

abdominal quadrants on tendon percussion. MRI of the brain and cervical and thoracic spinal cord was normal. Serum B12 level was 142 pg/mL (>200), and methylmalonic acid and homocysteine levels were 20,000 nmol/L (90 to 279) and >50 μmol/L (0 to 11.3), respectively. Neurologic examination 5 months after parenteral B12 repletion (1000 μg/month) was normal, with no myoclonus.

**Discussion.** Typically, as in our patients, spinal segmental myoclonus is rhythmic (0.5 to 3 Hz), is confined to muscles innervated by a few spinal segments, persists during sleep, and, once established, tends to persist indefinitely.<sup>1-3</sup> Stimulus sensitivity (i.e., reflex percussion, posture change) sometimes occurs.<sup>4</sup> Below-normal vitamin B12 levels are common in 20 to 30% of people aged >65 years, but few have neurologic manifestations.<sup>5</sup> The classic subacute combined degeneration associated with B12 deficiency begins with vacuolar swelling of midthoracic spinal cord myelin layers. Subsequently, axons in the descending pyramidal and ascending posterior column tracts degenerate.

We have found no previous reports of segmental myoclonus in patients with B12 deficiency, although a case of generalized myoclonus was described.<sup>6</sup> The severe abdominal myoclonus at the T8 to T12 levels developed in the first patient at the same time as the characteristic symptoms of B12-associated disease, and myeloneuropathy and myoclonus showed improvement after treatment even though symptoms had been present for 4 years. The second patient made a complete recovery after B12 repletion, possibly because of earlier detection and treatment. We speculate that injury to the myelin in the dorsal and lateral columns or, alternatively, some more subtle result of B12 deficiency affecting neuronal excitability may have led to the development of reflex-sensitive spinal segmental myoclonus in our patients.

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## Central Tapia's syndrome ("matador's disease") caused by metastatic hemangiosarcoma

*M. Krasnianski, MD; S. Neudecker, MD; A. Schlüter, MD; U. Krause, MD; and M. Winterholler, MD*

The Spanish otorhinolaryngologist Antonio Garcia Tapia described in 1906 a lesion of the vagus and hypoglossus nerves with contralateral hemiplegia in a matador with bullhorn-induced wounds of the neck.<sup>1</sup> Almost all later publications described Tapia syndrome as a peripheral disease of the X and XII cranial nerves. The extremely rare central Tapia syndrome is characterized by hemiparesis, often with hemihypesthesia contralateral to the cranial nerves palsies. It is caused by nuclear lesions of the nucleus ambiguus of the vagal and glossopharyngeal nerve and the nucleus of hypoglossal nerve<sup>2</sup> combined with a lesion of the pyramidal tract.<sup>3</sup> We report a patient with Tapia syndrome caused by metastatic hemangiosarcoma in the medulla oblongata.

**Case report.** A 77-year-old man was referred with distinct weight loss, progressive weakness of his right leg, hoarseness, and swallowing and speech disturbances for 3 months.

On admission, he was alert and oriented. Speech was hoarse and dysarthric; the soft palate moved asymmetrically to the right side. He had dysphagia with absence of the left gag reflex. Laryngoscopy showed paralysis of the left vocal cord. Cough reflex was reduced. The tongue was deviated to the left on protrusion with prominent left-sided weakness. He had no nystagmus. There was no disturbance of facial movement and sensation or of trapezius and sternocleidomastoid muscles, and no Horner syndrome. The patient had a moderate right hemiparesis. The deep tendon reflexes were increased on the right with a right-sided Babinski sign. There was no ataxia. Pain and temperature perception was impaired on the right side of the body. Cranial CT on the first hospital day demonstrated a hyperdense area in the left medulla oblongata with enhancement after IV contrast agent (figure, A).

The axial gadolinium-enhanced T1-weighted brain MRI revealed a mass lesion similar to vascular structures in the left medulla oblongata (figure, B).

The patient died several days later from pulmonary embolism.

Autopsy revealed a hemorrhage in the left medulla oblongata (figure, C). On histologic examination a metastatic hemangiosarcoma with a prominent hemorrhage was found (figure, D and E);

there were many small hemangiosarcoma metastases in small intestine.

**Discussion.** The history of progressive weight loss and swallowing and speech disturbances in this patient was caused by a metastatic brainstem tumor. The lesions of vagal and hypoglossal nerves combined with contralateral hemiparesis and hemihypesthesia were caused by the involvement of the left lateral (nucleus ambiguus) and medial (nucleus of hypoglossal nerve, pyramidal tract, and medial lemniscus) medulla oblongata and were compatible with those of Tapia syndrome<sup>1</sup> (figure, F).

Cranial CT and MRI revealed a mass in the left medulla oblongata that was a hemangiosarcoma with secondary hemorrhage at autopsy.

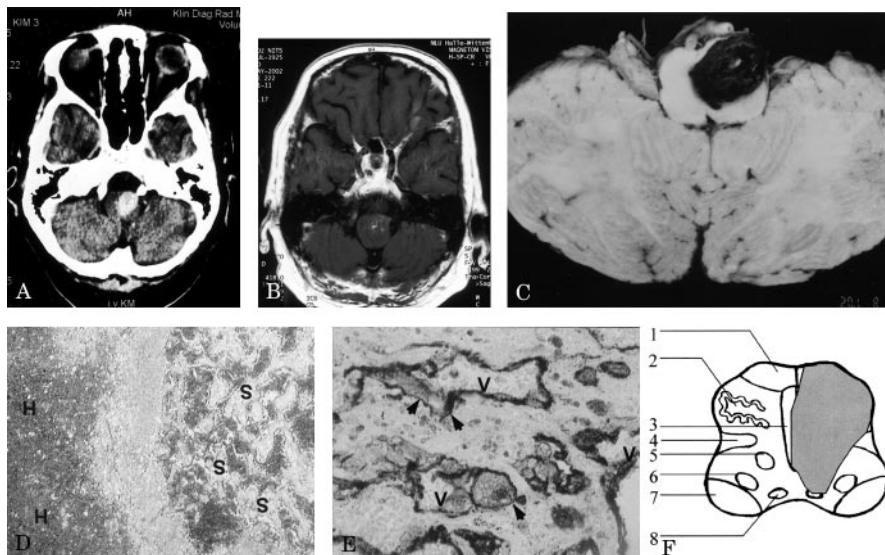
Tapia described this syndrome in a bullfighter who had been punctured by a bullhorn in the right side of his neck. He noted aphonia and difficulty swallowing his saliva. The bullfighter lost consciousness 2 hours later and awoke with a left hemiplegia that gradually improved. He also had paresis of the right side of the tongue and right vocal cord.<sup>1</sup>

The cause of the hemiplegia was not established in Tapia's case. Embolism from the traumatized carotid artery as well as dissection of the vertebral artery with brainstem infarction are possible. In two other patients of Tapia<sup>1</sup> and most publications about this syndrome, the peripheral variant of Tapia syndrome was described. The peripheral variant occurs in patients with trauma,<sup>1</sup> with tumor of the parotid,<sup>1</sup> and due to surgery.<sup>4</sup> In contrast, central Tapia syndrome, which is caused by lesions of the medulla oblongata, is very rare. Although the possibility of a central Tapia syndrome with paresis of X and XII nerves, contralateral hemiparesis, and possible hemihypesthesia in brainstem lesions was discussed, no etiology was established.<sup>2,3</sup> Only three patients with medial or mediolateral medullary infarction and the classic clinical features of central Tapia syndrome were described during the last 100 years: two patients with brainstem infarctions<sup>5,6</sup> and one with meningovascular lues.<sup>7</sup> Our case report is the first description of Tapia syndrome in a patient with brainstem tumor and adds this syndrome to the differential diagnosis of progressive swallowing dysfunction.

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*Figure. (A) Enhanced cranial CT shows a hyperdense area in the left medulla oblongata. (B) Axial gadolinium-enhanced T1-weighted MRI shows structures similar to vessels within the lesion. (C) Autopsy demonstrates a hemorrhage. (D) Histology reveals metastatic hemangiosarcoma (S) with prominent hemorrhage (H). (E) Higher magnification shows neoplastic vessels (V) filled with erythrocytes and lined with pleomorphic endothelial cells (arrows), which demonstrated a dark cytoplasmic stain in immunohistochemistry with CD 31 antibody. (F) Schematic drawing of the CT findings: 1. pyramidal tract; 2. nucleus olivaris; 3. lemniscus medialis; 4.*

*tractus spinothalamicus; 5. nucleus ambiguus; 6. nucleus and tractus spinalis nervi trigemini; 7. pedunculus cerebellaris inferior; 8. nucleus nervi hypoglossi.*

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## Autoimmune rippling muscle

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Muscle contraction is caused by electrophysiologic activity, whereas electrically silent myofiber shortening, physiologic contracture, occurs in varied situations including metabolic failure and calcium-ATPase deficiency. In one clinically fascinating form of physiologic contracture, rippling muscle (RM), mechanical stimuli cause electrically silent waves to propagate across a muscle.<sup>1,2</sup> RM can be dominantly inherited, as in families with caveolin mutations,<sup>3</sup> or can develop in patients with myasthenia gravis.<sup>4,5</sup> We report RM in a patient with autoimmune hemolytic anemia, but without either thymoma or a neuromuscular junction disorder. This patient demonstrates that acquired RM can occur in association with non-neurologic autoimmune disease.

**Case report.** A 60-year-old man of Irish and Polish descent had no neuromuscular symptoms, normal muscle bulk, normal strength, and no abnormal cramping until 2 years before evaluation, when he noted episodic localized tightening and rippling of muscles, sometimes producing involuntary limb twitches and impaired fine motor control. Physical exertion caused prolonged painful muscle stiffening with slow relaxation. His muscle mass and strength had increased over the preceding year. He denied sensory, cranial nerve, ventilatory, bladder, or bowel symptoms. Chronic lymphocytic leukemia (CLL) and type 2 diabetes mellitus were diagnosed 12 years before onset of muscle symptoms. Medications included insulin, lisinopril, naproxen, and gemfibrozil for several years. He denied drug abuse. His family history was negative for muscle disease, with no neuromuscular symptoms in his only sibling or either of his parents (his mother was 85 years old and his father died at 41 years); he had no children. General medical examination had normal results. Cranial nerve examina-

tion, sensory examination, reflexes, coordination, and gait were normal. Motor examination revealed normal muscle bulk, tone, and strength. Hand grip relaxed slowly, with no improvement after repeated trials. Pressing a thumb against a muscle for 2 seconds caused persistent visible mounding. Muscle percussion initiated a localized response followed by painful rippling that, in the thigh, traveled more than 10 cm. EMG showed no electrical activity as the wave of muscle movement propagated past the needle. EMG also showed normal insertional activity, with no fibrillations, fasciculations, myotonic potentials, or any other abnormal spontaneous activity; motor unit potential size and configuration, recruitment, and interference pattern were normal. Single fiber EMG of the extensor digitorum communis muscle had normal results: mean jitter of 20 pairs was 30  $\mu$ s (normal <33  $\mu$ s); range was 23 to 48  $\mu$ s (normal <55  $\mu$ s); fiber density was 1.6 (normal <2.0). Serum creatine kinase was normal. There was an immunoglobulin G kappa monoclonal gammopathy, a warm autoantibody to red blood cells, and a striated muscle antibody with a titer of 1:15,360. Tests for antineuronal antibodies types 1 and 2, Purkinje cell cytoplasmic antibody type 1, amphiphysin antibody, N- and P/Q-calcium channel antibodies, and AChR binding, blocking, and modulating antibodies were all negative. Abdominal and chest CT scans had normal results.

One year after the initial encounter, RM became more severe at the same time that the patient was diagnosed with autoimmune hemolytic anemia. Prednisone (prescribed for the hemolytic anemia) eliminated the muscle pain, reduced RM symptoms, and resulted in briefer, smaller amplitude percussion-induced rippling. Treatment of the anemia with chlorambucil and rituxan did not further improve his RM.

**Discussion.** Several features substantiate the autoimmune etiology of RM in this patient: subacute onset in midlife, no family history, presence of antistriated muscle antibodies, and RM severity

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