Paroxysmal dysarthria and ataxia after midbrain infarction

Matsui et al. report a patient with attacks of paroxysmal dysarthria and ataxia after midbrain infarction. SPECT showed hypoperfusion in the parietal lobe contralateral to hemiataxia during the period of paroxysmal attacks. See page 345

Finger-nose test with penlight attached to tip of a finger, during an attack (A) and between attacks (B).

Acquired episodic ataxia and dysarthria

Commentary by Joanna C. Jen, MD, PhD

The participation of the cerebellum in various feedback circuits involving sensorimotor pathways ensures smooth and precise voluntary and involuntary movement. The superior cerebellar peduncle carries efferent cerebellar projections to the red nucleus and thalamus, as well as the reticular formation. Ascending fibers project to the cortex, from which corticotopontine fibers originate to descend to the pontine nuclei, with postsynaptic pontocerebellar fibers carried by the middle cerebellar peduncle, thus closing an important feedback loop between the cerebellum and the cortex. Another feedback loop consists of projections from the red nucleus to the inferior olive, which projects to the cerebellum, which in turn projects back to the red nucleus, thereby indirectly modulating descending rubrospinal and reticulospinal tracts. It is well known that disruption at different levels of these circuits can lead to ataxia. Depimelinating lesions in multiple sclerosis can cause paroxysmal ataxia (presumably from ephaptic transmission of demyelinated axons), yet reports of ische-mic subcortical lesions causing paroxysmal ataxia have been rare.

The report from Matsui et al. describes a patient with a left midbrain infarct who, 6 weeks later, developed paroxysmal right arm ataxia and dysarthria with increasing frequency. The authors observed left parietal hypoperfusion by SPECT, which the authors correlated with frequent spells and that improved (but did not normalize) when the patient became free of symptoms after taking phenytoin. The authors hypothesized that sprouting and ephaptic transmission underlies paroxysmal ataxia in this case.

The brief and recurrent nature of the patient’s symptoms in this report is more similar to EA1 than EA2. EA1 symptoms are generally not responsive to medications. EA2 can be dramatically responsive to acetazolamide, and a recent report described three EA2 patients who responded to 4-aminopyridine (4-AP), a blocker of potassium channel. Of note, acetazolamide has been reported to be effective in treating paroxysmal symptoms in multiple sclerosis presumed to arise from ephatic transmission of partially demyelinated axons. Since demyelination affects the distribution of potassium channels, it may be interesting to look into how MS patients with paroxysmal symptoms may respond 4-AP.

References
This information is current as of July 26, 2004

<table>
<thead>
<tr>
<th>Updated Information &amp; Services</th>
<th>including high resolution figures, can be found at: <a href="http://www.neurology.org/content/63/2/197.full.html">http://www.neurology.org/content/63/2/197.full.html</a></th>
</tr>
</thead>
<tbody>
<tr>
<td>References</td>
<td>This article cites 4 articles, 3 of which you can access for free at: <a href="http://www.neurology.org/content/63/2/197.full.html##ref-list-1">http://www.neurology.org/content/63/2/197.full.html##ref-list-1</a></td>
</tr>
<tr>
<td>Permissions &amp; Licensing</td>
<td>Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: <a href="http://www.neurology.org/misc/about.xhtml#permissions">http://www.neurology.org/misc/about.xhtml#permissions</a></td>
</tr>
<tr>
<td>Reprints</td>
<td>Information about ordering reprints can be found online: <a href="http://www.neurology.org/misc/addir.xhtml#reprintsus">http://www.neurology.org/misc/addir.xhtml#reprintsus</a></td>
</tr>
</tbody>
</table>