Sleep, 20(8):630–631

Short Note

MRI Findings in Narcolepsy

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Summary: The neuropathology of narcolepsy is unknown. Recently, Plazzi et al. (1) reported magnetic resonance imaging (MRI) abnormalities in the pontine tegmentum of three patients with long-standing idiopathic narcolepsy. Considering the localization of the neuroradiological findings in the pontine reticular formation, where rapid eye movement (REM) sleep is generated, the authors suggested a causal relationship between narcolepsy and MRI abnormalities.

Frey and Heiserman, however, found pontine MRI abnormalities in only two of 12 patients with narcolepsy, both of whom had long-standing hypertension (2). Pullicino et al. noted similar pontine MRI abnormalities in patients with subcortical arteriosclerotic encephalopathy-like ischemic rarefaction of the pons (3). Thus, the changes noted by Plazzi et al. may have been caused by small-vessel disease rather than narcolepsy. To assess whether altered pontine MRI signals are a regular feature of idiopathic narcolepsy, we selected randomly from our database seven patients with narcolepsy with cataplexy. Of these seven, three agreed to have brain MRIs; their cases are described below. None had pontine MRI abnormalities. **Key Words:** Narcolepsy—MRI—Sleep—Sleep disorders—Brain imaging.

CASE STUDIES

Patient 1

This 59-year-old teacher developed overwhelming excessive daytime sleepiness (EDS) in high school. Cataplexy developed at age 40, associated with laughter or anger. She also had sleep paralysis and hypnagogic hallucinations. Neurologic examination at age 49 was normal. Polysomnographic findings included a sleep onset REM period (SOREMP) and a few periodic leg movements. The multiple sleep latency test (MSLT) was consistent with severe EDS (mean sleep latency = 2.4 minutes) and showed three SOREMPs. Human leukocyte antigens (HLA), DR2 and DQ1, were present. Treatment with scheduled naps, pemoline (150 mg/day), and methylphenidate (5–10 mg/day as needed) produced almost complete resolution of EDS and cataplexy. Brain MRI with special emphasis on the brainstem revealed a single 2-mm supratentorial nonspecific hyperintense signal abnormality.

Patient 2

This 62-year-old nurse developed EDS at age 45 along with cataplexy, sleep paralysis, and hypnagogic hallucinations. At the time of evaluation at age 55 in 1989, neurologic examination was normal. Polysomnographic findings included a SOREMP, obstructive sleep apnea (apnea–hypopnea index = 30), and periodic leg movements (34/hour). The MSLT was consistent with severe EDS (mean sleep latency = 2.7 minutes) and showed three SOREMPs. The patient was positive for HLA-DR2. Treatment with scheduled naps, continuous positive airway pressure at a pressure of 6 cm of water, methylphenidate (60 mg/day), and protriptyline (40 mg/day) led to improvement of EDS and cataplexy. Brain MRI with special emphasis on the brainstem was normal.

Patient 3

At age 37, this 57-year-old housewife developed EDS, severe cataplexy (usually following excitement

Accepted for publication June 1997.
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or anger), sleep paralysis, and hypnagogic hallucinations. Neurologic examination at age 49 was normal. Polysomnographic findings included very mild sleep apnea (apnea–hypopnea index = 11). Treatment with scheduled naps and combinations of methylphenidate with protriptyline, imipramine, fluoxetine, and clomipramine produced moderate improvement of EDS but only partial relief of cataplexy. Brain MRI with special emphasis on the brainstem was normal.

DISCUSSION

These patients are clinically similar to those reported by Plazzi et al. (1). The classical clinical tetrad was present in all three patients, and typical MSLT and HLA findings were found in two of them. Furthermore, all three patients were in their fifties and had long-standing narcolepsy; however, none had pontine MRI abnormalities.

Recently described pontine vascular changes may be the cause of the abnormal MRI findings reported by Plazzi et al. (1). Pullicino et al. (3) found pontine MRI hyperintensities in 19% of 85 subjects above the age of 60 (3). As in the patients reported by Plazzi et al. (1), these signal abnormalities were usually patchy and bilateral and localized to the paramedian ventrotegmental junction of the rostral pons. The pontine changes were clinically asymptomatic, were accompanied by supratentorial MRI abnormalities in only one-third of the patients, and were often associated with vascular risk factors.

Our results, along with those of Frey and Heiserman (2), suggest that pontine MRI abnormalities in elderly patients with idiopathic narcolepsy are not common and, when present, are probably not causally related to the sleep disorder. We believe that neuroradiological studies should be considered only when the history and examination suggest that narcolepsy may be related to a structural brain lesion (4,5).

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