Idiopathic Inflammatory Myopathies

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KEYWORDS

• Polymyositis • Dermatomyositis • Necrotizing myopathy • Inclusion body myositis
• Clinical presentation • Diagnosis • Pathology • Treatment

KEY POINTS

- The idiopathic inflammatory myopathies (IIM) consist of rare heterogeneous autoimmune disorders that present with marked proximal and symmetric muscle weakness, except for distal and asymmetric weakness in inclusion body myositis (IBM).
- Besides frequent creatine kinase (CK) elevation, the electromyogram confirms the presence of an irritative myopathy.
- Extramuscular involvement affects a significant number of cases with interstitial lung disease (ILD), cutaneously in dermatomyositis (DM), systemic or joint manifestations, and increased risk of malignancy, especially in DM.
- Myositis-specific autoantibodies influence the phenotype of the IIM. Jo-1 antibodies are frequently associated with ILD and the newly described HMG-CoA reductase antibodies are characteristic of autoimmune necrotizing myopathy (NM).
- Muscle abnormality ranges from inflammatory exudates of variable distribution to intact muscle fiber invasion, necrosis, phagocytosis, and, in the case of IBM, rimmed vacuoles and protein deposits.
- Despite many similarities, the IIM are fairly heterogeneous from the histopathologic and pathogenetic standpoints, and show some clinical and treatment-response differences.

EPIDEMIOLOGY

The idiopathic inflammatory myopathies are rare sporadic disorders with an overall annual incidence of approximately 1 in 100,000 (**Table 1**). Except for juvenile dermatomyositis (JDM), the IIM are diseases of the adult, and besides IBM these affect more

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Acronym	
APCs	Antigen-Presenting Cells
AZA	Azathioprine
BAFF	B Cell-Activating Factor
CK	Creatine Kinase
CMAP	Compound Muscle Action Potential
CS	Corticosteroids
DC	Dendritic Cells
DM	Dermatomyositis
EMG	Electromyography
IBM	Inclusion Body Myositis
IFIH1	Interferon-Induced Helicase
ILD	Interstitial Lung Disease
IVIG	Intravenous Immunoglobulin
JDM	Juvenile Dermatomyositis
MSAs	Myositis-Specific Antibodies
MTX	Methotrexate
MUAPs	Motor Unit Action Potentials
NM	Necrotizing Myopathy
PCP	Pneumocystis Carinii Pneumonia
PDC	Plasmacytoid Dendritic Cells
PM	Polymyositis
RA	Rheumatoid Arthritis
SANAM	Statin-Associated Autoimmune
SRP	Signal Recognition Particle
TGF	Transforming Growth Factor

women than men. In a Dutch study that excluded IBM, necrotizing myopathy (NM) represented 19%, whereas dermatomyositis (DM) and nonspecific myositis accounted for 36% and 39% of all IIM, respectively.[1] Unlike findings from other studies, polymyositis (PM) was reported to be uncommon, accounting for only 2% of IIM cases.[1] However, a PM clinical phenotype was the most common cause of PM disorder in the Mayo Clinic case series.[2] Indeed, 27 of 43 cases with pathologic PM had clinical features of PM, whereas 37% had phenotypic IBM with tissue inflammation but no rimmed vacuoles. Studies of the combined incidence of PM and DM from Israel, South Australia, and the United States (Allegheny County, Pennsylvania and Olmstead County, Minnesota) have yielded rates ranging from 2.2 to 7.0 per million population using a variety of methods.[3] The incidence of DM in South Australia is to 1.0 to 1.4 per million, whereas in Olmstead County it is 9.6 per million inhabitants. The incidence of PM in South Australia derived from muscle biopsy findings and review of medical records is 4 times higher than that of DM, respectively 4.1 to 6.6 per million versus 1.0 to 1.4 per million. A recent study indicates the prevalence rates in South Australia to be 1.97 and 7.2 per 100,000 for DM and PM, respectively. In a nationwide Taiwanese population survey between 2003 and 2007, the overall annual incidences of DM and PM were 7.1 (95% confidence interval [CI] 6.6–7.6) and 4.4 (95% CI 4.0–4.8) cases per million population. The incidence of DM and PM increased with advancing age and reached a peak at age 50 to 59 years.[4]