Case Report

Crossed Ataxia
A Case Report and a Diffusion Tensor Imaging Tractography Study

Julien Bally, MD; Pierre Mégevand, MD, PhD; Duy Nguyen, MD; Theodor Landis, MD; Cristina Granziera, MD, PhD

Background and Purpose—Ever since the seminal description of ataxic hemiparesis contralateral to a pontine lesion by Miller-Fisher, the question of why contralesional crossing pontocerebellar fibers do not more frequently produce ipsilesional hemiataxia was raised. The few cases of “quadrataxic hemiparesis” or bilateral leg ataxia remain exceptions.

Summary of Case—We report an even more unusual variant, namely “crossed ataxia” of the contralesional arm and the ipsilesional leg subsequent to an anteromedial pontine ischemic stroke.

Conclusions—MRI diffusion tensor imaging tractography shows that caudal contralesional crossing pontocerebellar fibers (those for the leg) travel through the lesion, whereas more rostral fibers (those for the arm) are spared. (Stroke. 2011; 42:e571-e573.)

Key Words: ataxia ■ brainstem stroke ■ clinical neurology ■ diffusion-weighted imaging ■ lacunar infarcts ■ MRI ■ tractography

Miller-Fisher first described “ataxic hemiparesis” (AH) in 1965 and redefined and named it in 1978.1,2 AH can have different etiologies (lacunar, embolic) and different locations (mainly internal capsule, corona radiata, thalamus, pons).3-4 The syndrome associates contralesional limb paresis and ataxia, the latter being sometimes initially hidden by the former and revealed as strength recovers. AH can be associated with dysarthria or with cranial nerve involvement. It generally involves both the arm and leg, but cases of AH affecting a single limb have been reported.3,5,6 AH due to pontine lesions is thought to be caused by the involvement of both the corticospinal tract and the corticopontocerebellar pathway, where corticopontine fibers synapse on neurons in the pontine nuclei that cross the midline and reach the cerebellum through the contralateral middle cerebellar peduncle. An open question is why ataxia ipsilateral to the lesion is not more frequent, because fibers crossing the midline from the contralesional pontine nuclei should also be involved in the lesion. This question has already been raised by Miller-Fisher who states: “Speculations on the mechanism underlying the contralateral cerebellar signs is limited to the suggestion that either the pontine nuclei sending fibers to the opposite cerebellar hemispheres are damaged or crossing fibers from the opposite pontine nuclei are interrupted. In either case it is not clear why the cerebellar signs are not bilateral.”2

Although rare, bilateral cerebellar signs have been reported as “quadrataxic hemiparesis”7 or bilateral leg ataxia.8 We report for the first time an even more unusual variant, namely “crossed ataxia” of the contralesional arm and the ipsilesional leg subsequent to an anteromedial pontine ischemic stroke situated at the caudal part of the middle third of the pons; in addition, we show that the pathophysiology of this new syndrome can be clarified using diffusion tensor imaging tractography.

Patients and Methods

This 53-year old male insurance manager without known cardiovascular risk factors presented with acute dysarthria and clumsiness of the right arm. At admission the patient was hypertensive and neurological examination showed marked ataxia as well as mild, predominantly distal paresis of the right arm and marked ataxia without paresis of the left leg. The right leg and left arm were unaffected. In addition, there was severe dysarthria without facial paresis. There was no nystagmus, no oculomotor deficit, no sensory abnormality of the face, and the rest of the cranial nerves examination was normal. Deep tendon reflexes were symmetrically normal, cutaneous plantar response was flexor on both sides, and touch and vibration sense were normal. The general examination was remarkable only for marked hypertension (blood pressure 215/110 mm Hg).

Brain MRI showed a left anteromedial pontine ischemic stroke situated at the caudal part of the middle third of the pons extending dorsally from the most ventral part of the pons (Figures 1 and 2). CT angiography as well as extra- and transcranial ultrasound examination found diffuse athromatosis without any significant vascular stenosis. Forty-eight-hour Holter monitoring did not reveal atrial fibrillation. Transthoracic echocardiography disclosed slight left atrial dilatation.
Three weeks after the stroke, an MRI including T1-/T2-weighted images and diffusion tensor imaging was acquired. Diffusion tensor imaging tractography was performed using a streamline based algorithm of the Trackvis software (www.trackvis.org). Tractography results showed that the caudal crossing fibers (the lower fibers in red on the picture) of the middle cerebellar peduncle pass through the ischemic lesion, whereas the rostral fibers travel above the lesion (Figure 3).

Discussion

The interest of the present case is 2-fold: (1) it is the first description of “crossed ataxia”; and (2) it shows that diffusion tensor imaging tractography may help in understanding the functional anatomy of the clinical picture, particularly that of the ipsilesional ataxia, whose origin is often discussed.

To explain the absence of ipsilesional ataxia, it has been hypothesized that because the crossing fibers run relatively widespread on their way to the middle cerebellar peduncle, a small lesion would affect only part of them, intact fibers providing compensation.9 Following this idea, it has been stated that only massive unilateral lesions, interrupting all crossing fibers, could cause ataxia ipsilateral to the lesion (associated with important contralateral paresis).5 Others have explained the absence of ipsilesional ataxia by the fact that the crossing fibers lead their way obliquely in the rostral–caudal plane, thus being spared by a lesion at a single level of the pons.10 However, cases of AH combined with ataxia of the nonparetic leg8 or arm and leg (so-called “quadrataxic hemiparesis”)7 have been reported in patients with a single lesion that did not reach over the midline.

The diffusion tensor imaging tractography results obtained from the reported case show that only the caudal part of the pontocerebellar axons, which have already decussated and which originate in the contralateral pontine nuclei, travel through the lesion, whereas the more cranial part of this fiber bundle is spared because it passes above the stroke area (Figure 3). Because the fibers forming the homolateral middle cerebellar peduncle, which has a broad rostrocaudal extension throughout the pons, are somatotopically organized,11,12 we hypothesize that the relatively small lesion in our case interrupted only fibers with a destination aimed at the control of the ipsilateral leg and spared those controlling the arm movements.
Source of Funding
This work has been supported by the Swiss National Science Foundation Grants 320030_132967/1 to T.L. and PZ00P3_131914/1 and the Stoicescu fellowship to C.G.

Disclosures
None.

References
Crossed Ataxia: A Case Report and a Diffusion Tensor Imaging Tractography Study
Julien Bally, Pierre Mégevand, Duy Nguyen, Theodor Landis and Cristina Granziera

Stroke. 2011;42:e571-e573; originally published online September 29, 2011;
doi: 10.1161/STROKEAHA.111.623553
Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2011 American Heart Association, Inc. All rights reserved.
Print ISSN: 0039-2499. Online ISSN: 1524-4628

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://stroke.ahajournals.org/content/42/11/e571

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Stroke can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Stroke is online at:
http://stroke.ahajournals.org//subscriptions/