantia nigra, as if these areas had been functioning in an uninhibited manner. Certainly the response to haloperidol suggests an “unopposed” dopaminergic etiology of hemiballismus. Painful tonic spasms appear unrelated to a dopaminergic mechanism, but may still, by another means, result from the “disinhibition” created by putaminal infarction.

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References


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**Figure 1.** CT showing small left putaminal infarction (arrow).
Ophthalmoplegia with Contralateral Hemiplegia. Occlusion of the Internal Carotid Artery Due to Thrombosis of an Intracavernous Aneurysm

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SUMMARY

A left ophthalmoplegia and right hemiplegia were due to thrombosis of an intracavernous aneurysm with occlusion of the left internal carotid artery. Ophthalmoplegia with contralateral hemiplegia is probably a rare syndrome. Thrombosis of and intracavernous aneurysm is probably a rare cause of occlusion of the internal carotid artery.

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OCCLOSIVE THROMBOSIS of the internal carotid artery (ICA) secondary to thrombosis in an aneurysm of the intracavernous ICA (ICICA) is probably very rare. It is not mentioned in current textbooks.1-3 We report one such case with a distinct clinical picture namely ipsilateral complete ophthalmoplegia with contralateral hemiplegia.

Case Report

A 65 year-old right handed woman was found at home seated on a chair with a right-sided weakness and speech disorder. She was admitted 24 hours later, on June 30, 1985.

She was alert with a right hemiplegia and aphasia. There were no gross sensory or visual field defect. She had a left ptosis and complete extrinsic and intrinsic paralysis of the left eye. There was no proptosis or chemosis. No bruit was heard over the neck and eyeballs. Cardiac examination was without significant abnormalities. BP: 160/80.

Unenhanced CT scan 1 hour after admission showed a left prerolandic low density area and a high density area in the left cavernous sinus region. On the same day angiography showed occlusion of the left ICA approximately 3 cms after its origin (fig. 1). A second CT with contrast and slices 6 mms thick showed a rounded image highly suggestive of a thrombosed large (23 mms diameter) aneurysm of the left ICICA (fig. 2). There was in addition erosion of the anterior clinoid process and of the lateral wall of the ethmoidal sinus (fig. 1 and 2). On July 4, a right carotid angiography showed 2 small berry aneurysms, at the bifurcation respectively of the right ICA and right middle cerebral artery.

The hemiparesis partially improved. The ophthalmoplegia was still complete when the patient was discharged to a Rehabilitation Unit on August 5, 1985.
Painful tonic spasms caused by putaminal infarction.
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