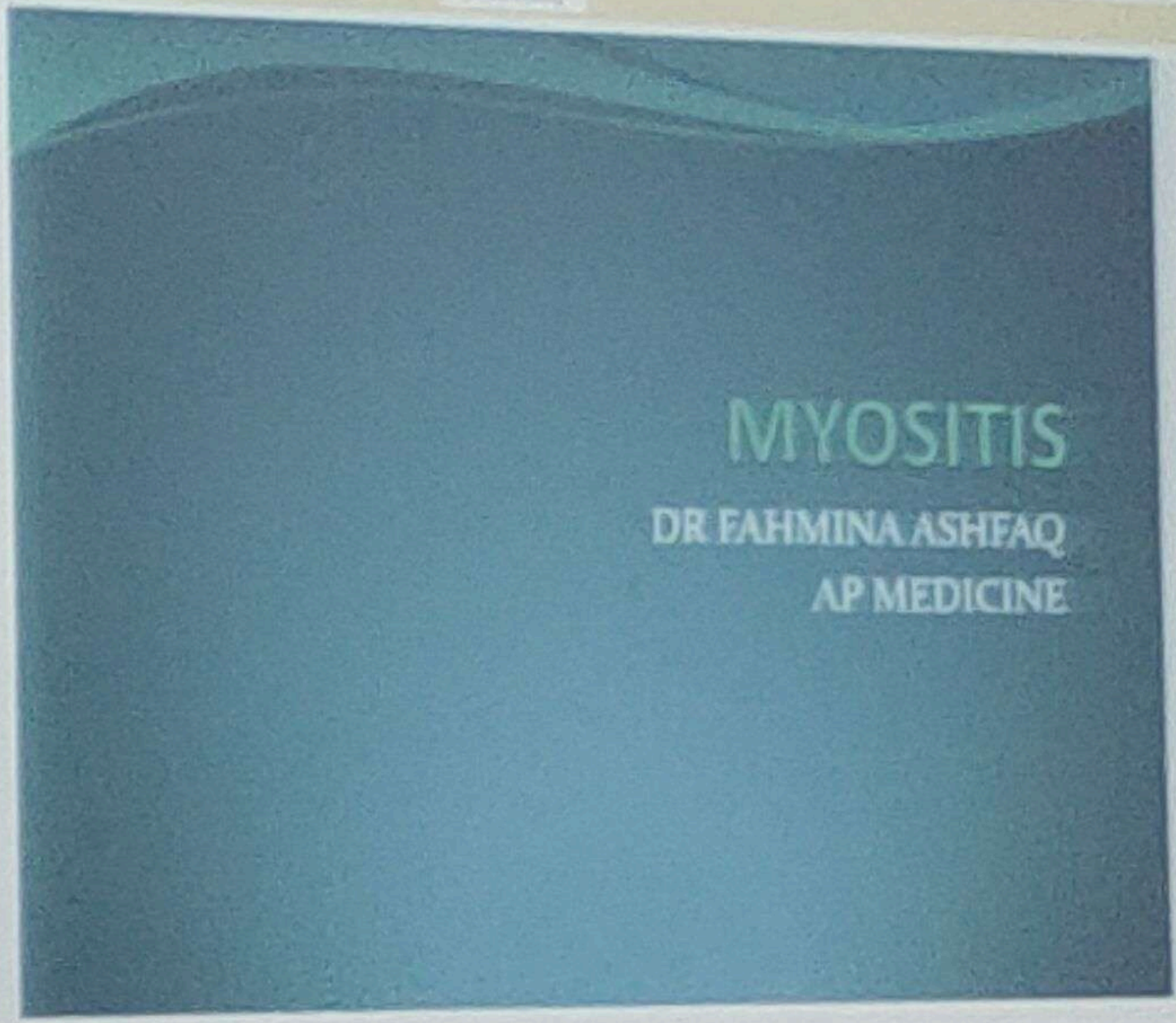
MYOSITIS
DR FAHMINA ASHFAQ
AP MEDICINE

POLYMYOSTIS:

Polymyositis is an idiopathic inflammatory myopathy characterized by the following

- Symmetrical, proximal muscle weakness
- Elevated skeletal muscle enzyme levels
- Characteristic electromyography (EMG) and muscle biopsy findings.

DERMATOMYOSITIS

- Clinically similar to polymyositis, dermatomyositis is an idiopathic inflammatory myopathy associated with characteristic dermatologic manifestations.

INCLUSION BODY MYOSITIS

- Inclusion body myositis is a slowly progressive, idiopathic, inflammatory myopathy with characteristic pathologic findings that is generally found in older men

NAM

- Necrotizing autoimmune myopathy (NAM) is a recently recognized form of idiopathic inflammatory myopathy that is identified by finding macrophage-predominant myocyte destruction, with few to no lymphocytes, on muscle biopsy. NAM has been associated with malignancy and statin use

polymyositis

CAUSATIVE AGENTS:

- human retroviruses
- human immunodeficiency virus (HIV)
- human T-cell lymphotrophic virus type I (HTLV-I)
- the simian retroviruses
- coxsackievirus B.

SYMPTOMS:

- Symmetrical, proximal muscle weakness with insidious onset
- Pain is not a predominant symptom
- Dysphagia (30%) and aspiration, if pharyngeal and esophageal muscles are involved
- Arthralgias may be associated
- Difficulty kneeling, climbing or descending stairs, stepping onto a curb, raising arms, lifting objects, combing hair, and arising from a seated position

- Weak neck extensors cause difficulty holding the head up
- Involvement of pelvic girdle usually greater than upper body weakness
- Cardiac involvement may cause symptoms of pericarditis or cardiomyopathy
- Characteristic rash on face, trunk, and hands seen in dermatomyositis only

CONSTITUTIONAL SYMPTOMS:

- Morning stiffness
- Fatigue
- Anorexia
- Fever (associated with antisynthetase antibodies such as anti-Jo-1)
- Weight loss

PULMONARY MANIFESTATIONS

- Pharyngeal and esophageal weakness may lead to aspiration pneumonia.
- Patients with polymyositis may experience exertional dyspnea secondary to weakness of chest wall muscles and diaphragmatic muscles.
- Patients receiving immunosuppressants are at an increased risk of infection.

- Interstitial lung disease occurs in 5-30% of patients
- Interstitial pneumonitis, bronchiolitis obliterans organizing pneumonia, and pulmonary capillaritis have been described in conjunction with polymyositis.

CARDIAC MANIFESTATIONS

- Rhythm disturbances, conduction defects, congestive heart failure, pericarditis, pulmonary hypertension, and myocarditis can occur.

JOINTS MANIFESTATIONS

- Arthritis is usually symmetrical and involves the knees, wrists, and hands (associated with antisynthetase antibodies)

GI MANIFESTATIONS

- Dysphagia
- Odynophagia
- Nasal regurgitation
- Reflux esophagitis
- Abdominal bloating
- Constipation

COMPLICATIONS

- Pulmonary (eg, aspiration pneumonia, interstitial lung disease, hypoventilation)
- Increased incidence of malignancy in dermatomyositis as well as polymyositis
- Dysphagia, weight loss, and malnutrition
- Ambulatory dysfunction
- Adverse effects of immunosuppressive therapy

DIAGNOSIS:

- Complete blood count (CBC) - leukocytosis or thrombocytosis;
- Erythrocyte sedimentation rate or C-reactive protein level - Elevated
- Elevated muscle enzyme levels
- Myoglobinuria
- Autoantibodies
- Positive rheumatoid factor - Found in more than 50% of patients

MUSCLE ENZYMES

- Serum creatine kinase (CK) levels are usually elevated in persons with polymyositis, ranging from 5-50 times the reference range.
- Lactate dehydrogenase
- Aspartate aminotransferase
- Alanine aminotransferas

ANTIBODY FINDING

- Antisynthetase antibodies (such as anti-Jo-1 antibodies) - antisynthetase syndrome may manifest as idiopathic inflammatory myopathy, interstitial lung disease, arthritis, Raynaud phenomenon, fever, and/or mechanic's hands
- Signal-recognition particle (SRP) antibodies - Approximately 4% of patients with polymyositis have antibodies to signal recognition particles (SRPs), which are associated with acute onset of severe weakness, increased incidence of cardiac involvement, and higher mortality rates.
- Anti-HMGCR autoantibodies in statin-induced auto immune myopathy

ELECTROMYOGRAPHY

- Evidence of membrane irritability, increased insertional activity, fibrillation potentials, positive sharp waves at rest
- Myopathic changes of motor unit action potential; decreased amplitude and duration; increased polyphasic potentials; bizarre, high-frequency, repetitive discharge

MUSCLE BIOPSY

- Muscle biopsy (eg, deltoid or quadriceps femoris) is crucial in helping to diagnose polymyositis and in excluding other rare muscle diseases.
- Muscle biopsy shows muscle fibers in varying stages of inflammation, necrosis, and regeneration

TREATMENT

- Prednisone is the first-line treatment of choice for polymyositis. Typically, the dose is 1 mg/kg/day, either as a single or in divided doses. This high dose is usually continued for 4-8 weeks, until the creatine kinase (CK) level returns to reference ranges.

- Immunosuppressive agents are indicated in patients who do not improve with steroids within a reasonable period (ie, 4 wk) or in whom adverse effects from corticosteroids develop

- Intravenous immunoglobulin (IVIG) has been used for the short-term treatment of steroid-resistant cases of polymyositis.