Case Discussion 2

Dermatomyositis

History of present illness:

A 60 year old male came with bilateral pain in his proximal upper and lower extremities with inability to comb his hair, climbing stairs or getting up from the chair.

Signs and symptoms

These symptoms started 4 months ago when the patient initially complained of having slowly progressive inability to climb stairs and comb his hair. These symptoms were associated with pain over the thighs and arms, with similar severity throughout the day (24 hours steady pain). In addition, the patient noticed hyperpigmentation over his face for the past 2 months along with previously complaint of fatigue and weight loss for the past 9 months. A history of constipation was also documented by the patient for one year period.

There is no history of trauma, ptosis, seizures, spasticity, muscle twitches, urinary incontinence, edema, hypertension or hematuria.

The patient is not smoker, and has no significant past medical or surgical history.

Physical examination

The patient is pale, conscious, oriented and not jaundiced.

The blood pressure measures 130/80, temperature is 37, and heart rate is 70.

The patient has pigmentation of upper eyelids (heliotrope rash), and over his knuckles (Gottron papules).

Examination of the trunk revealed erythema of the anterior chest “V-neck sign” and the upper back and shoulders “shawl sign”.

CNS and muscle examination: Tone is normal. Power is decreased mainly at shoulders and hips (quadripareisis). Hand grip is normal. Sensation and tendon reflexes are maintained.

No significant findings in other systems.

Lab investigations

-CBC: Hb: 9 mg/dL, MCV: 70 fL, MCHC: 29

-Creatine kinase (CK): 1000 IU/L (normal: 60-174)
-ANA (antinuclear antibody): elevated titer

-Anti-Mi-2, anti-Jo-1…etc. (myositis-specific antibodies (MSAs)): elevated

**EMG study**
Myopathic changes

**Muscle biopsy**
- Shrinkage (atrophy) of fibers near borders of fascicles
- Inflammatory cells around fascicles and between fibers
- Perivascular cuffing by inflammatory cells

**Histopathological differential diagnoses**
- Polymyositis
- Inclusion body myositis

**Types of dermatomyositis**
Juvenile and adult types

**Epidemiology**
The most common inflammatory myopathy in children
In adults about 20% has underlying malignancy: = paraneoplastic syndrome
Better prognosis in children

**Pathogenesis**
- Systemic autoimmune disease
- Deposition of complement attack complex in small blood vessels
- Type I interferon secretion
- Autoantibodies and lymphocytic inflammation

**Other skin manifestations**
- Erythematous lesions in photodistribution
- Pruritus
- Erythema of face
- Nail changes
- Hair loss

*Muscle and skin manifestations may occur together or one before the other

**Systemic manifestations**
- Fever, malaise and weight loss

**Other manifestations**
- Raynaud phenomenon
- Dysphagia and GERD in 30% of the cases

**Other organs that may be involved**
- Joints
- Esophagus
- Heart …more in adults form
- Lungs …interstitial lung disease in 10% …more in adults form

**Differential diagnoses**
- Neurological deficit
- SLE
- Myasthenia gravis
- Polymyositis
- Inclusion body myositis

**Therapy for muscle disease**
Corticosteroids and immunosuppressive agents

**Therapy for skin disease**
- Sun avoidance
- Sunscreens and photoprotective clothing
-Topical corticosteroids

-Antimalarial agents

-Immunosuppressive agents

Conclusion

-Our patient is old

-He has constipation

-He has anemia...he may lose blood in his stool

**Do colonoscopy for him. He may have colon cancer associated with dermatomyositis (paraneoplastic syndrome)**