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A case reflection on task specificity in a client with Writer's cramp ¹Walton M, ²Mathew T

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Abstract

This is a case reflection on task specificity of a client with Writer's Cramp (WC). The clients' condition is compared with the book picture. WC is a task-specific movement disorder that manifests itself as abnormal postures and unwanted muscle spasms that interfere with motor performance while writing. One symptom of WC, typically during the initial stage, is a tight grip on the pen. Slowly, handwriting becomes less legible. About half of the patients with simple cramps progress to having dystonia with other activities. Treatment for WC includes oral medications with anticholenergics; injection botulinum toxin; neurosurgery including neurostimulation, and rehabilitation. The surgical management of WC is Thalamotomy or globus pallidus internus deep brain stimulation (GPi DBS) in patients not responding to medical treatment.

Case Summary: This is the narrative of a 47 year old healthy female who was working as a faculty in a college and who was pursuing her PhD. She had obtained the 3rd rank in an All-India PhD entrance examination which was of multiple choice questions(MCQ) and got selected for the same. During the examination she experienced difficulty in circling the correct answers but was able to complete the paper as it was MCQ. A year later when it came to writing the pre- PhD exam which is of a three hour paper she found it difficult to complete the paper on time as she experienced severe pain and cramps in the right hand and wrist while writing. She found it difficult to write though she had all the answers on her finger tips. She was able to clear one paper with good marks, but the second paper she lost by four marks, so she had to repeat both the papers and was frustrated about the same. She requested the University to allow her to type or use a scribe, but was not granted permission for the same. But was given the permission to re write the exam based on medical grounds. That is when she approached the neurologist for help to solve her problem of being unable to write for a prolonged period of time, so that she could get treated and clear the exam in the next attempt after six months. The Neurologist examined her, and motivated her that there was treatment available for the same. On physical examination she had inability to sustain

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abduction of the fingers of the right hand for more than 5 seconds. She was also asked to write a few sentences and a video was taken. It was observed that she had an abnormal posture while holding the pen. She had a tight hand grip with extension of the wrist and flexion of the index finger while writing. The neurologist diagnosed her with Writer's cramp and advised her to take BOTOX treatment as a temporary relief for writing the next exam. This would help to improve her muscle power while writing. The client was treated with a single dose of Botulinum 50 IU (Botox) i.e., IM injections over six specific areas on specific muscles of the forearm of the right hand, three months before the date of examination. Immediately after the injection the client experienced a muscle power of 3/5 to 2/5 for almost two months. She expressed that she could not carry a bag it would slip out of her thumb and index finger grip; could not open a tap or lift anything heavy, especially lifting vessels while cooking. She could not even sign a cheque or any documents at work. She was explained about the same by the neurologist, that it would take time to increase the muscle power. She approached him twice in a three month period out of anxiety of being unable to write the exam. As the date for the exam approached the client practiced writing and found that she was holding the pen with a horizontal thumb grip while writing. On the day of the exam after much prayer she started the exam but was not able to write also due to stress of passing the exam which triggered further spasm. She almost decided to give up and move out of the exam hall, when she prayed and was determined to write how much ever she could. By God's grace she was able to complete the paper and passed the pre-PhD examination. After a five year period she successfully completed her PhD and wanted to share her experience in order to motivate others with a similar problem, as she was a teacher herself and many students may experience the same.

Introduction

Writer's cramp has been recognized for over a century, and originally was construed as a physical motor disorder. However, an unfortunate use of the descriptive term 'professional neuroses' to describe this and other similar task-specific conditions, coupled subsequently with fashions in psychiatry, led to the mistaken belief that writer's cramp was due to psychic rather than motor pathology. Evidence has accumulated in recent years showing that writer's cramp is a real focal motor disorder, with a close relation to dystonia. WC is also termed as Task Specific Dystonia (TSD). Based on extent of body parts involved it is also called Focal Hand Dystonia (FHD) Evidence from research and clinical cases shows that there is specific aetiology, clinical





features, pathophysiology and treatment of writer's cramp and analogous occupational cramps, with the aim of drawing the attention of neuroscientists to this common, bizarre and poorly understood condition.³

Definition

Writer's Cramp (WC) is a task-specific movement disorder that manifests itself as abnormal postures and unwanted muscle spasms that interfere with motor performance while writing¹.

The client had complaints of a tight pen grip, abnormal hand posture and experienced severe muscle spasms during a specific task of writing for prolonged period of time hence she was diagnosed with WC.



Classification

According to the new classification, WC is considered a sporadic focal dystonia with late adult onset between the ages of 30 and 50 years.⁴

The client was 47 years at the time of diagnosis and came to visit the neurologist as she could not complete a three hour paper of a pre PhD exam.

In clinical practice, an initial classification divided the patients into two groups, those with simple and those with dystonic WC, on the basis of the absence or presence of dystonia while performing other tasks. ⁵ However, this simple classification seems inappropriate, as there may be transitions from highly specific deficits, which only affect the writing of specific letters, to simple and then to dystonic WC, and eventually to multifocal dystonias. ⁶

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The present client can be considered to have simple dystonic WC as she did not experience difficulty in performing other tasks other than writing for prolonged period of time. However the client did have difficulty in writing specific letters such as alphabet like s, g, b, o, m, w, x and numbers such as 8, 3, 6 and 9.

Signs and symptoms/ Diagnosis

One symptom of WC, typically during the initial stage, is a tight grip on the pen. Hand–wrist flexors are more commonly involved than extensors, though hyperextension of the distal phalanges or even the fingers may occur.³ Slowly, handwriting becomes less legible. About half of the patients with simple cramps progress to having dystonia with other activities. Remissions are uncommon, and symptoms can progress to the other hand in about 5% of cases.³

In the present case the client also had a tight grip on the pen with extension of the wrist and flexion of the index finger while writing. Hand writing was illegible. Hand cramps was experienced. There was no progression observed in the other hand.

Symptoms appear at a mean age of 38 years.⁷ Generally, FD of the limb is rare, and prevalence has been estimated in a more recent meta-analysis to be 15 per 100,000 people.⁸ The prevalence rate of WC was reported to be 6.9 per 100,000 persons, whereas the incidence was 0.27 per 100,000 in one year.⁹

The present client had experienced this problem at the age of 37 years while appearing for a post graduate examination, but did not take it seriously, as she felt it was due to stress or poor illegible handwriting. But during PG, the requirements of the course were repeated assignments, hence due to continuous writing every day hand writing was neat and legible. But when the client experienced stress she did not have control over the untidy handwriting and had severe muscle spasms.

Patients with simple WC frequently reported discomfort in other daily activities, like typing





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on a computer keyboard or using a spoon. It is noted that frequent association of other features of segmental and generalized dystonia in patients with dystonic WC over 30 years ago and, nowadays, even the spread of motor symptoms to the opposite hand are reported.^{3.10}

The client had difficulty in holding the tooth brush and experienced mild tremors, and adopted a tight hand grip rather than a finger grip to avoid tremors while brushing. Similar grips were seen while holding a spoon or using a knife while cutting vegetables. No hand tremors were seen, except while holding an object and trying to perform a task. She experienced difficulty in writing on the blackboard during class and had to adopt a posture to avoid tremors. However she did not experience any difficulty while typing on a computer.

A clinical profile of a large cohort of patients with WC was done in India. A chart review of 125 patients (F:M = 16:109) with WC seen over 10 years at the National Institute of Mental Health and Neurosciences (NIMHANS), India. The results showed that the mean age of patients was 36.8 ± 14.3 , the mean age at onset (AAO) of symptoms was 31.5 ± 2.7 and the mean duration of difficulty in writing was 5.3 ± 6.3 . The most common presenting complaint while writing was pain in the forearm (56%), followed by tremulousness of hand (28.8%) and abnormal posturing of hand (15.2%). Along with writing difficulty, 5.6% had difficulty in typing. The mean AAO of writing difficulty was significantly earlier in women than in men (22.8 \pm 3.5 vs. 32.8 \pm 4.2 years, p < 0.001). While writing, excessive extension (41.6%) of wrist was the most common abnormality, followed by excessive flexion (37.6%) and extension (19.2%) of index finger. The study concluded that women with WC have a significantly earlier age of onset than men. Excessive extension of wrist with flexion of thumb and index finger were the most common abnormalities noted in WC. 12

Secondary motor disturbances with reduced range of motion in other task than writing have also been described in patients suffering from WC. These were related to the severity and duration of the disorder and explained by biomechanical abnormalities of the hand, possibly as a consequence of a combination of innate factors and psychological factors.

The subject did experience secondary motor disturbances as mentioned earlier but was not very significant in disturbing daily activities.

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The diagnosis includes physical examination, writing test i.e., as the client to write and observe the pen grip, posture of the hand and the hand writing, the ability to draw circles and certain alphabets. Next test is to keep hands forward with fingers in abduction, it will be observed that the affected hand's fingers will automatically adduct and cannot hold in abduction position for long. The other investigation is Electro-Myogram(EMG)

The client was asked to write, tight pen grip and abnormal posture of hand was observed. She was not able to hold fingers of the affected hand in abduction for more than five seconds. Illegible handwriting and difficulty in drawing circles was also observed.





Management

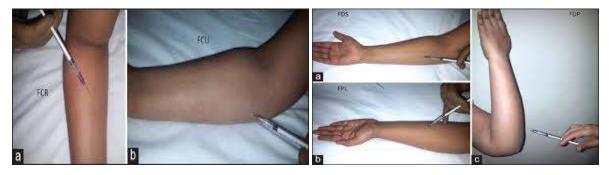
Treatment for WC includes oral medications such as anticholinergic drugs and benzodiazepines, injection botulinum toxin, neurosurgery including neurostimulation, and rehabilitation.² Role of injection botulinum toxin has been extensively studied in writer's cramp patients and found to be effective; however, selection of muscles and techniques of injection are crucial in getting best results. ¹⁰ Typical EMG-guided delivery of botulinum toxin to the active intrinsic hand musculature that is identified as giving rise to the dystonia has helped restore, to varying degrees, coordinated movement of the dystonic musculature. In some cases, this allows some return of writing function. Of note, it has been shown via PET studies that botulinum toxin

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does not reverse the cortical dysfunction associated with writer's cramp. 10

In the present case the client was treated with a single dose of Botulinum 50 IU (Botox) i.e., IM injections over six specific areas on specific muscles of the forearm of the right hand, 3 months before the date of examination. Immediately after the injection the client experienced a muscle power of 3/5 to 2/5 for almost two months. She expressed that she could not carry a bag it would slip out of her thumb and index finger grip; could not open a tap or lift anything heavy, especially lifting vessels while cooking. She could not even sign a cheque leaf or leave forms. The client was able to write the exam and complete the specific task for which she had taken treatment from the neurologist.



Videos after injection with Botilinium (attached separately)

Limb immobilization using a plastic splint, motor training by asking the patient to write using individual fingers, and sensory training by reading and practicing Braille have been used.^{2,10} Brain surgery includes lesionectomy and more recently deep brain stimulation of ventralis oralis (Vo) nucleus of thalamus. The surgical management of WC is Thalamotomy or globus pallidus internus deep brain stimulation (GPi DBS)in patients not responding to medical treatment. Astudy was done on eight patients (all men, age 16–47 years) with refractory focal hand dystonia who underwent either ventrooralis (Vo) thalamotomy (seven patients) or GPi DBS (one patient) using stereotactic techniques. Preoperative video recordings, writing movement score for dystonic posture and latency of dystonia (WMS), and symptom severity scores (SSSs) were evaluated at baseline and latest follow-up ranging from 1 to 4 years. The results revealed that all patients had difficulty in performing their most common tasks. The duration of symptoms







ranged from 6 months to 12 years. All patients obtained immediate postoperative relief from the dystonic symptoms, and the effect was sustained during the follow-up period. The WMS (range 0–28) improved from a mean of 14.5 before surgery to 2, whereas the SSS (maximum 43 and minimum 10) improved from a mean of 15.3 before surgery to 2 at the last follow-up. There were no surgical complications, morbidity, or mortality. The study concluded that Vo thalamotomy or GPi DBS offers successful symptom relief in patients with task-specific dystonia. ²

Conclusion Writer's Cramp is a focal hand dystonia which can be disturbing to the client when the need to perform certain specific tasks. This article was written to create awareness about WC, that it is a condition that may be prevalent in any student or faculty, it may show as symptoms only when the person is in stress or during the periods of examination. Treatment is available for temporary relief to achieve the specific task. The client mentioned above was able to perfom her day to day activities of writing, but had experienced difficulty during a three hour exam for which she took botilinuim injection and achieved her task of completing an exam successfully.

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