INTRODUCTION

• This case involves a patient presenting with a chief complaint of pain with movement of his extremities.
• It should remind us how important it is to know the classic cutaneous presentations of certain diseases.
• We will also highlight the importance of post-diagnostic evaluation in preventative healthcare.
• 32-year-old previously healthy Hispanic male
• Chief complaint- progressive muscle pain/weakness in all four extremities and a rash
• The rash began approximately two weeks ago on the bridge of his nose and cheeks.
• Two days later, the rash had progressively spread to the rest of his face, down his neck, across his upper back and on his chest.
• By the end of the week, the rash had progressed down both of his arms and he began to experience a dull aching pain in his proximal legs bilaterally.

• The deep aching pain progressed down both of his lower extremities and eventually spread to both upper extremities.
• The pain was exacerbated by any movement of his arms or legs, and the patient described that he experienced excruciating 10/10 pain by simply raising his arm off the bed.
• He was finally prompted to come to the hospital when it was too painful for him to go to the bathroom independently.
• The patient tried taking extra strength Tylenol which provided minimal, if any, relief of his symptoms. Respite from the pain was only achieved while he was completely immobile.
• The patient denied any recent illnesses, sick contacts or travel.
PAST MEDICAL HISTORY

• No past medical history
• Patient was previously healthy

PAST SURGICAL HISTORY

• No surgical history.
FAMILY HISTORY

- Father: Diagnosed with prostate cancer, died of natural causes at 78
- Mother: Alive and Healthy at age 84
- No Siblings

SOCIAL HISTORY

- Patient has never smoked or used any illicit drugs
- Drinks one or two beers per week with dinner
- Married, Lives with his wife, No children
- Sexually Active, Monogamous with wife
- Works from home as a cartoon animator
MEDICATIONS

• Extra Strength Tylenol 2 Caplets by mouth Q6 hours prn for pain, not exceeding 6 caplets in 24 hours, for the past 2 weeks
• No other Medications

ALLERGIES

• No Known Drug Allergies
REVIEW OF SYSTEMS

• General: denies weight loss and fatigue, chills, night sweats, malaise
• HEENT: denies headache, vision changes, dizziness, difficulty swallowing
• Cardiovascular: denies chest pain, palpitations, edema, syncope
• Respiratory: Denies any SOB, wheezing, cough
• GI: denies nausea, vomiting, hematemesis, dyspepsia
• GU: Denies hematuria, dysuria, polyuria, nocturia

REVIEW OF SYSTEMS CONTINUED

• Extremities: Admits to muscle weakness and pain exacerbated with movement in all four extremities. Denies edema of the upper or lower extremities.
• Skin: Admits to Rash on his face, neck, upper back, and upper extremities
• Neurologic: Denies any numbness or tingling. No loss of sensation.
• Lymphatic: Denies lumps or swollen lymph nodes
• Hematologic: Denies easy bruising or excessive bleeding
PHYSICAL EXAM

• Vitals: 132/88 (supine, left arm), 72 bpm, 98.8F, 18 breaths/min, 100% on room air
  • Wt: 130 lbs Ht: 5’11 BMI: 31.5 - obese
• General: Patient is laying in hospital bed supine with his eyes closed.
• HEENT: Normocephalic, atraumatic, mucous membranes moist, PERRLA. extraocular muscles intact, normal conjunctiva, sclera anicteric, no papilledema, hemorrhages or A-V nicking on retina, dentition normal
• Neck: Supple, non-tender, no rigidity, no carotid bruits, no JVD, no lymphadenopathy, no thyromegaly or thyroid nodules

PHYSICAL EXAM

• Cardiovascular: Regular rate and rhythm, normal S1, S2, no S3, S4, no murmurs. PMI is non-displaced.
• Pulmonary/Thorax: Lungs are clear to auscultation bilaterally. Good air movement, no wheezes or crackles noted, respirations non-labored, chest expansions symmetrical
• Abdomen: Soft, non-distended and non-tender to superficial and deep palpation, no rebound, no guarding, liver edge not palpable, no hernias or scars, no abdominal masses, normal bowel sounds x 4
PHYSICAL EXAM

- Extremities: no clubbing, cyanosis or edema, pulses: +2 radial, posterior tibial, dorsalis pedis bilaterally. Muscle strength 3/5 bilaterally in upper and lower extremities.
- Neurologic: Alert and Oriented x3 to person, place and time. CN II-XII intact.. Triceps, biceps, patellar, and Achilles reflex +2/4 bilaterally. Babinski absent. No difficulty speaking.
- Lymph: No anterior or posterior cervical lymphadenopathy
- Skin: Normal turgor, no cyanosis, capillary refill <3seconds, Rash on following slides
- Osteopathic Findings – Joints were stiff and produced audible “pops” when manipulated while testing passive range of motion. The cervical spine and upper thoracics were warm and boggy bilaterally upon palpation.
LABORATORY FINDINGS - CBC

- WBC: $7.3 \times 10^9/L$
- Hgb: 15.2 g/dL
- RDW: 12.0%
- MCV: 96 fL
- MCHC: 30g/dL
- Platelet: $283 \times 10^9/L$
LABORATORY FINDINGS - BMP

- Fasting Glucose: 98 mg/dL
- Sodium: 141 mmol/L
- Potassium: 4.2 mmol/L
- Chloride: 99 mmol/L
- Bicarbonate: 26 mmol/L
- Magnesium: 2.1 mEq/L
- Calcium: 9.7 mg/dL
- Phosphate: 1.0 mg/dL
- BUN: 20 mg/dL
- Cr: 1.01 mg/dL
- eGFR: >60 mL/min

LABORATORY FINDINGS

- Creatinine Kinase: 8,363 units/L
- Lactate Dehydrogenase: 402 units/L
- Troponin: <.02 ng/mL
- TSH: 0.68 micro-int.units/mL
LABORATORY FINDINGS

• ANA – Negative
• Anti-Jo1 – Negative
• Anti-dsDNA – Negative
• Sclrdm70 Ab – Negative
• RNP Ab – 0.3
• West Nile Serology – Negative
• HIV – Negative
• Hepatic Panel – Negative

LABORATORY FINDINGS

• Urinalysis – Negative for protein, leukocyte esterase, nitrites, glucose, ketones, blood, casts, urobilinogen
• UA pH – 7.0
• UA Specific Gravity – 1.010
• Myoglobin urine – Negative
LABORATORY FINDINGS

• Imaging
  • MRI of Upper and Lower Extremities revealed heterogeneous distribution of edema throughout muscle bellies bilaterally highly suspicious of rhabdomyolysis
  • No evidence of compartment syndrome

ASSESSMENT

• Inflammatory Myopathy
  • Suspect Dermatomyositis
  • Must rule out Motor Neuron disease, Myasthenia Gravis, Muscular Dystrophies, Infectious myopathies, and drug-induced myopathies

• Rhabdomyolysis
  • Secondary to Inflammatory Myopathy
  • Obese
PLAN

• Consult Surgery for Muscle Biopsy
• Continue Extra Strength Tylenol
  • Extra Strength Tylenol 2 Caplets by mouth Q6 hours prn for pain, not exceeding 6 caplets in 24 hours
• Begin Pulse Dose IV Methylprednisolone in Hospital
  • Solumedrol 1,000 mg = 1 vial, IV Piggyback Daily
  • Trend CK values while in hospital, discharge with significant down trending

PLAN CONTINUED

• Send home with Prednisone
  • Prednisone 20 mg tablet – three tablets orally per day for a total of 60mg daily
• Follow up in one week for results of muscle biopsy with repeat labs to trend CK
• Plan for cancer screening beginning with EGD & Colonoscopy
DISCUSSION

- Dermatomyositis (DM) is an autoimmune inflammatory myopathy.
- The gold standard diagnosis for DM is a muscle biopsy in patients with symmetrical proximal muscle weakness, elevated muscle enzymes, and a rash specific for dermatomyositis (described further on next slide).
- Muscle biopsies are obtained from a muscle that is weak on physical exam with either an open biopsy or a closed-needle biopsy, location of muscle biopsy may also be determined with MRI of extremities that reveal evidence of rhabdomyolysis.
  - Some tissue is fixed for routine light and electron microscopy.
  - Other tissue is frozen and prepared for immunohistologic assays for mutant proteins. Including: dystrophin for Duchenne/Becker dystrophy, Merosin for congenital muscular dystrophy, and Sarcoglycan for limb-girdle muscular dystrophy.

DISCUSSION

Classic Manifestations of Dermatomyositis

- Gottron’s papules are erythematous to violaceous papules that occur symmetrically over extensor (dorsal) aspects of the MCP and PIP joints. The lesions are often associated with scaling and may ulcerate. Gottron’s sign is similar, but refers to lesions on extensor surfaces of joints in sites other than the hands.
- Facial erythema in a malar distribution that may mimic SLE. In contrast to SLE, this rash will often include involvement of nasolabial fold.
- Photodistributed Poikiloderma including the Shawl Sign (upper back) and V sign (across the neck between the clavicles).
Gottron’s sign

Shawl Sign

V sign

Facial Erythema in malar distribution

Gottron’s sign

AFTER 5 DAYS OF HIGH DOSE STEROIDS
OSTEOPATHIC CONSIDERATIONS

• Dermatomyositis often causes proximal muscle weakness in the extremities, which can be addressed osteopathically.
• Combined aerobic exercise and strength training may have slight increases in muscle strength and aerobic capacity for people with dermatomyositis.
• In addition to physical therapy and exercises at home, patients may benefit from myofascial manipulations.

MALIGNANCY ASSOCIATIONS

• A 2017 systematic review and meta-analysis, in up to 20% of cases, dermatomyositis appears as a paraneoplastic syndrome associated with multiple malignancies such as ovarian, breast, prostate, lung, nasopharyngeal and colorectal cancer.
MALIGNANCY ASSOCIATIONS

• In a 2018 systematic review, DM manifested before the diagnosis of colorectal cancer in 21 out of 27 patients and had immediate improvement of DM symptoms after surgery.

MALIGNANCY ASSOCIATIONS

• A 2014 systematic review and meta-analysis identified the factors in DM patients that would increase their risk of malignancy:
  • Age >45
  • Male Sex
  • Cutaneous necrosis
  • Cutaneous vasculitis
DISCUSSION

Other Post Diagnostic Evaluations

- Cardiac Involvement- patients with clinical suspicion of cardiac muscle involvement such as signs or symptoms of heart failure or conduction abnormalities, may need to obtain an echo and ECG.
- Esophageal Dysfunction- patients with esophageal dysmotility or dysphagia, esophageal motility studies should be performed.
- Pulmonary Disease- DM has been associated with interstitial lung disease so patients with suspected pulmonary involvement with dyspnea or cough may require pulmonary function testing and a CT scan of the chest.
  - If patient does have interstitial lung disease = reduced chance of malignancy and vice versa.

CONCLUSION

- Every form of dermatomyositis presents with cutaneous findings, but not all cases present with myopathies.
- Muscle biopsy is needed for confirmatory tissue diagnosis
- If presenting with purely cutaneous findings, DM malar rash can be distinguished from lupus because it does not spare the nasolabial folds, and patients will also have classic signs such as Gotron’s papules, shawl sign, etc.
- It is paramount that a malignancy workup be performed and patient adequately screened for cancer, specifically colonoscopy.
- Specific Auto-antibodies can determine risk of certain malignancies and may also be used to predict prognosis.
REFERENCES

- Patchett DC, Grover ML. Mitochondrial myopathy presenting as rhabdomyolysis. JAOA 2011; 111:404-405.

ACKNOWLEDGMENTS

- Dr. O'Shaine Brown, M.D., PGY1, was my supervising internal medicine resident at Abrazo West Hospital while I was a student on service and assigned to this patient.