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Coexisting idiopathic cervical dystonia and primary vaginismus. A case report

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Cervical dystonia is seldom associated with other focal dystonias [4]. A patient with coexisting cervical dystonia and primary vaginismus led us to conjecture that primary vaginismus might be a form of dystonia.

A 32-year-old woman suffered from idiopathic right laterocollis. We recorded polymyography of the neck muscles, median nerve somatosensory evoked potentials (SEPs) and upper limb motor evoked potentials (MEPs). At age 38 the patient complained of introital pain during intercourse, vulvar pain and erythema diagnosed as type IV vaginismus [1, 9] complicated by vestibulitis. She also reported mild constipation. Hymen dilatation provided no benefit. We recorded pudendal nerve SEPs and the bulbocavernosus reflex (BCR), with the early response (R1) and late response (R2). Concentric needle electromyographic (EMG) activity was recorded from the levator ani (LA) muscle, between the anal and vaginal orifices, and the external anal sphincter (EAS) muscle. The motor unit potentials (MUPs) were collected and analyzed by standard "multi MUP analysis" implemented on the EMG system (Keypoint, Dantec Medical) [10]. Ten normal nulliparous females served as controls for BCR and EMG. During BCR examination subjects mildly contracted the pelvic floor. EMG was performed at rest, during straining and voluntary sphincter contractions. The patient was treated with botulinum toxin type A (BT-A) (Dysport®, Ipsen) injected into the LA. Our local Ethical Committee previously approved the protocol; subjects signed an informed consent.

Polymyography showed continuous involuntary activity from the right splenius and sternocleidomastoideus muscles. BT-A (300 MU) injections restored normal neck position. Upper limb SEPs and MEPs were normal.

Pudendal nerve SEPs were normal. BCR showed normal latency of R1; amplitude and duration of R2 were higher in the patient than in controls (Fig. 1a, Table 1). In the patient, EMG of the LA showed increased tonic activity at rest and paradoxical muscle activation during straining (Fig. 1b) and similar activity was recorded in the EAS muscle; no muscular hyperactivity was observed in controls (Table 2).

After BT-A treatment, the patient's myalgia diminished and intercourse and bowel movements normalized. EMG of LA and EAS muscles showed markedly reduced baseline hyperactivity and paradoxical activity with straining (Fig. 1c). Three months later the vaginismus returned and the patient interrupted pregnancy planning. The LA was injected again with BT-A (40 MU). The patient now repeats BT-A for vaginismus and laterocollis 3-monthly and has regular intercourse.

To our knowledge, coexisting idiopathic cervical dystonia and primary vaginismus in a patient successfully treated with BT-A injections is unique. Though not excluding a chance association, we leave open a possible multifocal dystonic disorder. First, our patient's EMG showed that laterocollis and primary vaginismus shared a common pattern of continuous hyperactivity, and lack of appropriate voluntary inhibition with dyssynergic LA and EAS muscle activation during straining. These features recall the muscle co-activation characterizing dystonia and may explain the patient's painful intercourse and constipation. Multifocal dystonia also fits in well with the prolonged late BCR, a central response [13] corresponding to the blink reflex R2, a response that is often abnormal in blepharospasm [2]. Our patient also responded well to 3-monthly repeated BT-A

injections. In vaginismus and genital pain syndromes, local BT-A treatment relieves spasms [3, 6, 11]. Possible reasons why our patient had a shorter-lasting response to BT-A than previously reported cases (3 months versus > 12) [6] could be that we injected BT-A at a lower dose or she had more severe LA spasms because of abnormal motor inhibitory mechanisms.

In cervical dystonia and vaginismus abnormal sensory inputs might cause neuroplastic changes within the CNS, thus altering the physiology of the sensorimotor program and/or a genetic permutation might constitute a risk factor so that any painful anatomic site becomes dystonic [4, 5, 7, 8, 12]. Our findings suggest further research into the pathophysiology of vaginismus.

Table 1 Pudendal SEPs and BCR recorded from the patient and ten controls matched for age. Values are means ± SD. Amplitude is expressed in uV, or mV when specified, latency and duration are in ms

	Patient		Controls	
Pudendal SEPs	P1-N1 Cortical		P1-N1 Cortical	
Latency	37.4		39.8 ± 1.3	
Bulbo-cavernosus reflex	First response R1	Late Response R2	First response R1	Late response R2
Latency	32	65	35.9 ± 9	60 ± 20
Amplitude	30.5	**535.7**	100 ± 70	150 ± 80
Duration	7.2	**135**	10.3 ± 8.5	41.2 ± 25

np not performed

Table 2 EMG data from LA and EAS musdes

Patient	Before BT-A		2 months after BT-A		Controls (10)	
Levator ani	Tonic activity		Tonic activity		Tonic activity	
MUPs at rest						
Amplitude uV	**428**		185		321 ± 69	
Duration ms	**13.6**		4.6		6.8 ± 1.5	
Area uV/ms	**812**		144		230 ± 189	
Poly (%)	**41**		20		16 ± 11.1	
MUPs (No)	**13**		5		4.4 ± 1.0	
Denervation	absent		**present**		absent	
Interference patterns	Voluntary contraction	Straining	Voluntary contraction	Straining	Voluntary contraction	Straining
Turns (No)	705	**448**	640	**283**	620 ± 153	No activity
Amplitude NV	394	**330**	362	**242**	390 ± 195	
Activity %	45	**21**	39	**11**	35 ± 6.9	
NSS/s (No)	540	**275**	478	**150**	428 ± 66	
Envelope uV	1233	**1028**	1099	**618**	1211 ± 486.4	
External anal sphincter	Tonic activity		Tonic activity		Tonic activity	
MUPs at rest						
Amplitude uV	**317**		176		138 ± 69.5	
Duration ms	**9.1**		5.3		5.5 ± 3.4	
Area uV/ms	**534**		157		111 ± 193	
Poly (%)	**16**		10		15.4 ± 8.3	
MUPs (No)	**11**		3		3.3 ± 0.4	
Denervation	absent		absent		absent	
Interference patterns	Voluntary contraction	Straining	Voluntary contraction	Straining	Voluntary contraction	Straining
Turns (No)	523	**228**	420	**133**	587 ± 146	No activity
Amplitude uV	348	**232**	289	**213**	361 ± 56	
Activity %	31	**9**	19	**5**	33 ± 13	
NSS/s (No)	333	**108**	220	**55**	416 ± 145	
Envelope uV	1094	**464**	954	**381**	1124 ± 228	

Activity (defined as percentage of time with EMG activity) number of polyphasic (with more than four phases) motor unit potentials (MUPs); *MUPs No* number of MUPs in 100 ms; *NSS* number of short segments; and *Envelope* MUP amplitudes with outliers removed. Bold characters indicate differences in patient's data from normal values (mean ± SD)

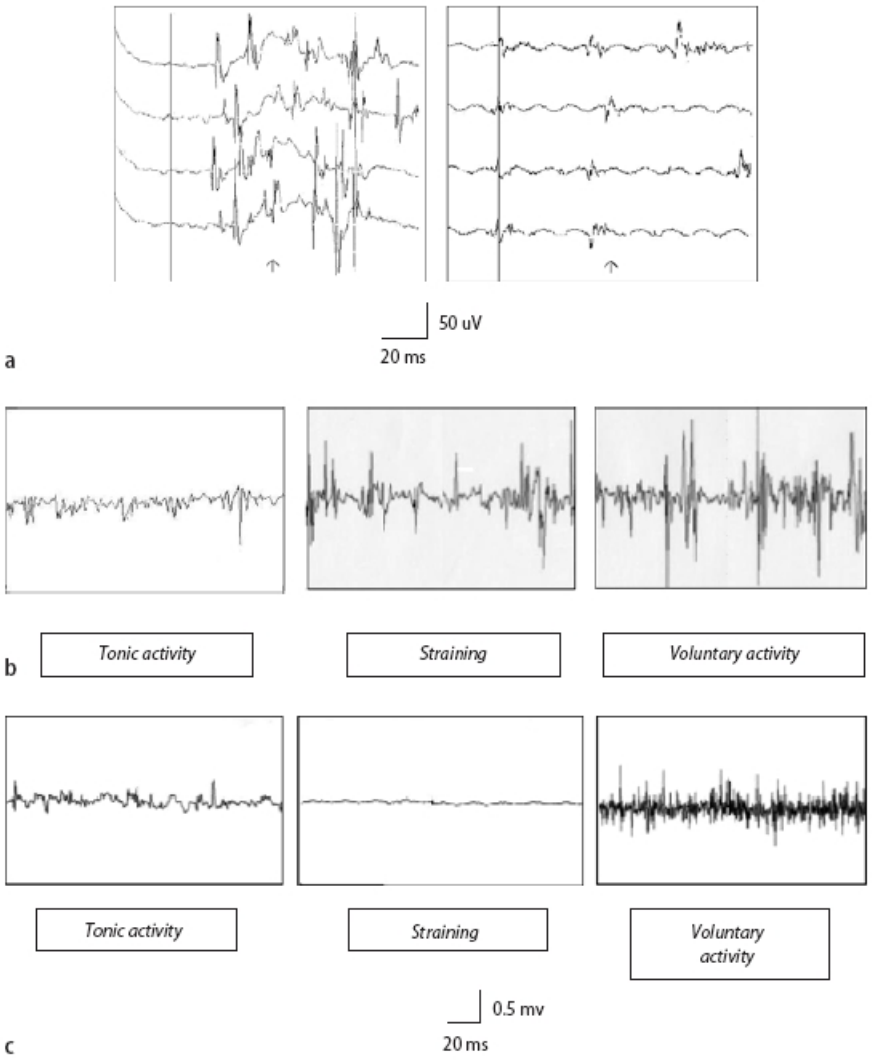

Fig.1 a Traces of the bulbocavernosus reflex (BCR) in the patient and a representative control. Line indicates R1 and arrow indicates R2. Note the increased duration and amplitude of R2 in the patient. **b** Traces of EMG activity from the LA muscle in the patient before BT-A injection: from right to left, increased tonic activity at rest, physiological interference pattern during voluntary contraction and subinterference pattern during straining. **c** Traces showing the EMG activity from the LA in the patient two months after BT-A injection into the LA muscle. Note the denervation activity (fibrillation) at rest and decreased voluntary muscle activity

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