other neurodegenerative disorders. However, the diagnosis of vCJD should be considered if hypogeusia and hyposmia are accompanied by changes of personality and other, more "typical" features of vCJD.

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Obsessive-compulsive characteristics in patients with writer's cramp

Writer's cramp is characterised by a muscular spasm in the hand of the writing arm, and is often provoked during specific tasks such as writing. Because of its highly task specific nature, some neurologists believed that writer's cramp was of psychogenic origin. However, recent electrophysiological and neuroimaging studies unanimously confirmed basal ganglia dysfunction in patients with writer's cramp. As a consequence, writer's cramp is currently regarded as a form of focal dystonia with neurophysiological pathogenetic mechanisms.[1]

On the other hand, in the classic psychiatric literature, clinicians sporadically pointed out obsessive-compulsive personalities in patients with writer's cramp. Bindman and Tibbetts described 10 patients with writer's cramp, nine of whom had obsessional personalities.[2] These findings have never been confirmed using reliable psychometric measures. However, because of the above mentioned evidence of basal ganglia pathophysiology in writer's cramp, together with the growing evidence of basal ganglia involvement in obsessive-compulsive disorder, it is of great interest to elucidate the relation between writer's cramp and obsessive-compulsive symptoms.

In the present study, we evaluated obsessive-compulsive symptoms in patients with writer's cramp. Obsessive-compulsive symptoms may simply be a psychological reaction to the writing impairment. To rule out this possibility, patients with writing impairment due to peripheral nerve lesions were included as a second group of control subjects, in addition to normal healthy control subjects.

Twelve consecutive patients with writer's cramp who visited the dystonia outpatient clinic of the Department of Neurology at Kyoto University from May 1998 to December 1998, were evaluated with the Yale-Brown obsessive-compulsive scale (Y-BOCS). All patients had been treated with muscle afferent block for more than 3 months.[3] As a consequence, their writing disabilities were moderately improved. All of them could write their names in four to six Chinese characters at the time of assessment. A disease control group consisted of seven patients with carpal tunnel syndrome, three with cervical spondylotic radiculopathy, one with chronic inflammatory demyelinating polyneuropathy, and one with myasthenia gravis. All these patients showed disabilities in finger movements due to peripheral nerve lesions of an upper limb, and were recruited from the outpatient clinic of the Department of Neurology at Kyoto University. A healthy control group, age and sex matched to the writer's cramp group, consisted of office clerks or secretaries of Kyoto University and Shiga University. None of the subjects had received antidepressive or neuroleptic medication for at least 6 months before the study. None of the subjects had a history of any psychiatric diseases.

For these three groups, obsessive-compulsive symptoms were evaluated by trained psychiatrists, based on the semistructured interviews assessing Y-BOCS. The scores of the writer's cramp group and two control groups on Y-BOCS were compared using a two tailed Kruskall-Wallis test for non-parametric data. p Values lower than 0.05 were considered to be significant. When the result was significant, the analysis was repeated with each pair of groups, and the p values were multiplied by three to correct for multiple comparisons (two tailed Mann-Whitney tests). Informed consent was obtained from all the participants after the procedure of the study had been fully explained.

The 12 patients with writer's cramp, 12 patients in the disease control group, and 12 normal controls did not differ in age or sex ratio (table 1). Mean Y-BOCS scores were 5.8 (SD 5.5, range 0–18) for the writer's cramp group, 1.0 (SD 1.4, range 0–4) for the disease control group, and 1.0 (SD 1.0, range=0–2) for the normal control group, respectively. There was a significant effect of diagnosis on the total scores of Y-BOCS (Kruskal-Wallis test, χ^2=5.99, p=0.012), with significant differences between the writer's cramp group and disease control group (p=0.030, corrected for three comparisons) or normal control group (p=0.036) but not between the disease control group and the normal control group (p>0.99). The compulsive symptoms of the writer's cramp group were mainly related to cleaning and ordering.

The result of our study showed that patients with writer's cramp had higher obsessive-compulsive symptoms than the disease control group or normal control group. Although according to the classic literature the patients with writer's cramp often have obsessive personality features,[2] the present study is the first to confirm the clinical finding using objective psychometric measures.

The disease control group consisted of patients with peripheral nerve lesions, who had discomfort in writing comparable with the patients with writer's cramp in this study, who were treated with muscle afferent block.[3] Thus the current result does not support the idea that physical distress and social disability due to writing impairment is the main cause of the obsessive-compulsive features of patients with writer's cramp, but supports the notion that writer's cramp and obsessive-compulsive symptoms would develop due to common pathophysiological mechanisms. However, it should be noted that our patients with writer's cramp had been under the treatment for more than 3 months and the duration of disease was relatively short. In most cases of writer's cramp without treatment, it is known that writing disability is extremely severe, causing a significant effect on their lifestyle. The present result does not deny the possibility that the condition could cause abnormal psychopathology in writer's cramp.

Recent studies reported abnormal metabolic activity in basal ganglia together with frontal and anterior cingulate areas in patients with obsessive-compulsive disorder. These data suggest basal ganglia dysfunction as a neural basis of obsessive-compulsive symptoms (for a review, see Saxena *et al*[4]). Therefore, we suspect that basal ganglia dysfunction is the common neural basis of writer's cramp and obsessive-compulsive symptoms. In other forms of focal dystonia such as spasmodic torticollis and blepharospasm, high obsessive-compulsive symptoms were reported.[5] Thus, the basal ganglia hypothesis of obsessive-compulsive symptoms may be applied to focal dystonia in general. As obsessive-compulsive symptoms of the patients in this study were not severe enough for the diagnosis of obsessive-compulsive disorder, and due to the small sample size, these findings should be interpreted with caution. Larger studies are necessary in clarifying the pathophysiology of writer's cramp and associated obsessive-compulsive symptoms.

Table 1 Ages, disease duration, and sex ratio of writer's cramp group and control groups

	Age range (y)	*Mean age (SD) (y)*	*Disease duration range (y)*	*Mean duration (SD) (y)*	*Sex ratio (male/female)*
Writer's cramp	21–70	42.6 (16.5)	1–9	4 (2.6)	8/4
Disease control	22–66	44.9 (16.2)	1–5	2 (1.6)	7/5
Normal control	22–70	43.1 (16)	—	—	6/6

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Cervical spondylotic myelopathy and Kennedy syndrome mimicking amyotrophic lateral sclerosis

A 62 year old male patient presented with a longstanding history of slowly progressive limb weakness, speech, and swallowing difficulties. In 1983 a diagnosis of amyotrophic lateral sclerosis had been made. At that time his physical examination showed tongue atrophy with fibrillations, proximal limb weakness, and brisk lower limb tendon reflexes. Electromyography showed abnormal "spontaneus activity" with fibrillations and positive sharp waves in muscles of all limbs. His further medical and family history was unremarkable.

In January 2000 neurological examination showed mild facial weakness, marked atrophy and fibrillations of the tongue, severe dysarthria and dysphagia, atrophy, and weakness of the shoulder girdle and arm muscles, and an unsteady and broad based gait. Apart from brisk knee jerks, deep tendon reflexes were absent and plantar responses were negative. Sensory testing was normal. General physical examination showed slight gynaecomastia. Laboratory testing showed raised creatine kinase (305 U/l) and lactate dehydrogenase (195 U/l) concentrations. Needle EMG demonstrated positive sharp waves and fibrillation potentials and long duration polyphasic motor unit potentials with increased amplitudes in muscles of all limbs. By contrast, motor and sensory nerve conduction studies gave normal results.

As cervical myelopathy is an important differential diagnosis in patients with suspected motor neuron disease, cervical MRI was performed. As shown in figure 1, MRI disclosed marked cervical spondylosis with appreciable narrowing of the spinal canal between C3 and C6. In addition, T2 weighted images showed intramedullar changes with foci of high signal intensity at the level of C5 indicating myelopathy. Although these changes may readily explain the weakness in his upper limbs, the cause of bulbar symptoms and denervation in his lower limbs remained unclear.

The presence of slight bilateral gynaecomastia prompted us to look for androgen receptor gene mutations, which cause X linked spinal bulbar muscular atrophy.[1] This disorder, also known as Kennedy syndrome, is caused by an unstable expansion of a CAG repeat in exon 1 of the androgen receptor gene (Xq11–12). The androgen receptor is highly expressed in motor neurons of the brain stem and spinal cord. The CAG repeat expansion is thought to confer a toxic gain of function to the androgen receptor protein resulting in irreversible damage of brain stem and spinal cord motor neurons. In addition, the impaired ability to transactivate androgen sensitive genes of the mutated receptor may account for endocrine features such as gynaecomastia or testicular atrophy in spinal bulbar muscular atrophy.[2–4]

Genetic analysis in our patient showed one allele carrying an abnormally expanded CAG repeat (44; normal range 16–33) thus confirming the diagnosis of Kennedy syndrome.

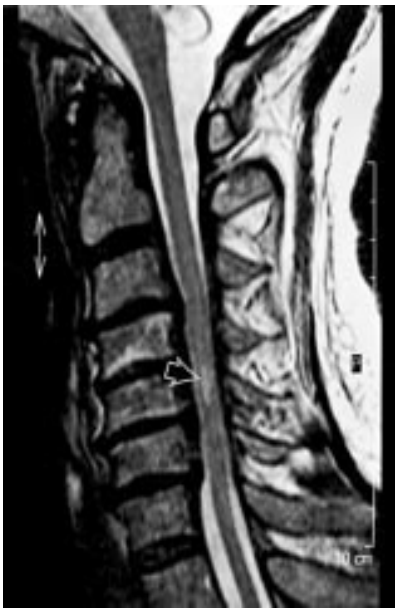

Figure 1 Spondylotic ridging at the C3-C6 level causing spinal stenosis is shown in the sagittal T2 weighted MR image. Additionally increased signal intensitiy is seen within the spinal cord at the level of C5.

The present patient with coexisting cervical spondylotic myelopathy and Kennedy syndrome highlights the diagnostic value of an intensified investigation including cervical MRI and androgen receptor gene analysis in patients with an unusual clinical presentation of motor neuron disease.

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Posturally evoked vomiting without nystagmus in a patient with Arnold-Chiari malformation

Arnold-Chiari malformation type I (ACM I) is a developmental anomaly of the rhombencephalon characterised by displacement of the cerebellar tonsils into the foramen magnum and elongation of the medulla. It usually presents in adult life with head motion induced oscillopsia, ataxia, headaches, cervical pain, or Valsalva induced dizziness.[1]

Various ocular motor abnormalities have been reported in patients with ACM I. Among these, downbeat nystagmus and periodic alternating nystagmus are the most common. Other often encountered ocular signs, such as gaze evoked nystagmus, rebound nystagmus, and impaired smooth pursuit, reflect cerebellar involvement.[2 3]

Vertigo of vestibular origin, being peripheral or central, is usually accompanied by nystagmus and nausea, or vomiting, and is often influenced by head position.[3 4] The entity of central positioning vomiting without, or little, vertigo and nystagmus (posturally evoked vomiting (PEV)) was first reported by Drachman *et al* and later recognised by Brandt and Baloh and Halmagyi.[3–5]

Posturally evoked vomiting is generally poorly known as a warning symptom of a posterior fossa lesion and is often misinterpreted.[5] Whereas it has been documented in patients with posterior fossa tumours, it has not been reported in patients with developmental abnormalities. We report on a patient with ACM I where PEV was the most prominent presenting symptom.

This 57 year old woman was seen in our vertigo clinic because of gait unsteadiness and postural vomiting. Her history included an aortic valve replacement for aortic insufficiency, nephrolithiasis, and peptic disease. She was treated with warfarin.

For years, she had dizziness and severe nausea while looking up. During the past months severe nausea and vomiting appeared when she tilted her head to either side and down. Lately, she had become unsteady. She also complained about left high pitched tinnitus and intermittent pain in the left shoulder.

On examination her eyes were properly aligned with a full range of movements. No primary or gaze evoked nystagmus was seen, with and without Frenzel's glasses. The saccadic eye performance was normal, but the smooth pursuit in both the horizontal and the vertical plane was saccadic. The vestibulo-ocular reflex, examined by a doll's eyes movement, head thrust test, and dynamic visual acuity test, was normal. Mild dysmetria on finger-nose testing and finger-finger testing was found bilaterally. The deep tendon reflexes were brisk in the upper and normal in the lower limbs. The plantar toe responses were flexor. Sensation was normal. The gait was atactic and the Romberg test negative.

On testing the eyes in the Dix-Hallpike position to either ear, as well as in the head down position, the patient reported severe nausea, and became pale and perspired. However, no nystagmus was seen either by direct observation, or with Frenzel's glasses.

The symptoms persisted with repetition of the positioning.

An electronystagmogram (ENG) documented saccadic eye tracking in the horizontal plane. The optokinetic nystagmus was asymmetric with little increment after increased speed velocity of the target. When supine and with her head turned to the left, nystagmus of 7°/s, beating to the left, was recorded. No nystagmus was recorded on Hallpike testing.The caloric test was within normal limits.