

eTable 1 Demographic and clinical characteristics of SLONM patients and controls

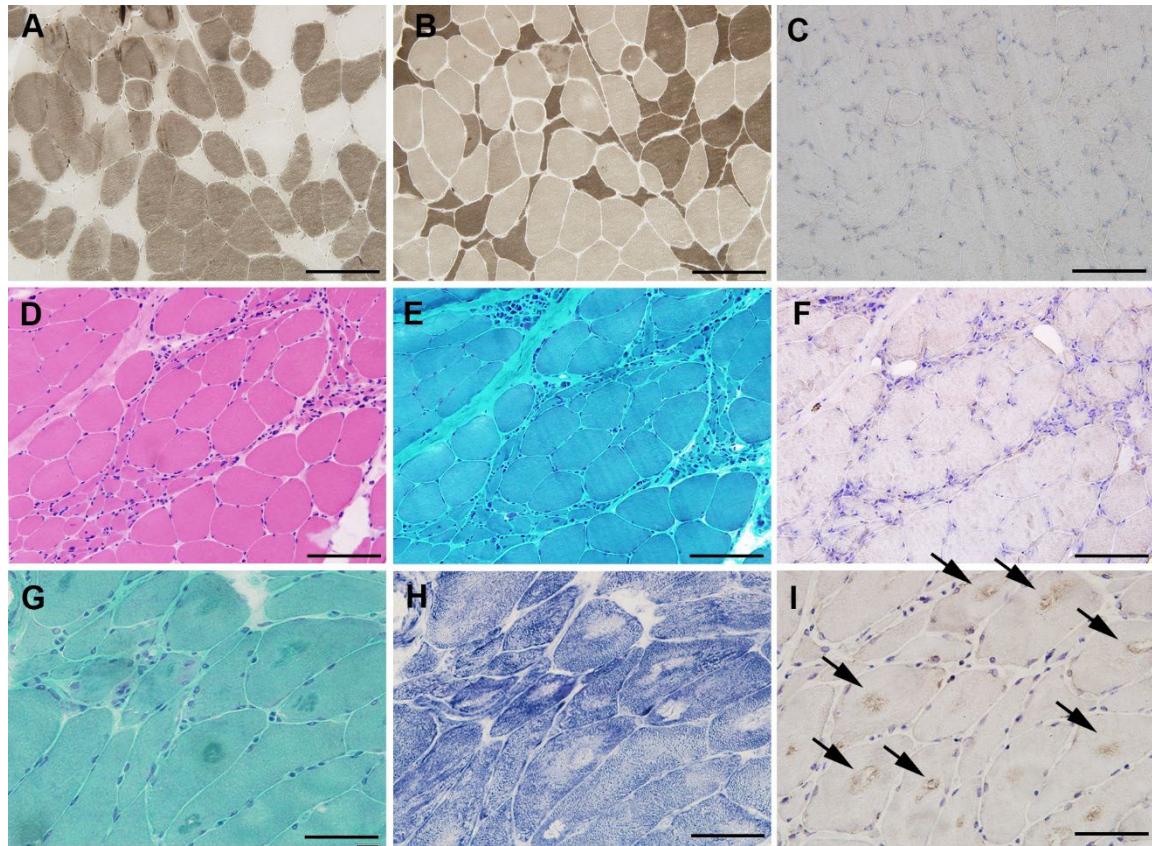
Abbreviation: SLONM=sporadic late-onset nemaline myopathy; IMNM= immune-mediated necrotizing myopathy; NC=normal

	SLONM (n=17)	Neurogenic atrophy (n=22)	Type II fiber atrophy (n=22)	Mitochondrial myopathy (n=5)	IMNM (n=5)
Age (y)**	54 (52, 63)	52 (50, 59)	53.5 (42, 66)	18.5 (14, 21)	47 (46, 48)
Course (mo)**	6 (5, 24)	10 (5, 36)	6.5 (3, 48)	18 (6, 108)	6 (3, 12)
Sex (male)*	6 (35.29)	16 (72.73)	6 (27.27)	5 (83.33)	2 (40.00)
Muscle weakness*	17 (100)	21 (95.45)	15 (78.95)	4 (66.67)	5 (100.00)
NE weakness*	9 (52.94)	4 (30.77)	1 (6.67)	1 (20.00)	1 (20.00)
NF weakness*	2 (11.76)	3 (13.64)	4 (18.18)	1 (16.67)	2 (40.00)
Proximal weakness*	17 (100)	20 (90.91)	14 (63.64)	4 (66.67)	5 (100.00)
Distal weakness*	5 (29.41)	16 (72.73)	8 (36.36)	2 (33.33)	3 (60.00)
CK (U/L)**	130 (97, 197)	133 (58, 400) ^a	127 (25.1, 331) ^b	678 (301, 1055) ^c	7521 (1000, 15000) ^d
EMG:	n=16	n=19	n=12	n=4	n=4
Myopathic change*	14 (87.5)	2 (10.53)	4 (33.33)	3 (75.00)	4 (100.00)
Neuropathic change*	1 (6.25)	16 (84.21)	5 (41.67)	1 (25.00)	0 (0.00)
Spontaneous activities*	12 (75.00)	3 (15.79)	1 (7.69)	0 (0.00)	0 (0.00)

control; CK=creatin kinase.

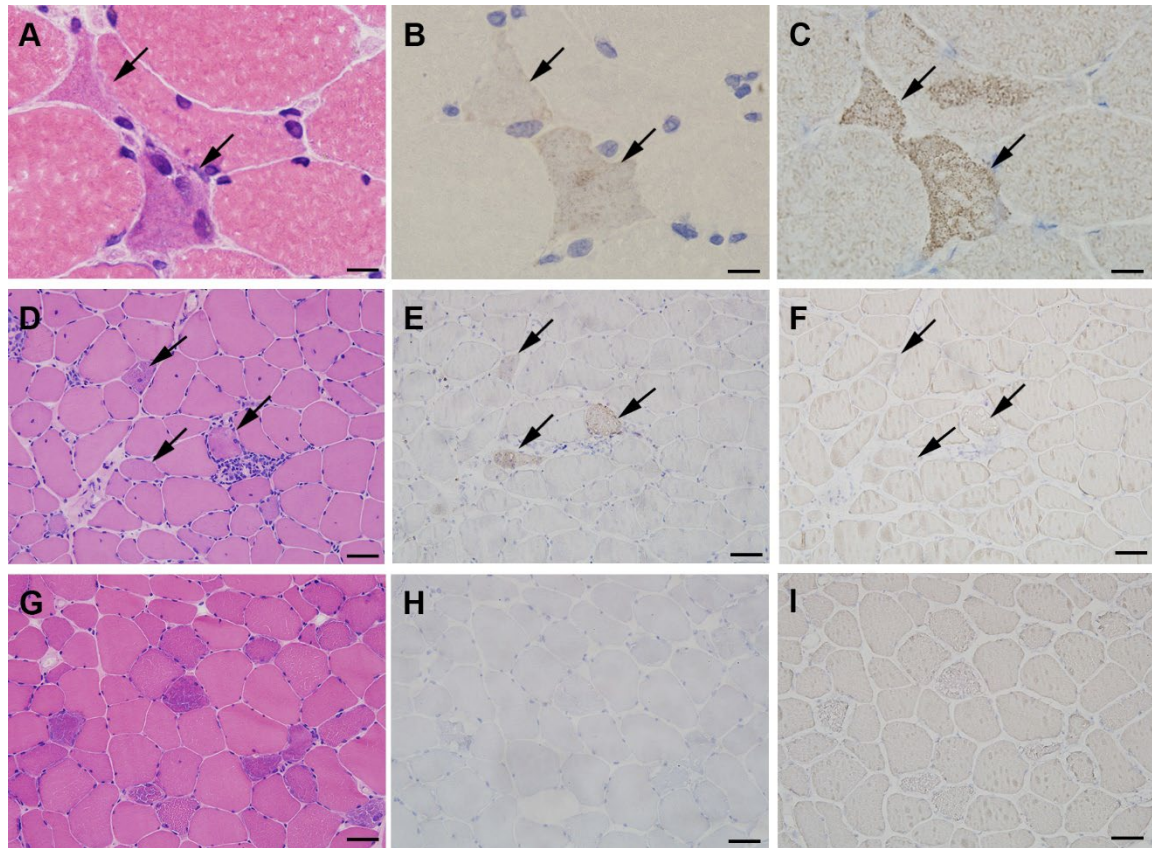
*N (%); ** Median (interquartile range).

^a n=14; ^b n=16; ^c n=2; ^d n=5; ^e n=1.



efigure 1. α -Actinin expression in type 2 atrophy and neurogenic atrophy

Type 2 fiber atrophy stained with ATPase at PH 4.3 (**A**) and PH 10.4 (**B**), showing no α -actinin expression (**C**). Neurogenic fiber atrophy stained with HE (**D**) and MGT (**E**), exhibiting no α -actinin expression (**F**). Target fibers in neurogenic atrophy stained with MGT (**G**) and NADH-TR (**H**), demonstrating α -actinin expression limited to the concentric zone (**I**, arrows). Scale bars: A-F, 50 μ m; G-H, 20 μ m.



efigure 2. α -Actinin expression in SLONM, IMNM and mitochondrial myopathy

Rod-bearing fibers with the basophilic cytoplasm in SLONM on HE (A), positive for both anti-NCAM (B) and anti- α -actinin staining (C). Regenerative fibers with the basophilic cytoplasm in IMNM on HE (D), positive for anti-NCAM staining (E) but negative for anti- α -actinin staining (F). Ragged-red fibers with basophilic cytoplasm on HE (G), negative for both anti-NCAM (H) and anti- α -actinin staining (I). Scale bars: A-C, 10 μ m; D-I, 50 μ m

