Current PSYCHIATRY

CASES THAT TEST YOUR SKILLS

Lonely and grieving her daughter's death, Ms. M confronts diminishing coordination and increasing involuntary limb movements. Doctors say she has conversion disorder. Is this diagnosis correct?

One patient's 'moving' story

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HISTORY 3 'UNCONTROLLABLE' MONTHS

Ms. M, age 57, presents to the ER complaining of coordination problems and involuntary limb movements that have gradually worsened over 3 months.

Two months ago, Ms. M's primary care physician and neurologist diagnosed her with conversion disorder. Brain MRI at the time showed mild chronic ischemic changes; cervical spinal cord MRI was normal. The neurologist referred Ms. M to a psychiatrist, who prescribed duloxetine, dosage unknown. She started having suicidal thoughts and trembling after starting the medication, so she stopped taking it after 1 week.

Physical exam shows upbeat nystagmus, inconsistent sensory findings, limb ataxia that is more pronounced on the right side, and uncontrollable limb movements, particularly of the right arm.

Ms. M is divorced, lives alone, and works as a medical secretary. Four months ago, she marked the fifth anniversary of her daughter's death from a drug overdose at age 20. Her parents, whom she cared for, died within the last 3 years. Her son recently left home to attend graduate school, and she is estranged from the rest of her family. She endorses depressed mood and grief over her daughter's death but says she has no one with whom to talk. She also feels persistent guilt, as she was out on a date when her daughter tried to call home shortly before her death.

The limb movements and lack of coordination are increasingly interfering with Ms. M's life. She

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often uses her left hand to stop the right from moving and to guide it in simple tasks, such as opening doors. She can no longer hold a cup of coffee in her right hand or stand on stools at work to reach overhead shelves. At presentation, Ms. M's imbalance and involuntary movements are so severe that she cannot walk. A coworker drove her to the ER.

Ms. M's presenting symptoms suggest:

a) multifocal intracerebral process (such as ischemia, demyelination, toxic-metabolic problem, or prion disease)

b) paraneoplastic syndrome

c) conversion disorder

The authors' observations

A neurologist who evaluates Ms. M in the ER is concerned about her vertical nystagmus, which, unlike horizontal nystagmus, is almost always pathologic. The neurology service admits her for further evaluation.

Ms. M's age, recent normal MRIs, physical presentation, and lack of other findings suggest a paraneoplastic syndrome. Ataxia associated with subacute cerebellar degeneration can indicate an occult malignancy and is closely linked to gynecologic and breast cancers. Cerebellar degeneration often begins with loss of coordination, can be unilateral, and can appear as intention myoclonus.[1]

Also considered are:

- opsoclonus-myoclonus, which presents with ataxia, myoclonus, and random chaotic eye movements. This paraneoplastic disorder is less common in adults than in children, however.[1]
- alien hand/limb syndrome, in which the limb unintentionally performs seemingly purposeful movements, often prompting the patient to restrain the limb with the other hand. This syndrome, however, usually localizes to a lesion in the medial frontal lobe or corpus callosum. Ms. M's brain MRIs show no such lesion.

The psychiatry consult/liaison (C/L) service is asked to assist with Ms. M's care because of her prior conversion disorder diagnosis and her ongoing grief, depression, and anxiety.

How should the C/L team evaluate Ms. M?

a) perform basic imaging studies

b) perform extensive neurologic evaluation

c) assume conversion disorder with no further evaluation

TREATMENT SEARCHING FOR ANSWERS

We order an extensive neurologic workup for Ms. M, focusing on causes of inherited and acquired ataxias. The evaluation includes:

- brain and cervical spine MRIs to check for focal cerebral and spinal lesions
- EEG to search for seizure activity and slowing characteristic of encephalopathies
- urine heavy metal testing for toxic processes
- thyroid-stimulating hormone testing for hypothyroid-associated ataxia.

We also measure serum copper and ceruloplasmin to rule out Wilson's disease, pyruvic acid and lactic acid to check for a mitochondrial disorder, vitamin E and gamma tocopherol to rule out ataxia with vitamin E deficiency and Bassen-Kornzweig syndrome (abetalipoproteinemia), and endomysial/gliadin antibodies to evaluate for gluten ataxia.

Paraneoplastic workup includes chest, pelvic, and abdominal CT; a gynecologic exam; and a mammogram. All results are negative or equivocal.

We also order blood tests for paraneoplastic antibodies, evidence of opsoclonus/myoclonus, and spinocerebellar ataxia genetic testing; and a CSF check for protein 14-3-3 levels suggestive of prion disease. These tests, run at specialized laboratories, take 4 to 6 weeks.

Ms. M remains hospitalized for 7 days for evaluation. Her movement problems persist, though they often abate when she is distracted. Her

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upbeat nystagmus appears intermittent. Her affect is diverse, often shifting between tearfulness and inappropriate laughter.

Based on interviews with Ms. M, the C/L team sees prominent depressive symptoms including marked difficulty sleeping, appetite loss, and excessive guilt over her daughter's death. She also seems indifferent towards her disabling motor symptoms.

The C/L team diagnoses Ms. M with chronic and acute adjustment disorder and major depressive disorder. She is initially hesitant to take another antidepressant but agrees to try mirtazapine, 15 mg nightly, to treat her depression, decreased appetite, and sleep problems. After 2 days, mirtazapine is increased to 30 mg nightly as she is tolerating it and is willing to try a higher dosage.

Possible differential diagnoses include:

a) conversion disorder

b) unidentified neurologic disorder

c) malingering or factitious disorder

The authors' observations

No neurologic or pathologic explanation is found for Ms. M's symptoms. Imaging reveals no lesions to explain her intermittent upbeat nystagmus, which localizes to the pons and caudal medulla.[2]

Conversion disorder. Ms. M, however, appears to meet DSM-IV-TR criteria for conversion disorder (*Box*), which is thought to result from intense psychological distress in persons who can only express such emotions somatically. Her complaints had specific precursors: she was newly separated from her son and had marked the anniversary of her daughter's death, which intensified her persistent mourning. We link both circumstances temporally to symptom onset. Also, lack of interest in her serious motor symptoms could be the "la belle indifference" typical of conversion disorder.

Ms. M, however, appears highly suggestible. Her physical symptoms improve soon after her attending psychiatrist suggests that treating her depression will decrease her movements. The neurologists also notice day-to-day fluctuations in her gait disturbance and jerking movements. Distraction techniques produce objective improvement in both symptoms.

Is Ms. M faking her symptoms? Mental disorders and medical field employment both increase the risk of factitious disorder.[3] In caring for her parents, Ms. M often felt unappreciated and may be trying to enter the sick role that they had filled.

In malingering, the patient seeks external incentives for feigned behavior. The role of secondary gain must be considered, as Ms. M's illness has reunited her with her son, who visits her regularly at the hospital.

Ms. M's evaluation, however, uncovers no evidence that she is intentionally producing symptoms.

FOLLOW-UP THE ANSWER BECOMES CLEAR

One month after discharge to inpatient rehabilitation, Ms. M is readmitted to the neurology unit. Her uncontrollable limb jerks and ataxia are worse, and she appears demented and near mute. At that time, we learn that the CSF sample sent during her first admission is positive for protein 14-3-3.

Ms. M is diagnosed with Creutzfeldt-Jakob disease (CJD), a spongiform encephalopathy secondary to prion disease. She dies 6 days later. Sporadic CJD is confirmed at autopsy.

The authors' observations

The literature lists no comprehensive differential diagnosis for conversion disorder, probably

Box

DSM-IV-TR diagnostic criteria for conversion disorder

A. One or more symptoms or deficits affecting voluntary motor or sensory function that suggest a neurological or other general medical condition.

B. Psychological factors are judged to be associated with the symptom or deficit because the initiation or exacerbation of the symptom or deficit is preceded by conflicts or other stressors.

C. The symptoms or deficit is not intentionally produced or feigned (as in factitious disorder or malingering).

D. The symptom or deficit cannot, after appropriate investigation, be fully explained by a general medical condition, or by the direct effects of a substance, or as a culturally sanctioned behavior or experience.

E. The symptom or deficit causes clinically significant distress or impairment in social, occupational, or other important areas of functioning or warrants medical evaluation.

F. The symptom or deficit is not limited to pain or sexual dysfunction, does not occur exclusively during the course of somatization disorder, and is not better accounted for by another mental disorder.

Specify type of symptom or deficit:

With motor symptom or deficit
With sensory symptom or deficit
With seizures or convulsions
With mixed presentation

Source: *Diagnostic and statistical manual of mental disorders (4th ed-text rev)*. Copyright 2000. American Psychiatric Association. Reprinted with permission.

because presentations are diverse and the symptoms overlap with innumerable neurologic and medical conditions. This is underscored by the broad differential diagnosis for Ms. M's ataxia.

In a study to identify organic syndromes initially diagnosed as conversion disorder,[4] 10 of 85 patients (11.8%) were initially misdiagnosed and later found to have dyskinesia, amyotrophic lateral sclerosis, multiple system atrophy, extrapyramidal syndrome, multiple sclerosis, dementia, Parkinson's disease with psychogenic aggravation, lung cancer with cerebral metastases, and radicular syndrome. CJD and conversion disorder also share many symptoms (*Table, page 80*).

Correct diagnosis of conversion disorder calls for ruling out neurologic and medical conditions. Ms. M's upbeat nystagmus prompted aggressive neurologic evaluation. Although horizontal nystagmus has been reported rarely in conversion disorder,[5] vertical nystagmus has not. One case report[6] describes vertical nystagmus as the first clinical sign of CJD.

Leading clinical symptoms of CJD include progressive dementia, myoclonus, cerebellar ataxia, visual problems, and extrapyramidal signs.[7] Ms. M's uncontrollable movements and jerks, although not classically myoclonic, were similar to this common finding. She did not present with dementia, but her rapidly progressive end-stage mental status changes were characteristic of CJD.

Sporadic CJD accounts for 84% of transmissible spongiform encephalopathies. Genetic, iatrogenic, and variant CJD forms (linked to bovine spongiform encephalopathy, or "mad-cow disease") account for other cases.[8] Psychiatric symptoms are a more-common manifestation of variant CJD[9] but have been reported in sporadic CJD.[10]

Eventually, Ms. M's upbeat nystagmus, persistent abnormal movements, rapidly progressive dementia, and elevated CSF protein 14-3-3

Table

Conversion disorder, sporadic Creutzfeldt-Jakob disease share many symptoms

Complaint	Conversion disorder	Sporadic CJD
Paralysis	May not follow motor pathways	No
Myoclonus	Yes	Cardinal manifestation
Ataxia	May be bizarre in character	Present in 25% to 30% of patients, reflecting multiple disease subtypes
Hyperreflexia	No	Yes (40% to 80% of patients)
Dysphagia	Yes	No
Vomiting	Yes	No
Aphonia	Yes	No
Diplopia	Yes	Rare
Nystagmus	Rare	Yes
Blindness	Hysterical blindness detectable by ophthalmologic examination	Rare
Deafness	Yes	Rare
Anesthesia	Yes	No
Paresthesia	Yes	No
Depression	Yes	Yes
Other psychiatry diagnoses	Yes	More common in variant CJD
Progressive dementia	No	Cardinal manifestation
Temporal relationship with stress	Yes	No
Left-side symptoms more common	Yes	No

made the CJD diagnosis. Protein 14-3-3 is 94% sensitive and 84% specific for diagnosing CJD.[11] Ms. M's EEG findings did not suggest CJD, but these findings are less sensitive and occur later than the CSF findings.[11]

Finally, conversion disorder is almost always acute, not slowly progressive as with Ms. M.

GETTING THE DIAGNOSIS RIGHT

DSM-IV-TR criteria state that conversion disorder symptoms cannot be otherwise explained "after appropriate investigation," but what constitutes "appropriate" is unclear. Extensive inpatient evaluation eventually produced the correct diagnosis for Ms. M, but such a detailed evaluation may be too expensive and expansive for every patient with conversion disorder symptoms.

In the conversion disorder study,[4] the 10 misdiagnosed patients received one to eight supplemental diagnostic techniques before being correctly diagnosed. In five of the patients, however, the general neurologic examination was identi-

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fied as the diagnostic technique responsible for final diagnosis.

These findings suggest that a neurologic examination is key to evaluating complaints that suggest conversion disorder and to identify neurologic conditions. The results can also suggest somatic disorders, as exam findings will reflect patients' perceptions of neurologic processes. For example:

- patients with conversion motor symptoms may have tonic contractures of antagonistic muscles to "paralyze" certain joints
- those with conversion sensory symptoms rarely have sensory impairments that follow known innervation patterns.

When exam results suggest a neurologic process rather than conversion disorder, the workup must target the abnormal findings. Determine the need for evaluation on a case-by-case basis.

Motor complaints, such as localized paralysis or abnormal movements, should be evaluated with a brain MRI to look for lesions along the motor or cerebellar tracts. Sensory pathways can be further investigated with brain MRI and the relevant evoked potential(s) (visual, brainstem, or somatosensory).

Order EEG for patients with convulsions, particularly prolonged EEG monitoring with a video component, and measure serum prolactin immediately after an episode. In some cases, the neurologic exam alone or in conjunction with these initial studies can make the diagnosis. If the clinical situation warrants, more-detailed evaluations may be necessary.

Bottom Line

Order a multidisciplinary assessment for patients who meet DSM-IV-TR criteria for conversion disorder to rule out other medical and neurologic conditions. Abnormal neurologic findings should prompt further evaluation. Reconsider the diagnosis regularly, particularly if symptoms progress.

Related resources

- Wise MG, Rundell JR. *Clinical manual of psychosomatic medicine: a guide to consultation-liaison psychiatry*. Arlington, VA: American Psychiatric Publishing; 2005.
- National Institute of Neurological Disorders and Stroke. Creutzfeldt-Jakob disease fact sheet. www.ninds.nih.gov/disorders/cjd/detail_cjd.htm.

DRUG BRAND NAMES

Duloxetine • Cymbalta

Mirtazapine • Remeron

DISCLOSURE

The authors report no financial relationship with any company whose products are mentioned in this article, or with manufacturers of competing products.

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