

**Supplementary Table S1.** Summary of SLE patients with intracranial calcification.

Author and year	Sex and age (y)	Disease courses (y)	Neurological Presentations	Brain CT findings	CSF examination	Autoantibodies	Gene mutation	Other complications	Treatment	outcome
This case	F/47	16	slurred speech	bilateral calcification in the <b>basal ganglia, white matter</b> of the cerebral hemispheres, and <b>cerebellar</b> hemispheres symmetrically	NA	ANA +; anti-dsDNA +; anti-SSA +; anti-ribosomal P +	NA	nephritis	NA	NA
Sait H, 2021 [1]	F/15	simultaneously	bilateral 6th and left upper motor neuron 7 <sup>th</sup> cranial nerve palsy, left hemiparesis with visual acuity limited to projection of light in both eyes	Intraparenchymal hemorrhage involving right frontoparietal region; bilateral <b>basal ganglia</b> calcification	NA	ANA 4+; Anti-dsDNA +; anti-SSA +	ACP5	Spondyloenchondrodysplasia with immune dysregulation	NA	NA
Zhang YX, 2020 [2]	F/53	13	dysarthria, bradykinesia, rigidity, hyperreflexia of the upper extremities, and abnormal tandem gait	multiple patchy and punctate lesions in the <b>bilateral lenticular nucleus and cerebellar dentate nucleus</b> , as well as radiating lesions along the <b>periventricular area</b>	normal	ANA 2+; anti-dsDNA 563 IU/ml, anti-RNP 2+; anti-SSA 3+; anti-Ro52 3+	NA	NA	Methylprednisolone	NA
Kara B, 2020 [3]	M/25	6	spastic paraparesis, mild intellectual disability	<b>periventricular, frontal subcortical and basal ganglia</b> calcification	NA	ANA+; anti-dsDNA+; anti-Smith +, anti-Smith/RNP +	ACP5	Spondyloenchondrodysplasia; claustrophobia	Corticosteroid, azathioprine, HCQ and rituximab. Heparin, acetyl salicylic acid.	Improved
Soh D, 2019 [4]	F/60	10	hypophonia, rigidity, bradykinesia, stooped posture, reduced arm swing and stride length, mild broad-based gait, and postural instability	calcification involving the dentate nuclei, basal ganglia, periventricular white matter, and subcortical white matter.	NA	NA	SLC20A2	Vertical supranuclear gaze palsy	NA	NA
Garg T, 2018 [5]	F/2.5	NA	Delayed speech and development	bilateral symmetrical calcifications in the <b>gangliothalamic</b> region	NA	ANA 3+; Anti-dsDNA 2+; Anti-Smith Ab 4+; anti-SSA 4+; anti-U1-RNP 4+	NA	NA	Oral steroids and HCQ	NA
Chang RS, 2015 [6]	F/54	NA	Shuffling gait, bradykinesia and rigidity	diffuse <b>basal ganglia</b> calcification with significant cerebral atrophy	NA	NA	NA	Parkinsonism; lupus nephritis; depression	Dopamine	NA
Troedson C, 2013 [7]	F/1.5	4	longstanding left 6 <sup>th</sup> nerve palsy, microcephaly and mild global delay	Basal ganglia calcification	Elevated neopterin	ANA 1:640; Anti-dsDNA +; Anti-Smith +; Anti-Ribosomal P +	NA	Malar rash, fevers, hepatitis, sterile cystitis	Prednisone and azathioprine.	Improved
Martinovic-Kaliterna D, 2013 [8]	F/65	17	insomnia and mild depression	bilateral symmetric intracranial calcifications in the cerebrum, <b>basal ganglia</b> , periventricular <b>white matter</b> and <b>cerebellum</b>	NA	ANA +, Anti-dsDNA +; negative antiphospholipid Ab and anti-ribosomal P Ab	NA	Fahr's disease?	Steroids, HCQ, and Zolpidem.	Improved
Jankovic M, [9]	F/64	6	confusion and hyporesponsiveness	Extensive bilateral and symmetric calcifications of <b>nucleus dentatus, cerebellum, striatum, pallidum</b> , and periventricular <b>white matter</b>	normal	Anti-dsDNA -	NA	idiopathic hypoparathyroidism and hypothyroidism; M. chelonae infection; Fahr's syndrome; septic shock.	Methylprednisolone, azathioprine	Died 2011
Ramantani G, 2011 [10]	M/12	6	severe developmental delay, spastic tetraplegia, multiple contractures, and severe scoliosis, dystonia and refractory epilepsy	<b>Periventricular</b> hypodensities with spotty calcifications around slightly dilated ventricles	NA	ANA+; anti-dsDNA +; ANCA +	SAMHD1 (AGS5) gene	Aicardi-Goutières syndrome	corticosteroids, azathioprine, etanercept	NA
Sibbitt WL Jr, 2010 [11]	M/56	10	hyperreflexia, gait disturbance, dysarthria, emotional lability, depression, cognitive decline	extensive calcification in the <b>thalamus, putamen, caudate nucleus, white matter</b> , and posterior <b>gray matter</b>	NA	ANA 1:640; anti-dsDNA 1:640; Anticardiolipin Ab +; lupus anticoagulant: positive	NA	Fahr's disease; antiphospholipid antibody syndrome; sepsis	NA	Died
Masuda Y, 2010 [12]	F/56	13	mental deterioration, bilateral pyramidal sign, parkinsonism, and intentional tremor	bilateral calcification in the <b>ganglia, dentate nuclei</b> , and the <b>white matter</b> of the cerebral hemisphere symmetrically	High IL-6	ANA +; Anti-SSA+; anti-SSB+; anti-ribosomal P Ab +	NA	NA	NA	NA

# Letters to the Editors

Author and year	Sex and age (y)	Disease courses (y)	Neurological Presentations	Brain CT findings	CSF examination	Autoantibodies	Gene mutation	Other complications	Treatment	outcome
Andres RH, 2008 [13]	F/53	20	psychotic episodes, progressive depression, insomnia, psychomotor slowing, gait disturbance and tremor of both hands. Rigidity of the upper extremities, ataxia, tetrahyperreflexia and bilateral pyramidal tract signs.	extensive calcification of the <b>basal ganglia</b> , <b>centrum semiovale</b> , <b>cerebellar</b> hemispheres and <b>brainstem</b>	NA	NA	NA	NA	high-dose immunosuppressive therapy.	Deteriorate
Navarro V, 2008 [14]	M/7		increased tone and hyperreflexia in lower limbs	two homogenous foci of calcification in the deep <b>white matter</b> of the frontal lobes bilaterally, and calcification of the <b>basal ganglia</b> particularly involving the caudate and putamina	normal	ANA+; Anti-dsDNA +	NA	spondyloenchondrodysplasia	Corticosteroids, chloroquine and cyclophosphamide, azathioprine.	Improved
Cañas CA, 2008 [15]	F/18	5	refractory seizures, severe dementia, psychomotor agitation, language involvement, and right hemiparesis	cerebral atrophy and multiple <b>paraventricular</b> , <b>basal ganglia</b> , <b>cortex</b> , cerebral <b>white matter</b> , and <b>cerebellum</b> calcifications; secondary hydrocephalus	Normal	ANA+; anti-dsDNA +; anti-Smith +; anti-RNP +	NA	NA	Steroids; azathioprine, cyclophosphamide, rituximab, lasmapheresis, IVIG.	Progressed
Filloux V, 2003 [16]	F/70	2	progressive loss of cognitive capacities and a right motor deficit with a pyramidal syndrome; important cognitive impairment with dementia and tetraplegia	Basal ganglia calcification	Elevated protein	ANA+; Anti-dsDNA+; lupus anticoagulant -; anticardiolipin Ab -	NA	Fahr's disease	Steroids, methotrexate	Died
Dale RC, 2008 [17]	M/0.8	3y before	progressive microcephaly, cortical blindness, myoclonic epilepsy, spastic and mental retardation	ventriculomegaly in addition to <b>periventricular</b> , <b>subcortical</b> and <b>basal ganglia</b> calcification	NA	ANA 1:132; anti-dsDNA 2350 IU/ml; Anti-Ro +; Anti-La +; Anti-C1q +; Anti RNP +; Anticardiolipin Ab+ Anti-dsDNA 2050 IU/ml; anticardiolipin Ab +	NA	NA	NA	Died
	M/0.4	1y before	progressive microcephaly, cortical blindness, myoclonic epilepsy, spastic and mental retardation	ventriculomegaly in addition to <b>periventricular</b> , <b>subcortical</b> and <b>basal ganglia</b> calcification (identical to his brother)	NA		NA	NA	NA	Died
Mathur A, 2000 [18]	F/27	Simultaneously	left sided hemiplegia	infarction in right frontoparietal region and a ring enhancing lesion in right basal ganglia	NA	Anti-dsDNA +	NA	NA	Oral prednisone	NA
Stuart BM, 1998 [19]	F/16	13	Headache, anosmia	extensive symmetrical calcification in the <b>basal ganglia</b> , the <b>internal capsule</b> , both <b>cerebellar</b> lobes, the <b>brainstem</b> and the <b>cerebrum</b>	Paraprotein, anti-astrocyte Ab >1:1600	ANA 1:3000; Anti-astrocyte Ab 1:125000	NA	NA	Steroids, HCQ. Improved	NA
Matsumoto R, 1998 [20]	F/22	10	psychosis and/or depression; mild left hemiparesis with a left extensor plantar response and confusion	moderate perisulcal atrophy and asymmetric granular calcification in the <b>basal ganglia</b> , <b>internal capsule</b> and <b>subcortical</b> and periventricular <b>white matter</b>	Increased cell count, protein and IgG; myelin basic protein 29.0 ng/ml; IL-6 4450 pg/ml; antiribosomal P Ab +	ANA +; Anti-dsDNA +; anti-Smith +; anti RNP Ab +	NA	respiratory failure	Methylprednisolone.	Died
Raymond AA, 1996 [21]	M/17	NA	Organic brain syndrome	<b>Basal ganglion</b> calcification	NA	NA		NA	NA	
	M/19	NA	Headache	<b>Basal ganglion</b> calcification	NA	NA		NA	NA	
	F/28	NA	Headache, seizures	Calcification in <b>basal ganglia</b> , <b>centrum semiovale</b> , and <b>cerebellum</b> . Infraction in right external capsule	NA	NA		NA	NA	
	F/29	NA	Unexplained syncope	<b>Basal ganglion</b> calcification, cerebral cortical atrophy	NA	NA		NA	NA	
	F/44	NA	Left optic neuritis	<b>Cerebral cortical</b> atrophy, calcification	NA	NA		NA	NA	
	F/45	NA	Chronic psychosis, seizures	<b>Basal ganglion</b> calcification, <b>cerebral cortical</b> atrophy, left periventricular infarction	NA	NA		NA	NA	
	F/47	NA	seizures	<b>basal ganglion</b> , <b>centrum semiovale</b> , <b>cerebellum</b> calcification; cerebral cortical atrophy	NA	NA		NA	NA	

Author and year	Sex and age (y)	Disease courses (y)	Neurological Presentations	Brain CT findings	CSF examination	Autoantibodies	Gene mutation	Other complications	Treatment	outcome
	F/48	NA	Organic brain syndrome	Calcification: <b>basal ganglion, cerebellum</b> ; cerebral cortical atrophy; Infraction: right caudate and external capsule, left internal capsule and external capsule	NA	NA		NA	NA	
Yamamoto K, 1992 [22]	F/32	14	dysarthria, moderate right hemiparesis with positive right Babinski sign and sensory disturbance in the right half of her body	Bilateral calcification in the <b>lentiform nuclei</b> , caudate nuclei, <b>thalamus</b> , <b>dentate nuclei</b> and <b>white matter</b> of cerebellar hemispheres	Elevated protein	ANA 1:640		NA	Oral corticosteroid	Improved
Daud AB, 1988 [23]	F/18	simultaneously	right sided paraesthesia with ipsilateral facial nerve paresis	ventricular and subarachnoid space widening and calcification in the right <b>basal ganglia</b> , right <b>paraventricular</b> area and also the frontal <b>cortex</b> of both hemispheres	NA	ANA +	NA	NA	steroid and anti-inflammatory therapy	NA
	F/21	6	Psychosis, hemiplegia	a resolving hypodense area in the right frontoparietal region, diffuse cortical atrophy and solitary <b>left paraventricular calcification</b>	NA	NA	NA	NA	NA	NA
Nordstrom DM, 1985 [24]	F/20	7	depressed sensorium, mental slowness and choreiform movements, spastic paraparesis, myelitis manifested by paraplegia of legs	perisulcal atrophy and basal <b>ganglia</b> calcifications	Elevated protein	Positive DNA binding; Coomb's test +	NA	NA	Prednisone.	Improved
	F/19	5	depressed sensorium, grand mal seizures	mild perisulcal atrophy and <b>basal ganglia</b> calcifications;	Elevated protein; oligoclonal bands	Positive DNA Binding	NA	NA	steroids and antiseizure medication. Improved	NA
Anderson JR, 1981 [25]	F/0.4	1y before	generalized hypertonia, cerebral and subarachnoid hemorrhage	moderate symmetrical dilatation of the ventricular system and bilateral calcification in the <b>basal ganglia</b> .	NA	Anti-dsDNA +	NA	Glomerulonephritis, Libman-Sacks endocarditis	NA	NA

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