

Dog	Sex	Age at onset (months)	Owner reported clinical signs	Sleep disturbances	Other history	General physical examination	Neurological examination abnormalities	Clinical pathology performed	Advanced imaging	Response to medication	Alive / Dead (post-mortem examination)
1	F	12	Excessive response to loud noises; heightened anxiety; inappropriate barking; ball-guarding	From 12 months; progressive	Previous reactive lymphadenopathy of unknown etiology	Small in stature (comparison to a littermate-sibling of same sex, neuter status and similar body condition; 15.7kg vs. 21.2kg)	At 32 months (progressive at later visits) fine head tremor, crossed extensors in all limbs, brisk tendon reflexes in all limbs. At 37 months (progressive at later visits) stiff stilted gait with hypermetria in all limbs. At 47 months developed keratoconjunctivitis sicca with bilateral corneal ulceration	Hematology, serum biochemistry, CSF analysis, infectious disease screening (<i>Toxoplasma gondii</i> ; <i>Neospora caninum</i> ; <i>canine distemper virus</i>), bile acid stimulation test, ACTH stimulation test, serum cobalamin, urine organic metabolites	MRI (n=3); progressive caudate nucleus cavitation	Imepitoin (12-30mg/kg orally twice daily) – equivocal response reported by owners Levetiracetam (15mg/kg orally three times daily) - equivocal response reported by owners Acetaminophen (12mg/kg orally twice daily) – no perceived response.	Euthanized at 48 months (Yes)
2	MN	7	Excessive response to loud noises; separation anxiety; minimal tail movement; reluctant to exercise at 33 months; ataxia at 60 months; fecal and urinary incontinence; food obsession	Lifelong; progressive	Lipoma	Grade 2/6 left systolic heart murmur and relative bradycardia (ECG, echocardiography unremarkable)	At 62 months manic mentation; ataxic in all limbs (hind > fore); hypermetria of the forelimbs; decreased hopping in all limbs; proprioceptive deficits in the hindlimbs; crossed extensor of the hind limbs; bilateral mild reduction in menace	Hematology, serum biochemistry, CSF analysis, infectious disease screening (<i>Toxoplasma gondii</i> ; <i>Neospora caninum</i> ; <i>canine distemper virus</i>), plasma ammonia, serum thiamine, urine organic metabolites	MRI (n=1); marked caudate nucleus cavitation	Prednisolone (maximum 0.54mg/kg twice daily orally then tapered to 0.54mg/kg every other day) – variable, partial improvement in ataxia	Alive at 95 months (n/a)
3	M	12	Altered interaction with stimulae (people / noises); swam vertically in water	From 32 months; progressive	SRMA at 10 months (relapse at 32 months)	Unremarkable	At 32 months depressed mentation; dysmetria in all limbs (fore > hind); mild bilateral lateral strabismus	Hematology, serum biochemistry, serum thiamine, CSF analysis	MRI (n=1); marked caudate nucleus cavitation	Prednisolone (2mg/kg/day then tapered) – behavior worse on treatment	Alive at 49 months (n/a)
4	M	24	Generalized ataxia, gait abnormalities; aggression	Not reported	None	Unremarkable	At 36 months ataxia and hypermetria in all limbs (hind > fore); mild muscle atrophy; mild proprioceptive deficits in all limbs	Not performed	Not performed	None	Euthanized at 37 months (No)
5	M	12	Inappropriate (excessive) response to stimulation; fecal and urinary incontinence at exercise; dog-dog aggression; swam vertically in water; unable to wag tail; difficulty on stairs / jumping	From 12 months	None	Unremarkable	Circling to right; left sided proprioceptive deficits; ataxia and hypermetria in all limbs (hind > fore); slightly reduced menace response on left	Hematology, serum biochemistry, plasma amino acid levels, urine organic metabolites	Not performed	None	Alive at 36 months (n/a)
6	F	<24	Loss of initial training and house breaking, separation anxiety; food obsession; loss of tail movements; episodes of “blinking out”	Yes	Episode of aspiration pneumonia; hind limb lameness.	Unremarkable	No record of complete neurologic exam	Not performed	Not performed	Meloxicam (0.1 mg/kg once daily orally) for lameness	Alive at 36 months (n/a)
7	M	6	Inappropriate (excessive) response to stimulation from two months; ataxia from six months; swam vertically in water from 6 months; poor concentration in training from 6 months; aggression (dog-dog from 6 months; dog-human from 24 months); reduced tail movements; circling behavior	Yes	Distichiasis (surgically corrected at 36 months)	Unremarkable	Ataxia and hypermetria in all limbs (hind > fore)	Not performed	Not performed	None	Euthanized at 39 months (No)

8	F	3	Inappropriate (excessive) response to stimulation (manic, spins, running back and forth) from three months; poor training, ataxia, and loss of tail movements from 18 months; swam vertically in water; food / water obsession	From four months; progressive	None	Unremarkable	At 58 months, continuously alert (failed to relax); pacing gait; possible mild head tilt to left; ataxia and hypermetria in all limbs (hind > fore); reduced tail movement; mildly reduced hopping (hind > fore);	Hematology, serum biochemistry, serum thyroxine, CSF analysis	MRI (n=1); mild caudate nucleus cavitation	Carprofen (2 mg/kg twice daily orally) – no improvement	Euthanized at 58 months (Yes)
9	M	18	Inappropriate (excessive) response to stimulation from three months; swam vertically in water from four months; poor concentration in training from six months; separation anxiety from three months; aggression, (dog-dog from 12 months; dog-human from 42 months); ataxia from 12 months; fecal and urinary incontinence	From 18 months	None	Unremarkable	Ataxia and hypermetria in all limbs (hind > fore)	Hematology, serum biochemistry	Not performed	None	Euthanized at 42 months (Yes)

Supplementary Table S2