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RAEDER SYNDROME PRODUCED BY EXTENSION OF CHRONIC INFLAMMATION TO THE INTERNAL CAROTID ARTERY

Raeder paratrigeminal syndrome (painful Horner syndrome with trigeminal nerve involvement) is a poorly recognized complication of multiple disorders including carotid aneurysm, cancer, herpes zoster, maxillary sinusitis, and chronic otitis media. Although brain MRI has revealed carotid artery narrowing,¹ dissection,^{2,3} and aneurysm,⁴ Raeder syndrome produced by inflammatory disease surrounding the carotid artery has not been verified by brain imaging until now.

Case reports. A 51-year-old man noted a sensation of right ear fullness and vertigo; neurologic examination and brain MRI scan (figure, A) was normal. Three months later, he developed right-sided otitis media, right facial numbness, stabbing headaches, and intermittent drooping of the right eyelid. Examination revealed a partial Horner syndrome (right-sided ptosis and miosis without anhidrosis) and right-sided trigeminal-distribution hypalgesia. CT angiogram showed no evidence of carotid artery dissection. Brain bone window CT (figure, B), proton-density MRI (figure, C), and contrast-enhanced T1-weighted MRI (figure, D) showed complex fluid collection and opacification in the mastoid air cells consistent with mastoiditis extending to the area around the internal carotid artery. Blood cultures were negative. The CSF was normal. The patient was treated with IV vancomycin 1 g twice daily for 3 days and piperacillin/tazobactam 3.375 g 4 times daily for 3 days, followed by oral doxycycline 100 mg twice daily and levofloxacin 720 mg daily for 5 weeks, as well as prednisone 60 mg for 3 days, 40 mg for 3 days, 20 mg for 3 days, and 10 mg for 3 days. Three months later, headaches and trigeminal-distribution hypalgesia had resolved, although the partial Horner syndrome was still evident. Brain imaging revealed near-complete resolution of inflammation in the mastoid air cells and around the carotid artery (figure, E and F).

A 78-year-old woman with long-standing subcortical arteriosclerotic encephalopathy (Binswanger disease) complained of pain over the left zygoma for

7 years. Neurologic examination revealed severe dementia and gait ataxia. There was a partial left Horner syndrome and hypalgesia in the maxillary distribution of the left trigeminal nerve. In addition to brain atrophy and ischemic small artery disease, an air-fluid level and inflammatory changes consistent with a left maxillary sinusitis was revealed by brain CT. The presence of a pacemaker precluded MRI. Treatment with clindamycin 150 mg 4 times daily for 3 months led to pain relief. Examination by a neurologist and an ophthalmologist 10 weeks after antibiotic treatment indicated loss of facial hypalgesia and complete resolution of the Horner syndrome.

Discussion. Herein, brain imaging in 2 cases of Raeder syndrome revealed chronic mastoiditis and sinusitis, respectively. The outcomes after antibiotic treatment were favorable, with both patients experiencing considerable relief of pain and loss of facial hypalgesia. Furthermore, the Horner syndrome completely disappeared in the second patient although not in the first patient, perhaps due to permanent damage to sympathetic nerve fibers around the carotid artery. Nevertheless, in the first patient, the fortuitous requests for brain MRI before the onset of Raeder syndrome, during acute disease and after antibiotic treatment, revealed the development of inflammation and extension of chronic mastoiditis to the area around the carotid artery, with resolution after treatment. Although MRI could not be performed due to a pacemaker in the second patient, the case is presented to emphasize that antibiotic treatment of chronic sinusitis produced relief of pain along with complete resolution of facial hypalgesia and Horner syndrome.

In 1924, Raeder⁵ reported the relationship between “paralysis of the oculopupillary sympathetic nerves and pain in the trigeminal nerve distribution implicating the paratrigeminal area of the middle fossa of the cranium.” Although Horner⁶ had described the features of involvement of the cervical sympathetic nerves in 1869, Raeder recognized that the clinical syndrome of Horner may result from a lesion anywhere along the course of these sympathetic fibers, and that the completeness of the syn-

Figure Raeder paratrigeminal syndrome: Brain imaging before and during acute disease and after antibiotic treatment

In all images (A-F), the long arrow points to the carotid artery, short arrows indicate mastoid air cells, and arrowheads indicate areas around the carotid artery. Postganglionic sympathetic nerve fibers travel in the adventitia of the internal carotid artery (not resolvable by imaging) before exiting more cephalad between Dorelo canal and Meckle cave and become associated with the first division of the trigeminal nerve in the cavernous sinus. (A) Three months before acute disease, a T2-weighted MRI shows normal mastoid air cells and no abnormal signal around the carotid artery. (B) During acute disease, high-resolution temporal bone CT shows new soft tissue opacification in the mastoid air cells and around the carotid artery. (C) During acute disease, proton density MRI shows a heterogeneous signal in the mastoid air cells and around the carotid artery, consistent with inflammation. (D) During acute disease, postcontrast T1-weighted MRI shows heterogeneous enhancement mixed with low T1 signal in the mastoid air cells and around the carotid artery, consistent with inflammation. (E) Three months after antibiotic therapy, repeat proton density MRI shows near complete resolution of inflammatory changes in the mastoid air cells and around the carotid artery compared to acute disease (C). (F) Three months after antibiotic therapy, repeat post-contrast T1-weighted MRI shows near complete resolution of inflammatory changes in the mastoid air cells and around the carotid artery compared to acute disease (D).

drome varies with the location of the lesions. For example, in our patients, the absence of anhidrosis is because sympathetic fibers for sweat follow the external carotid artery while those controlling the dilator papillae and superior palpebral muscle follow the internal carotid. The diverse etiologies and functional anatomy in Raeder syndrome are well-described.⁷ There is increased awareness of disease intrinsic to the carotid artery as a cause of Raeder syndrome. In particular, carotid dissection,^{2,3} aneurysm,⁴ and stenosis¹ have been verified by imaging. Yet a PubMed search back to 1967 revealed no reports of Raeder syndrome caused by chronic inflammatory sinus disease that were supported by brain imaging. Recognition that periarterial inflammation can cause Raeder syndrome should alert the clinician to request relevant imaging studies.

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SPONTANEOUS INTRACRANIAL HYPOTENSION AS AN INCIDENTAL FINDING ON MRI

Spontaneous intracranial hypotension due to a spinal CSF leak is an enigmatic cause of not only headache but also coma, parkinsonism, and dementia.¹ We report a group of patients who were found to have the typical MRI findings of spontaneous intracranial hypotension without any clinical symptoms.

Case reports. *Case 1.* A 44-year-old man requested screening for intracranial aneurysms after his sister had an aneurysmal subarachnoid hemorrhage. MRI showed diffuse pachymeningeal enhancement and brain sagging (figure). CT myelography showed an opening pressure of 8 cm H₂O and an extensive tho-

racic CSF leak (figure). The patient denied any current or previous neurologic symptoms. He had never undergone a spinal tap and there was no history of significant trauma. Neurologic examination was normal. No treatment was advised, but he requested an epidural blood patch because of the demonstration of the extensive CSF leak and a published report of “more clear thinking” following epidural blood patching.² No improvement was noted following a lumbar epidural blood patch using 15 mL of blood. His postprocedural back pain resolved over a 10-day period. A 1-year follow-up MRI is pending.

Case 2. A 65-year-old woman with history of breast cancer was involved in a motor vehicle crash sustaining isolated facial trauma. Head CT showed an intracranial mass. MRI showed a parietal homogeneously enhancing dural-based mass, consistent with meningioma. No other abnormalities were noted. CT of the chest, abdomen, and pelvis was normal. MRI 6 months later showed the appearance of diffuse pachymeningeal enhancement (figure). The patient denied any current or previous neurologic symptoms. She had never undergone a spinal tap. Neurologic examination was normal. Magnetic resonance myelography showed numerous large thoracic spinal meningeal diverticula (figure), common findings in spontaneous intracranial hypotension. No treatment was advised. MRIs performed 6 and 12 months later showed gradual resolution of the pachymeningeal enhancement.

Case 3. A 72-year-old woman noted transient monocular blindness. Carotid ultrasonography showed a 90% ipsilateral extracranial internal carotid artery stenosis. MRI showed diffuse pachymeningeal enhancement and sagging of the brain (figure). The patient denied any current or previous neurologic symptoms, except for the amaurosis fugax. She had never undergone a spinal tap and there was no history of significant trauma. Neurologic examination was normal. No treatment was advised for the spontaneous intracranial hypotension. The patient underwent an uneventful carotid endarterectomy. She declined any spinal imaging or follow-up MRIs.

Discussion. We report 3 patients with spontaneous intracranial hypotension found as an incidental finding on MRI. To our knowledge, this is a previously unreported finding. Two of these patients had no neurologic symptoms and the third patient only had amaurosis fugax. This is a symptom not related to spontaneous intracranial hypotension.

In previous studies of incidental findings on brain MRI, spontaneous intracranial hypotension has not been reported.^{3–5} It should be noted that one of the most characteristic findings of spontaneous intracra-

Figure Spontaneous intracranial hypotension as an incidental finding on MRI

Case 1: axial T1-weighted gadolinium-enhanced MRI shows pachymeningeal enhancement (A) and CT myelography shows an extrathecal thoracic CSF collection (B). Case 2: coronal T1-weighted gadolinium-enhanced MRI shows a dural-based left parietal mass and pachymeningeal enhancement (C) and thoracic magnetic resonance myelogram shows multiple large meningeal diverticula (D). Case 3: axial T1-weighted gadolinium-enhanced MRI shows pachymeningeal enhancement (E) and sagittal T1-weighted MRI shows sagging of the brain (F).

nial hypotension on MRI is pachymeningeal enhancement while in most studies no IV gadolinium was administered. Sagging of the brain, a pathognomonic feature of spontaneous intracranial hypotension, can mimic a Chiari type 1 malformation and it cannot be excluded that some individuals who were diagnosed with an incidental Chiari malformation in fact had spontaneous intracranial hypotension.⁶ More likely explanations for the absence of findings of spontaneous intracranial hypotension in studies of incidental findings on MRI are that 1) the MRI findings of spontaneous intracranial hypotension usually are transient and 2) the incidence of spontaneous intracranial hypotension is low. It is of interest to note in this context that one of our patients had a family history of intracranial aneurysm. Intracranial aneurysms are more common in patients with spontaneous intracranial hypotension than in the general population and this is likely related to a generalized—probably genetically determined—connective tissue disorder,⁷ making this individual possibly more likely to develop spontaneous intracranial hypotension.

The finding of asymptomatic spontaneous intracranial hypotension further corroborates previously made observations that there is no good correlation between findings on brain MRI and severity of symptoms due to spontaneous intracranial hypotension. It has been well-established that brain MRI findings may resolve over time as the headache of spontaneous intracranial hypotension becomes chronic, although this is regularly associated with the headache losing its positional features. Conversely, symptoms of spontaneous intracranial hypotension may resolve completely over time while brain MRI findings persist. Furthermore, up to 20% of patients with spontaneous intracranial hypotension never develop any abnormalities on MRI of the brain.¹ The

reasons for this individual variability remain to be determined.

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