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LOWER URINARY TRACT FUNCTION IN SPINOCEREBELLAR ATAXIA 6

Hypothesis / aims of study

To investigate lower urinary tract (LUT) function in a rare, pure cerebellar, hereditary neurologic disease spinocerebellar ataxia type 6 (SCA6).

Study design, materials and methods

We recruited 11 patients with genetically diagnosed SCA6. They were 5 men, 6 women; mean age, 59.6 (36-69) years; mean disease duration, 8.9 (3-15) years; mean CAG repeat length 24.3 (21-26, normal< 18). Four had positional vertigo; all had cerebellar ataxia. We performed urodynamics/sphincter electromyography (EMG) in all subjects, irrespective of the presence of LUT symptoms (LUTS) with patients' informed consent.

Results

We excluded two cases of SCA6 with comorbid diseases (diabetes, decreased bladder sensation with large post-void residuals; and prostate hyperplasia, outlet obstruction by pressure-flow analysis) from this study. As a result, LUTS was observed in 5 of 9 patients (56%), urinary incontinence in 3 (33%), and urinary retention in none. Urodynamic abnormalities were seen in 3 of 9 patients (33%), including increased bladder sensation in 2, detrusor overactivity in one (11%), and weak detrusor on voiding in 2, but post-void residual in none. Sphincter EMG revealed neurogenic change in 5 of the 8 patients (63%) on whom the test was performed.

Interpretation of results

Cerebellar vermis and inferior olivary nucleus are the main pathology in SCA6. Experimental studies have shown that cerebellum has a role in regulating micturition. However, detrusor overactivity was observed in only 1/9 cases, indicating that cerebellum's effects on micturition in humans seem to be limited. Spinal cord pathology has recently been reported in SCA6, which is relevant to sphincter EMG abnormalities in our study.

Although in a few patients, our study showed that LUTS are uncommon in SCA6 and none had urinary retention, which is in clear contrast to common LUTS and urinary retention in multiple system atrophy. However, we cannot differentiate SCA6 from MSA by sphincter EMG abnormalities alone. Our study results contribute to clinical differential diagnosis of ataxic neurologic diseases.

Concluding message

LUTS are uncommon in SCA6 and none had urinary retention, which is in clear contrast to common LUTS and urinary retention in multiple system atrophy. However, sphincter EMG abnormalities can be observed in both SCA6 and MSA. Our study results contribute to clinical differential diagnosis of ataxic neurologic diseases.

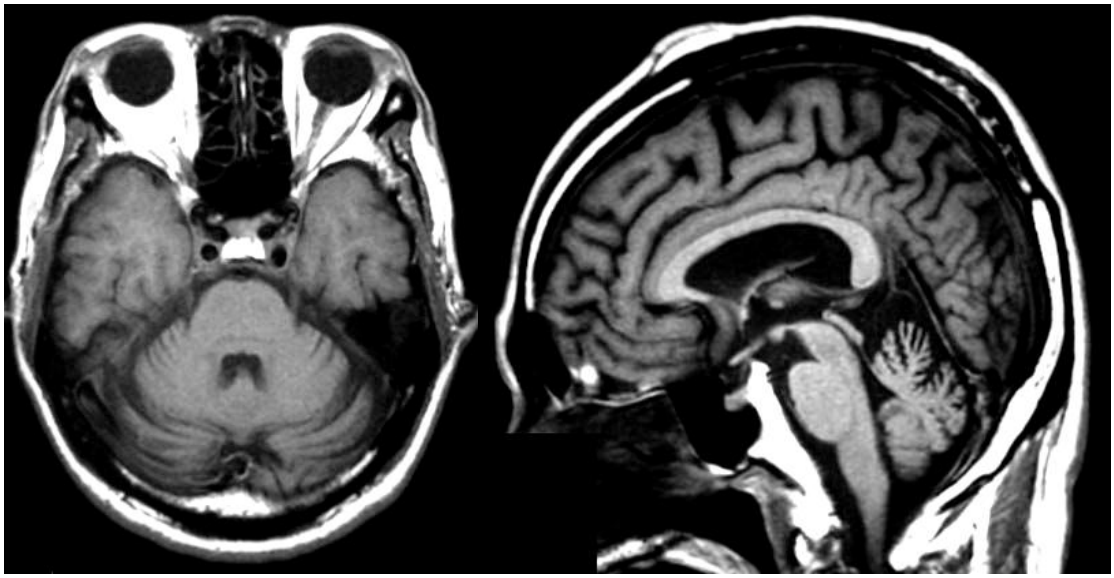

Figure 1 Brain MRI of the representative case.
Axial and sagittal views of T1-weghted images show marked cerebellar atrophy.

patient						neurological finding						lower urinary tract symptom					
No.	age	sex	illness	CAG repeat length	comorbid disease	vestibular		ataxia				daytime	nighttime	urinary	urinary incontiennce		voiding
			duration (years)	length [<18]		vertigo	nystag-mus	speech	upper extremity	gait	SARA (0-38)	frequency [<8]	frequency [<2]	urgency	frequency	type	difficulty
1	36	F	3	24		+	+	+	+	+	12	6	-	-	-	-	-
2	53	F	9	26		-	+	+	+	+	28	6	1	-	-	-	-
3	53	M	5	not mentioned		-	+	+	+	+	np (walk with cane)	7	1	-	-	-	-
4	57	F	12	26		+	+	+	+	+	23	8-10	1	+	weekly	urge & stress	-
5	61	M	15	23		-	+	+	+	+	23	7	1	-	-	-	daily
6	62	F	7	not mentioned	diabetes (HbA1C 7.5)	-	+	+	+	+	np (independent walk)	13	3	+	weekly	urge & stress	weekly
7	64	M	11	26		-	+	+	+	+	29	6	1	+	monthly	urge	-
8	67	M	8	26		-	+	+	+	+	22	6	-	-	-	-	-
9	67	F	7	21		+	+	+	+	+	27	8-10	1	-	-	-	-
10	67	M	17	not mentioned	prostatic hyperplasia	+	+	+	+	+	np (walk with cane)	10	3	-	monthly	unwitting	daily
11	69	F	4	22		-	+	+	+	+	20	8-10	2	-	monthly	stress	-

Table 1 Patients and lower urinary tract symptom (LUTS).

patient						urodynamics						sphincter electromyograhy		
No.	age	sex	illness	CAG repeat	comorbid disease	post-void	first sensation	bladder capacity	detrusor	pressur-flow analysis		detrusor-	MUP analysis	
			duration (years)	length [<18]		residual (ml)	(ml) [100-300]	(ml) [200-600]	overactivity	weak detrusor on voiding	outlet obstruction	sphincter dyssynergia	mean duration (ms) [<10.0]	MUP with duration>10 ms (%) [<20]
1	36	F	3	24		-	166	355	-	+	-	-	7.65	30
2	53	F	9	26		-	84	261	-	-	-	-	5.84	0
3	53	M	5	not mentioned		-	150	450	-	-	-	-	10.36	50
4	57	F	12	26		-	109	440	-	-	-	-	np	
5	61	M	15	23		-	251	397	-	np	np	-	11.44	60
6	62	F	7	not mentioned	diabetes (HbA1C 7.5)	250	88	400	+	+	-	-	12.00	50
7	64	M	11	26		-	118	472	-	-	-	-	8.54	30
8	67	M	8	26		-	91	328	+	+	-	-	9.08	50
9	67	F	7	21		-	120	296	-	-	-	-	6.65	20
10	67	M	17	not mentioned	prostatic hyperplasia	-	94	130	+	-	+	-	7.41	10
11	69	F	4	22		-	103	283	-	-	-	-	6.56	20

Table 2 Urodynamics.

CAG repeat, cytosine-adenine-guanine repeat; SARA, The Scale for Assessment and Rating of Ataxia; MUP, motor unit potential.

Specify source of funding or grant	**nothing**
Is this a clinical trial?	**No**
What were the subjects in the study?	**HUMAN**

Was this study approved by an ethics committee?	**Yes**
Specify Name of Ethics Committee	**Ethics committee of Sakura medical center Toho-university**
Was the Declaration of Helsinki followed?	**Yes**
Was informed consent obtained from the patients?	**Yes**