

gray and white matter, consistent with a diagnosis of AxD. An autopsy on patient 2 showed severe atrophy of medulla and spinal cord with Rosenthal fiber deposition. (Graff-Radford J, Schwartz K, Gavrilova RH, et al. Neuroimaging and clinical features in type II (late-onset) Alexander disease. **Neurology** 2014 Jan 7;82(1):49-56).

COMMENTARY. Alexander disease is characterized as infantile, juvenile, and adult forms or as type I with early-onset or type II of late-onset. MRI features of infantile type I consist of frontal white matter changes, a periventricular rim, and signal abnormality involving the basal ganglia, thalamus, and brainstem. The imaging features of type II late-onset AxD are different from those of type I and show predominant brainstem involvement with significant atrophy of the medulla, middle cerebellar peduncle, and spinal cord. The differential diagnosis of involvement of the middle cerebellar peduncle include AxD type II, spinocerebellar ataxia, Wilson disease, liver cirrhosis, adrenoleukodystrophy, and neoplasm. Palatal tremor (palatal myoclonus) is suggestive of late-onset AxD [1] but was rare in the Mayo Clinic series.

#### References.

1. Schwankhaus JD, et al. *Neurology*. 1995 Dec;45(12):2266-71.

## SLEEP DISORDERS

### EVOLUTION OF CHILDHOOD NARCOLEPSY AND CATAPLEXY

Investigators at University of Bologna and other centers in Italy, Finland, USA, and UK, performed clinical, polysomnographic, and cataplexy-video assessments at diagnosis (mean age of 10 +/- 3 years) and after a median follow-up of 3 years. At diagnosis children with narcolepsy with cataplexy showed an increase of sleep time during the 24 h; at follow-up, sleep time and nocturnal sleep latency shortened. Hypotonic phenomena decreased over time and were age dependent. At onset, childhood narcolepsy with cataplexy is characterized by abrupt increase of total sleep over the 24h, generalized hypotonia and motor overactivity, and hypocretin 1 deficiency. With time, cataplexy evolves into classic presentation (brief muscle weakness episodes triggered by emotions), whereas total sleep time across 24h decreases, returning to more age-appropriate levels. (Pizza F, Franceschini C, et al. Clinical and polysomnographic course of childhood narcolepsy with cataplexy. **Brain** 2013 Dec;136(Pt 12):3787-95).

COMMENTARY. Narcolepsy with cataplexy is characterized by abrupt onset and sudden weight gain that partially remits over time. Childhood onset narcolepsy/cataplexy is more than just a sleep disorder [1]. It is reported in 3 cases in association with paraspinal neuroblastoma [2], and is linked to H1N1 vaccination [3].

#### References.

1. Kotagal S, Kumar S. *Sleep*. 2013 Feb 1;36(2):161-2.
2. Sinsico C, et al. *Pediatr Neurol*. 2013 Jul;49(1):64-7.
3. Thebault S, et al. *Curr Opin Pulm Med*. 2013 Nov;19(6):587-93.