Case Presentations: Body Integrity Identity Disorder

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Objectives:
• Define the condition: Body Integrity Identity Disorder (BIID)
• Discuss the neurological and psychological components of BIID pathophysiology
• Review the medical and psychiatric management as it relates to this case and considerations for all psychiatric trauma patients
On life and limb: One man’s chilling desperation to feel whole

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Objectives

• Define the condition: Body Integrity Identity Disorder (BIID)
• Discuss the neurological and psychological components of BIID pathophysiology
• Review the clinical features, diagnosis, and management of rhabdomyolysis.
• Review the medical and psychiatric management as it relates to this case.
Initial Evaluation

- Male, early 30s, transferred from OSH as Trauma 2 Activation
- Submerged LLE in dry ice x 7.5 hours
- Ultimate intention: amputation of leg

- “I have never identified with my left leg…”
- “I know this sounds crazy....I have a neurologic condition..”
- “I would have flown to Asia to have a surgeon do it, if I had the money.”
- Has never sought psychotherapy... “the only solution is removing my leg.”
Physical Exam

Vitals: BP 122/85, HR 106, RR 12, T 37.5 SpO2 100% RA
General: A&Ox3, Appearing non-toxic, NAD

LLE:
- Red demarcation 15 cm superior to patella
- Frozen solid, ischemic, mottled appearance
- No detectable pulse (doppler or palpation)
- No sensation or motor function

CT Angio:
- Normal femoral and popliteal arteries
- No flow distal to posterior tibial/peroneal arteries origins
Case Study
ED Assessment/Plan

• Ortho and Vascular Surgery Consulted
  – Options: 1. TPA/Heparin revascularization
             2. Vascular Surgery Intervention
             3. Guillotine Amputation per Ortho

• Leg non-salvageable
  – Plan: Above knee amputation
  – Passive Rewarming, Pain control, IV fluid resuscitation
  – Psychiatric consult
  – High Rhabdomyolysis Risk
Body Integrity Identity Disorder

Condition in which one does not identify with one part of his/her body, causing emotional distress, and often desire to amputate the healthy appendage.
Terminology

- Apotemnophilia
- Body Integrity Identity Disorder
- Xenomelia
History

• 1977 Money et al – First case report of two patients who desired amputation of healthy limb
  – Coined Apotemnophilia (paraphilia)

• 2000 Furth and Smith – “amputee identity disorder”
  – Disorder of body identity (not paraphilia)
  – Robert C. Smith, MD
    • 1st surgeon to electively amputate (1997, 1999)
2005 study surveying 52 participants with desire to amputate healthy limb

Aimed to answer key questions
- Characteristics
- Motivation
- Etiology
- Delusional component
- Treatment
Demographics

• 67% recruited from internet; 33% by other participants
  – Average age 48.6 years (23-77)
  – Men (n=47); Female (n=4)

• Age of onset childhood or adolescence

• Leg preference, above the knee most common
  – Left > right

• 90% higher level of education
Amputations/Attempts

- 27% obtained amputation
  - shotgun, chainsaw, wood chipper, dry ice
  - Elective surgical amputation

- Significant negative impact on life with risk of life-threatening injury or death
### Table 1. Reasons provided (in open-ended narrative) for wanting amputation

<table>
<thead>
<tr>
<th>Reason</th>
<th>Reported as primary reason (n=52)</th>
<th>Reported as secondary reason (n=48) (four subjects reported no secondary reason)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Restoring true identity as an amputee</td>
<td>63% (n=33)</td>
<td>10% (n=5)</td>
</tr>
<tr>
<td>Feeling sexually excited or aroused</td>
<td>15% (n=8)</td>
<td>52% (n=25)</td>
</tr>
<tr>
<td>The attention it draws</td>
<td>4% (n=2)</td>
<td>6% (n=3)</td>
</tr>
<tr>
<td>Body sculpting, aesthetics (interested in surgically changing fingers and toes because of superior aesthetic of the amputee look)</td>
<td>4% (n=2)</td>
<td>2% (n=1)</td>
</tr>
<tr>
<td>Feeling satisfied inside (increased sense of well-being)</td>
<td>2% (n=1)</td>
<td>29% (n=14)</td>
</tr>
<tr>
<td>To overcome adversity</td>
<td>2% (n=1)</td>
<td>2% (n=1)</td>
</tr>
<tr>
<td>Related to being sexually attracted to amputees – to know how it feels</td>
<td>2% (n=1)</td>
<td>2% (n=1)</td>
</tr>
<tr>
<td>To be ‘special’</td>
<td>2% (n=1)</td>
<td>0</td>
</tr>
<tr>
<td>My parents and others would accept me more</td>
<td>2% (n=1)</td>
<td>0</td>
</tr>
<tr>
<td>Makes me feel safe (reported as sequelae of childhood sexual abuse)</td>
<td>2% (n=1)</td>
<td>0</td>
</tr>
<tr>
<td>‘Legs look ugly’</td>
<td>2% (n=1)</td>
<td>0</td>
</tr>
</tbody>
</table>
Tell me how this makes you feel…

“I can feel exactly the line where my leg should end and my stump should begin. Sometimes this line hurts or feels numb.”

“I feel myself complete without my left leg… I’m over-complete with it.”

“[After the amputation] I would have the identity that I’ve always seen myself as.”
Differential Diagnosis

• **Psychotic Disorders** – *This is crazy, right?* Perhaps not.
  
  – BIID acknowledges the reality that the limb is theirs and has a functional purpose
  – Phantom limb pain occurs in non-psychotic patients
  – Delusional to believe amputation will resolve inner turmoil?
Differential Diagnosis

- **Body Dysmorphic Disorder:**
  - Repetitive behaviors/thoughts related to a preoccupation with perceived flaw in physical appearance

- **Factitious Disorder**
  - Feigning illness to achieve end goal of amputation vs to assume the “sick victim” role

- **Neurological Conditions**
  - “I know this sounds crazy, but I have a neurological condition.”
Parietal Lobe Involvement

- Parietal lobe Function: sensation, visual-spatial processing, and integration of sensory input
- R. Parietal Lobe damage = Contralateral neglect
- Insular Cortex = involved in the integration of bodily feelings, including ownership and sexual arousal
• Explanation for leg > arms predominance, left-sided preference, and specific line of demarcation?
  – ↓ activation of right superior parietal lobe of affected leg
  – Dysfunction of right anterior insula
BIID Treatment

- Psychotherapy
- Medication – SSRI
  - Fluoxetine
- Vestibular Caloric Stimulation
  - ↑ contralateral parietal cortex activity
- Deep Brain Stimulation
- Elective Amputation...?
Case Study
Hospital Course

- **24 hours of admission**
  - **Psychiatric consult pre/post op**
    - Not suicidal, delusional, or interested in additional self-harm
  - **Above the knee amputation**
    - Transferred to STICU post-op
    - Peak: CK 16k, lactate 1.8, Cr 1.52, WBC 28k
    - Oliguric (in ED)
    - Post-op bleeding at VAC site, Anemic, hypotensive
    - Returned to OR for revision and ligation of bleeders
Rhabdomyolysis

• Syndrome that develops through muscle necrosis and subsequent release of intracellular toxins into the systemic circulation

• Early recognition and treatment is key to mitigating renal dysfunction
Etiology

• Skeletal muscle damage
  – Trauma
    • crush injury
    • prolonged immobility
    • ischemia reperfusion (vascular injury)
    • electrical injury
  – Nontraumatic
    • Exertional
    • Infections (influenza, HIV, Epstein-Barr, bacterial)
    • Toxins (Snake venom, carbon monoxide, mushroom poisoning)
    • Hereditary (glycogen storage diseases – NMS, malignant hyperthermia)
Pathophysiology of Rhabdomyolysis

• Trauma -> muscle damage -> release of intracellular components
  – Myoglobin & Creatine Kinase
  – Potassium, phosphate, uric acid, lactate dehydrogenase

• Myocyte injury/ATP depletion
  – Inhibits Na⁺/K⁺ pump -> Na⁺/Ca²⁺ exchange -> ↑ intracellular Na⁺ and Ca²⁺
  – Excess intracellular Na⁺ promotes cellular swelling and further myocyte damage
  – Excess intracellular Ca²⁺ promotes sustained myofibrillar contraction and further ATP depletion
  – Lyses of cell membranes
Diagnosis

- High index of suspicion
- Clinical triad
  - Muscle aches
  - Weakness
  - Tea-colored urine
- Physical exam
  - Evidence of trauma
  - Evaluate extremities for compartment syndrome
  - Pressure necrosis of skin

- Laboratory studies
  - CK = most sensitive marker
  - UA dipstick + with few or no RBCS
  - Urine myoglobin +
  - Pigmented cast cells
  - Anion gap metabolic acidosis
Complications of Rhabdomyolysis

**Acute Kidney Injury**
Direct effect of myoglobin +/- hypovolemia
Rx: IV fluids, bicarbonate, hemodialysis

**Compartment Syndrome**
Muscle ischemia and fluid sequestration
Rx: Fasciotomy

**Disseminated Intravascular Coagulation (Late)**
Thromboplastin release and thrombotic microangiopathy
Rx: Fresh frozen plasma

**Hyperkalemia**
Potassium released from damaged muscles and decreased clearance from acute kidney injury
Rx: IV fluids, diuresis, Kayexalate, calcium gluconate, glucose-insulin, hemodialysis

**Hypovolemia**
From sequestration of fluids in the muscles
Rx: IV fluids

**Hyperphosphatemia**
Muscle breakdown
Rx: Diuresis, hemodialysis

**Hypocalcemia**
Inward flux and binding to phosphatidylinositol
Rx: Avoid giving calcium, as calcium likely to increase later without treatment

**Hypercalcemia (Late)**
Efflux from damaged muscles and slow clearance if acute kidney injury present
Rx: IV fluids, diuresis
Creatine Kinase

- Intracellular enzyme – stores energy for ATP
- 5 times upper limit of normal (1,000 U/L)
  - 500 U/L – 75,000 U/L
- Serum concentration rises in first 12 hours post-trauma, peaks at day 3, and normalizes around day 5.
- CK elevation correlates with severity of muscle injury, but use as a predictor for AKI remains unclear
Pathophysiology of Acute Kidney Injury

Acute Kidney Injury (AKI)

- Myoglobin release
  - Direct renal cytotoxicity
  - Vasoconstriction
  - Tubular obstruction
- Hypovolemia
  - Ischemia
CK utility in predicting AKI and mortality

- Single-center, retrospective analysis
- 6 year period, 202 patients
- CK >1,000 U/L
- ARF: serum creatinine > 4mg/dL, >0.5mg/dL per day, 3x baseline, UOP <0.3 mL/kg per hour, anuria x 12 hours
CK utility in predicting AKI and mortality

Results:
- Peak CK < 5000 U/L for 65% (133)
  - 9% (12) developed ARF
  - 1 patient required RRT
- Peak CK 5000 -10,000 U/L for 9% (19)
  - 31% (6) developed ARF
  - 0 required RRT
- Peak CK >10,000 U/L for 25% (50)
  - 22% (11) developed ARF
  - 8% (4) required RRT

Conclusion:
- Initial and peak values may not be a reliable predictor for AKI
- Peak CK >5,000 associated with increase risk of AKI
- Peak CK >10,000 associated with increased risk of RRT
- Degree of correlation between CK level and rhabdomyolysis outcome remain unclear
Management Goals for Rhabdomyolysis

- Prevent further skeletal muscle damage
- Prevent acute kidney injury
- Identify and treat life-threatening complications
- No RCTs with definitive guidance
Fluids, Fluids, Fluids

• **Volume resuscitation**
  – Mainstay for treating rhabdomyolysis and preventing AKI
  – Dilutes nephrotoxins
  – Prevent myoglobin accumulation in renal parenchyma
  – Goal UOP 2 - 3 ml/kg/hr (100 - 200 ml/hr)

• **Selecting fluid therapy**
  – Cho et al 2007, prospective randomized trial (28 patients)
    • Lactated Ringer vs Normal Saline @ 400ml/hr
    • No AKI in either group
    • NS group hyperchloremic metabolic acidosis
Urine Alkalization

• Sodium Bicarbonate
  – Prevent the precipitation of myoglobin in renal tubules
  – Decreases cast formation and tubular obstruction
    • Decrease acidic environment
    • Goal urine pH >6.5
  – Benefits of alkalinization in animal studies with sodium bicarb compared to isotonic IVF alone

• Benefit of urine alkalinization in human clinical trials?
Mannitol

- **Proposed benefits**
  - Induces osmotic diuresis
  - Free radical scavenger
    - Renal protection from myoglobin
  - Decreasing compartment pressures

- **Potential risks**
  - Osmotic nephrosis
    - Large accumulated doses
    - Renal vasoconstriction
    - Tubular toxicity

- **No clear benefit in patients with rhabdomyolysis**
• 2004 retrospective study
• 2,083 trauma ICU patients
• No difference in rates of RF, need for RRT, or mortality between bicarbonate/mannitol group vs isotonic fluid administration among pts with CK >5,000 U/L
  – Some indication of benefit in pts with CK >30,000 U/L
Additional Management – electrolyte abnormalities

- **Hyperkalemia**
  - Check serum $K^+$ often
  - EKG
  - Calcium, insulin/glucose, sodium bicarb, kayexalate, hemodialysis

- **Hypocalcemia (early)**
  - Only correct if symptomatic (tetany, arrhythmias) or if hyperkalemic

- **Hypercalcemia (late)**
  - IVF, consider diuresis

- **Hyperchloremia**
  - Consider switching from NS to LR or adding sodium bicarbonate

- **Hyperphosphatemia**
  - IVF, diuresis, phosphate binders
Summary - Rhabdomyolysis

Pathophysiology
- Direct renal cytotoxicity
- Vasoconstriction
- Tubular obstruction

Evaluation
- Determine volume status
- Serial CK, BMP
- CK 5 times upper limit of normal (1,000 U/L)

Management
- Volume resuscitation
  - NS or LR
  - Consider sodium bicarb in addition to NS

- No RCTs demonstrate improved mortality with bicarb or mannitol
- Goal UOP 2-3 ml/kg/hr
- Monitor electrolytes
- EKG
- Monitor for complications
  - AKI
  - Hyperkalemia
  - Compartment syndrome
  - DIC
Case Study
Disposition

- PTD 5: D/C to inpatient rehab
  - Ortho and Psych continue to follow
  - Phantom Limb pain
- PTD 12: Discharged Home
  - Crutches, wheelchair
  - Denies need for psych follow-up
  - Return to graduate program
- Follow-up
  - Fitted for prosthesis
  - Physical Therapy
  - Phantom Limb pain persists
  - Remains full-time student
Thank You
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