

evaluations as the narcolepsy group, including polysomnography and MSLT. Nevertheless, with any of the other neurologic disorders, a concomitant diagnosis of narcolepsy is relatively improbable based on their overall clinical presentations. Low CSF hypocretin levels were consistently found in all narcoleptic subjects. In addition, these levels were occasionally found prior to classic narcoleptic signs and symptoms. There may be a true, relatively independent diagnostic utility in measuring CSF hypocretin levels when narcolepsy is suspected in children.

From the Department of Pediatrics (Dr. Arie), Chiba Rosai Hospital, and Division of Neurology (Dr. Tanabe), Chiba Children's Hospital, Departments of Neuropsychiatry (Drs. Kanbayashi, Mishima, Hishikawa, and Shimizu) and Pediatrics (Dr. Sawaishi), Akita University School of Medicine, Department of Pediatrics (Dr. Kimura), Akita Red Cross Hospital, and Department of Pediatrics (Dr. Watanabe), Akita Nakadori General Hospital, Japan; and Center for Narcolepsy (Dr. Nishino), Stanford University, Palo Alto, CA.

Received July 18, 2003. Accepted in final form July 12, 2004.

Address correspondence and reprint requests to Dr. J. Arie, Department of Pediatrics, Chiba Rosai Hospital, 2-16 Tatsumidai-Higashi, Ichihara-shi, Chiba 290-0003, Japan; e-mail: junko-a@muf.biglobe.ne.jp

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Aortic dissection presenting with transient global amnesia-like symptoms

C. Gaul, MD; W. Dietrich, MD; B. Tomandl, PhD; B. Neundörfer, PhD; and F.J. Erbguth, PhD

Diagnostic criteria of transient global amnesia (TGA) are witnessed attacks, clear-cut anterograde amnesia during the attack, absent clouding of consciousness and loss of personal identity, no accompanying focal neurologic symptoms or epileptic features, resolution of attacks within 24 hours, and no recent head injury or active epilepsy.¹ For the etiology of TGA, four main hypotheses have been considered: TIA, epilepsy, migraine, and transient venous ischemia.¹⁻³ None of these hypotheses fully explains the mechanism of this episodic disease, but the accepted neuroanatomic correlate of TGA is the mediobasal temporal lobe and hippocampus. We present two patients with aortic dissection who provide evidence for an ischemic pathogenesis in TGA.

Case reports. **Patient 1.** A 47-year-old man was found confused and disoriented. On examination at admission, he asked repetitive questions and was alert but completely disoriented to time and place and only partially oriented to person. The cranial nerve examination showed only a slight anisocoria. The pronator drift test revealed a discrete motor deficit of the left side, accompanied by a mildly increased reflex activity. The cranial CT was unremarkable. The EEG revealed no epileptic discharges. Some hours later, the patient was reoriented with an amnesic gap for the attack's duration, and the neurologic deficit had completely resolved. Because of persistent hypotension, a chest radiograph was taken, which revealed a widening of the mediastinum. A CT scan of the chest and abdomen revealed a dissecting aneurysm (Stanford type A) of the aortic arch starting at the aortic valve, involving both carotid arteries and the left subclavian artery, and continuing into both iliac arteries (figure).

Patient 2. A 61-year-old woman was taken to hospital because of acute chest pain. On admission, she had retrograde amnesia for the past few hours, anterograde amnesia with inability to learn new facts, and repetitious questioning. Neurologic examination revealed a mild right facial paresis. Cranial CT was unremarkable. Five hours after onset of symptoms, she was reoriented with an amnesic gap. Because of the initial thoracic pain and our knowledge of the first reported patient, an aortic dissection was considered. Chest radiograph was normal, but a CT scan of the chest and abdomen revealed a dissection of the aorta (Stanford type A) starting at the aortic valve, involving all supra-aortic branches, and ending above the left renal artery.

Discussion. Patients with TGA can be distinguished into three groups: "pure TGA" patients who fulfill all diagnostic criteria; patients with probable epileptic amnesia; and patients with probable transient ischemic amnesia. The third group includes patients with additional neurologic deficits during the attack as in our patients.¹ Although we found no proof of ischemic lesions in the cranial CT in our patients, the minor neurologic deficits suggested cerebral ischemia. MRI including diffusion-weighted MRI

(DWI) would have been helpful to characterize the etiology, but both patients underwent immediate surgery without possibility for further diagnostics. We presume that an aortic dissection can cause a TGA subtype with ischemic etiology, which can be called transient ischemic amnesia. The underlying mechanism may be an embolic vascular occlusion in the posterior circulation, thus causing an embolic TIA with an unusual TGA-like TIA syndrome. The ischemic hypothesis in TGA was enforced by bitemporal hypoperfusion found in brain SPECT.⁴ However, patients with TGA have fewer thromboembolic risk factors and smaller risk of cerebral infarction compared with those with TIA.¹ The DWI findings provide conflicting results concerning a possible ischemic mechanism.⁵ In one study using DWI in 10 patients with TGA, 7 patients showed an elevated signal intensity in the left or in both temporomesial regions. This was interpreted as a hint of the possible etiologic role of spreading depression.⁶ One-third of patients with TGA also have migraine. The low recurrence rate of ~8% in TGA and the different age distribution are arguments against migraine as a pathogenic mechanism.¹ The weakest evidence is



Figure. Multiplanar reconstructions from spiral CT: multiple dissection membranes within the aortic arch are demonstrated with involvement of the supra-aortic branches (arrows).

found for an epileptic genesis because of the longer duration of TGA attacks and the absence of epileptic changes. Some authors favor a mechanism of retrograde venous congestion from the superior vena cava to cerebral veins.² Both of our patients had no history of migraine, epilepsy, or stroke but did have hypertension for several years.

As shown, aortic dissection is a possible cause of TGA. For any patient who does not fulfill the criteria of pure TGA, it is necessary to exclude other probable causes such as aortic dissection, especially if there is any history of chest pain, unexplained hypotension, or pathologic findings on chest radiographs.⁷

From the Department of Neurology (Dr. Gaul), Martin-Luther-University, Halle-Wittenberg; Departments of Neurology (Drs. Gaul, Dietrich, and Neundörfer) and Neuroradiology (Dr. Tomandl), Friedrich-Alexander-University, Erlangen-Nuremberg; and Department of Neurology (Drs. Dietrich and Erbguth), Nuremberg, Germany.

Received May 17, 2004. Accepted in final form August 10, 2004.

Address correspondence and reprint requests to Dr. Charly Gaul, Department of Neurology, Martin-Luther-University Halle-Wittenberg, Ernst-Grube-Strasse 40, 06097 Halle/Saale, Germany; e-mail: Charly.Gaul@gmx.de

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Bilateral carotid artery dissection with thyrotoxicosis

C.R. Campos, MD; M. Basso, MD; E.F. Evaristo, MD; F.I. Yamamoto, MD; and M. Scaff, PhD

Spontaneous internal carotid artery dissection (ICAD) is a major cause of stroke in young adults.¹ Environmental factors, such as minor trauma and mechanical stretching, may trigger the dissection, especially in patients with an underlying arteriopathy. Despite the direct vascular effects of thyroid hormones (THs),^{2,4} the predisposition for arterial dissection has not been identified in Graves disease (GD). We describe two women with bilateral ICAD and GD with fatal outcome, one with histopathologic study.

Case reports. **Patient 1.** A 45-year-old woman with irregular treatment for GD was admitted after 6 hours of right hemiparesis and aphasia. She had no vascular risk factors. On admission, her blood pressure was 130/80 mm Hg, and her pulse was 150 beats/min. She presented a mild nonfluent aphasia and right-sided hemiparesis. EKG showed sinus tachycardia. Brain CT showed an ischemic infarct in left middle cerebral artery (MCA) territory. Angiography detected bilateral irregular high-grade stenosis starting ~2 cm distal to the carotid bulbs and extending to the base of the skull, suggesting ICAD. Anticoagulation was started. After 24 hours, the patient deteriorated with left-sided hemiparesis and right hemianopia. MRI showed infarctions in the territory of the right anterior cerebral artery and both MCAs. One month

later, she deteriorated with decerebrate rigidity and died. Brain CT revealed diffuse edema and midline shift.

Patient 2. A 33-year-old woman with poorly controlled GD was admitted after 3 days of left hemiparesis and aphasia. She had no vascular risk factors. She was febrile; her blood pressure was 180/110 mm Hg, and her pulse was 146 beats/min. She had left-sided hemiplegia, right-sided hemiparesis, global hyperreflexia, left Babinski sign, right conjugated gaze palsy, and drowsiness. EKG showed sinus tachycardia. Brain CT revealed infarctions in both MCA territories. Angiography showed bilateral high-grade stenosis starting ~3 cm distal to the carotid bulbs and extending to the base of the skull, suggesting ICAD. Anticoagulation was started. She deteriorated during the next 24 hours with decerebrate rigidity. Brain CT showed marked infarctions in both internal carotid artery territories, edema, and midline shift. She died 2 days later.

Postmortem examination macroscopic findings were diffuse and small hemorrhagic lesions at pons, severe infarctions in both MCA territories, and edema with bilateral uncal and tonsillar herniation. The circle of Willis and its major branches were unremarkable. Both internal carotid arteries presented multifocal media degeneration with intracellular and extracellular vacuoles, sometimes coalescing to form cysts, and areas of marked wall thinning. There were also deposits of Alcian blue/periodic acid–Schiff-positive substances in the degenerating media. The intima was focally thickened, and the internal elastic lamina was absent in some areas and reduplicated and distorted in others (figure).

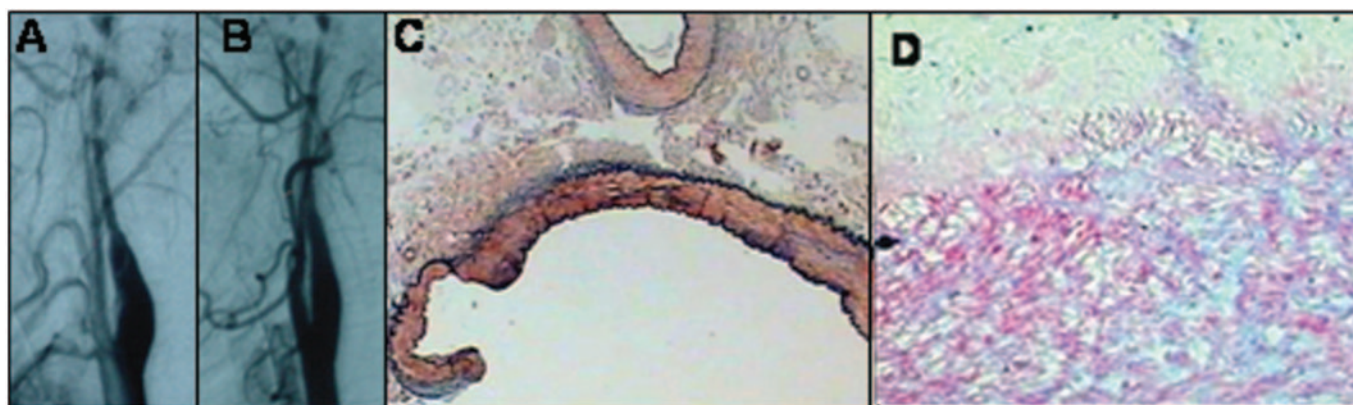


Figure. Angiography showing right (A) and left (B) internal carotid artery dissection. (C) Cross-section of the right internal carotid artery. There is focal tunica media thinning and an area of duplication and distortion of the internal elastic lamina on the left edge of the artery (Elastica–Verhoff stain, $\times 25$). (D) Cross-section of the left internal carotid artery. Tunica media showing vacuolar degeneration with intracellular and extracellular vacuoles and deposits of Alcian blue/periodic acid–Schiff-positive substances ($\times 400$).

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Neurology 2004;63;2442-2443

DOI 10.1212/01.WNL.0000148581.00912.1A

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