Gastrointestinal and Hepatic Disease in the Inflammatory Myopathies

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KEYWORDS

• Corticosteroids • Creatine kinase • Dermatomyositis • Inclusion body myositis • Oropharyngeal dysphagia • Polymyositis

KEY POINTS

- Oropharyngeal dysphagia due to the involvement of pharyngeal and proximal esophageal musculature is the most common gastrointestinal symptom in myositis, and severe cases are generally associated with inclusion body myositis.
- Inflammatory myopathy should be considered in the differential diagnosis of unexplained dysphagia because it may occur without other muscular or skin manifestations.
- Because of increased risk of occult malignancies, especially in the first year, age-appropriate cancer screening such as colonoscopy is recommended upon the diagnosis of inflammatory myopathy.
- Recurrent or worsening myopathy or skin symptoms in dermatomyositis patients warrants reevaluation for occult malignancy.
- Elevation of aminotransferases in inflammatory myopathy patients can be of skeletal muscle origin rather than from the liver.

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INTRODUCTION

The idiopathic inflammatory myopathies are a group of acquired heterogeneous autoimmune disorders characterized by the hallmark feature of muscle weakness. Proximal muscle weakness, when present, is often disabling. Because of insidious nature of the disease and nonspecific symptoms such as malaise and fatigue, diagnosis can frequently be delayed. The diagnosis is particularly challenging when the nonmuscular organ involvement such as gastrointestinal, pulmonary, or cardiac manifestations precedes muscle weakness. The extent and severity of associated systemic organ involvement varies with each type of inflammatory myopathy. Dermatomyositis (DM), due to the characteristic skin rash, may be better recognized than other inflammatory myopathies. Because of such challenges and the heterogeneity of inflammatory myopathies, understanding the pathogenesis and nonmuscular systemic organ involvement is critical for the diagnosis and management of this unique group of patients. The diagnosis of inflammatory myopathies is based on elevated serum muscle enzymes, electromyographic abnormalities, and presence of inflammatory infiltrate in the skeletal muscle resulting in symmetric proximal muscle weakness.[1]

The inflammatory myopathies are classified into 3 major categories: polymyositis (PM), DM, and inclusion body myositis (IBM). However, 2 other subtypes, necrotizing autoimmune myositis and overlap syndrome have been increasingly recognized as distinct entities.[2] This classification is largely based on clinical, immunopathologic, and dermatologic characteristics.[3]

Epidemiology

The prevalence of inflammatory myopathies is not very precisely known and estimated largely based on epidemiologic studies that do not perform systematic sampling of the general population. Nevertheless, the evidence suggests that PM and DM are very rare diseases with estimated incidence of 1 to 10 new cases per million persons per year.[4–8] In the United States, based on the medical claims data from a large managed care database, the adjusted annual incidence rate of inflammatory myopathies is 5.8 to 7.9 per 100,000 person-years and annual prevalence is 14.0 to 17.4 per 100,000, which is higher than elsewhere.[9] Women are affected at a higher rate than men, generally at 2:1 ratio, and as high as 5:1 during childbearing years.[4,8] The adult inflammatory myopathies usually occur in middle age with an average age of onset for DM and PM ranging from 52 to 56 years. In contrast, IBM occurs in later years with mean age of onset of 67 years, and it is more common in men.[8,10] On the other hand, juvenile DM is the most common (85% cases) form of juvenile inflammatory myopathies with a median age of onset of 7.5 years.[11]

The systemic organ involvement associated with inflammatory myopathies includes arthritis, cardiac arrhythmias, pulmonary and gastrointestinal symptoms. A myriad of gastrointestinal disorders involving oropharynx, esophagus, stomach, liver, small intestine, colon, and rectum has been reported in inflammatory myopathy patients (**Table 1**). In addition, there is a substantial increase in risk of occult malignancies, including the gastrointestinal cancers in myopathy patients, especially in those with DM. In this review, the authors focus on the gastrointestinal and hepatic manifestations associated with inflammatory myopathies.