Unvaccinated & Stuporous
Catatonia as a Clinical Feature of Subacute Sclerosing Panencephalitis

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March 23, 2019
Northern California Psychiatric Society
Disclosures

Relevant Financial Relationships
None

Off-Label Usage
None
Learning Objectives

1. Identify the main **characteristics** of catatonia

2. List three **differential diagnoses** for underlying etiology of catatonia as it pertains to this case

3. Recognize when & how to diagnose **subacute sclerosing panencephalitis**, which can present with catatonia
Case Presentation: History of Present Illness

- 40-year-old male Filipino immigrant with no psychiatric history
- Chief complaint: memory problems and falls for 3 months
- Reported severe anxiety, depression, amotivation, fatigue, alogia, auditory hallucinations, and thoughts of suicide
  - Started on risperidone recently for auditory hallucinations
- Rapid decline in functioning over 3 months (subacute)
- Had measles as a child in the Philippines
Case Presentation: Examination

- Mental status exam: mutism, stupor, rigidity & waxy flexibility, ambitendency, automatic obedience, negativism, withdrawal from eating and drinking; auditory hallucinations
- Frontal release signs: palmar grasp and palmomental reflex
- Neurological signs: frequent myoclonus, hyperekplexia, new left facial droop
- Vital signs are within normal limits
Case Presentation: 
Investigations

- Serum rubeola IgG positive, elevated titers; no IgM
- Cerebrospinal fluid:
  - Protein high, IgG index high, otherwise normal
  - Rubeola (measles) IgG level >300 mg/dL (high)
  - Creatinine Kinase (CK) levels elevated, down trending

Negative studies: CBC, BMP, HIV, RPR, blood cultures, heavy metals, ANA, ceruloplasmin, serum iron
Case Presentation: Magnetic Resonance Imaging
Electroencephalogram
Catatonia: Introduction

• Motor dysregulation disorder
  • Three forms: retarded, excited, malignant
• Catatonia is a syndrome, not etiological diagnosis
  • Failure to diagnose and investigate etiological causes can lead to significant morbidity and potential mortality
• Conceptualized as “playing dead” behavior in primitive mammals (e.g., Virginia opossum, duck-billed platypus)
### Catatonia: Diagnosis

**DSM-5 requires at least three of the following:**

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stupor</td>
<td>Not relating to external environment</td>
</tr>
<tr>
<td>Mutism</td>
<td>Minimal or no verbal response to most questions</td>
</tr>
<tr>
<td>Catalepsy</td>
<td>Passive induction of posture held against gravity</td>
</tr>
<tr>
<td>Waxy flexibility</td>
<td>Ease of movement after initial resistance when moving joint</td>
</tr>
<tr>
<td>Negativism</td>
<td>Opposition to or ignoring instructions. Stimulus-bound motor response.</td>
</tr>
<tr>
<td>Posturing</td>
<td>Spontaneous and active maintenance of posture against gravity</td>
</tr>
<tr>
<td>Mannerism</td>
<td>Odd, circumstantial caricature of normal actions</td>
</tr>
<tr>
<td>Stereotypy</td>
<td>Repetitive, too frequent non-goal-directed movements</td>
</tr>
<tr>
<td>Agitation</td>
<td>Too many movements without corresponding stimuli</td>
</tr>
<tr>
<td>Grimacing</td>
<td>Weird face expressions</td>
</tr>
<tr>
<td>Echolalia</td>
<td>Mimicking another’s speech</td>
</tr>
<tr>
<td>Echopraxia</td>
<td>Mimicking another’s movement</td>
</tr>
</tbody>
</table>

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Catatonia: Differential Diagnosis

20-25% are due to an additional medical condition

- Autoimmune: paraneoplastic/autoimmune encephalitis, SLE
- Toxic/metabolic: heavy metals, Wilson’s disease, porphyria
- Neurodegenerative: Dementia with Lewy Body, acute disseminated encephalomyelitis
- Infectious: Creutzfeldt-Jakob Disease, SSPE
- Iatrogenic: neuroleptic malignant syndrome (risperidone)
## Subacute Sclerosing Panencephalitis: Diagnosis

<table>
<thead>
<tr>
<th>Dyken Criteria</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Clinical</td>
<td>Progressive, subacute metal deterioration with typical signs like myoclonus</td>
</tr>
<tr>
<td>2. EEG</td>
<td>Periodic, stereotyped, high voltage discharges</td>
</tr>
<tr>
<td>3. CSF</td>
<td>Raised gamma globulin or oligoclonal pattern</td>
</tr>
<tr>
<td>4. Antibodies</td>
<td>Raised measles titers in serum ($\geq 1:256$) or CSF($\geq 1:4$)</td>
</tr>
<tr>
<td>5. Brain biopsy</td>
<td>Suggestive of panencephalitis</td>
</tr>
</tbody>
</table>
Case Presentation: Hospital Course

• Treated with benzodiazepines → improved catatonia

• Intrathecal catheter for interferon as treatment for SSPE

• Two months of treatment, eventual decline requiring ICU care for autonomic instability and vegetative state
  • Palliative comfort level care was decided by family

• Patient passed one day after allowing for natural death
Summary

40M with subacute anxiety, psychosis, then catatonic and neurological symptoms, investigation revealed SSPE

- Catatonia is a motor dysregulation syndrome requiring at least 3 characteristic signs to diagnose
- SSPE can present clinically with catatonia
  - Only through detailed history and through examination can underlying serious medical conditions such as SSPE be diagnosed and treated