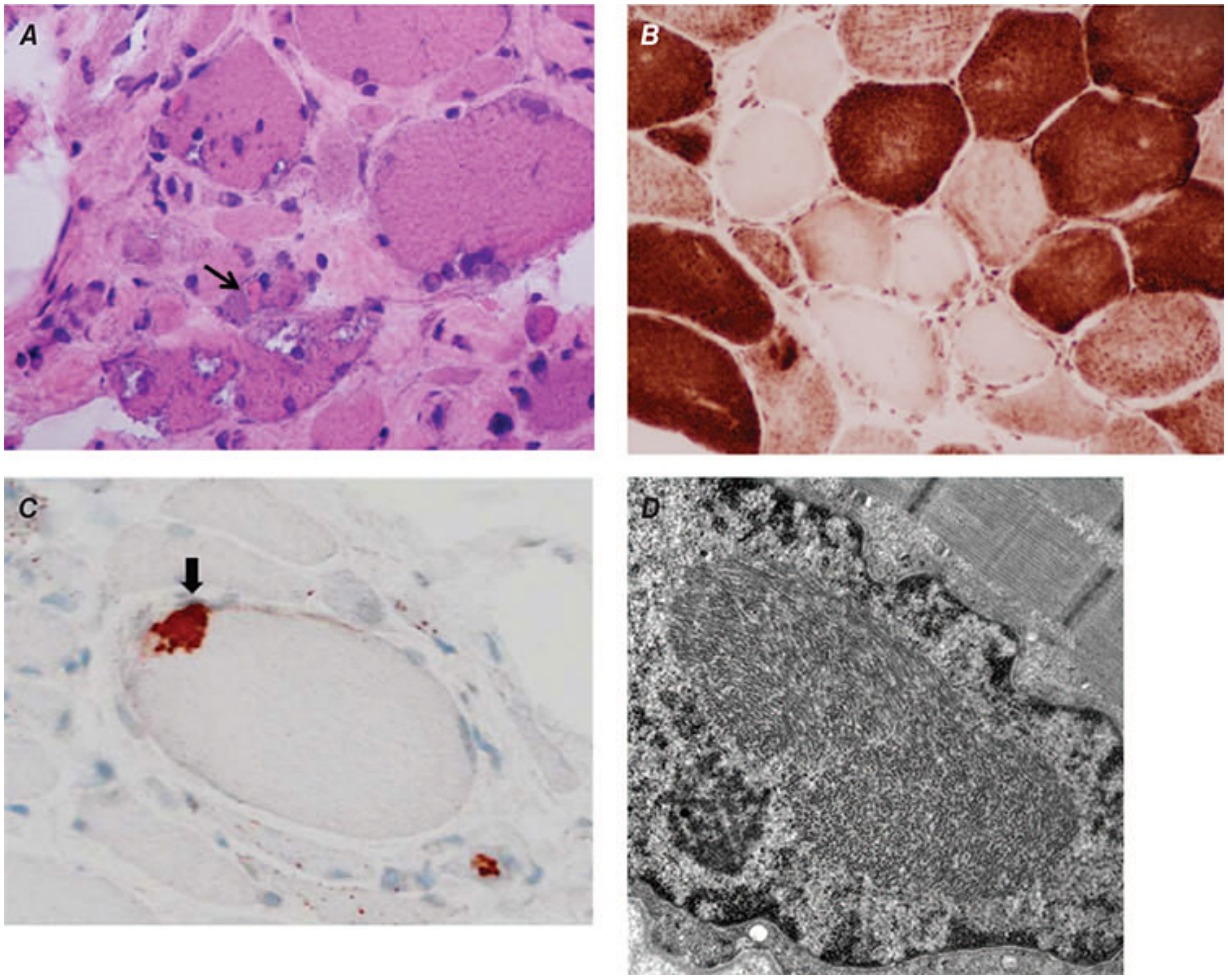

FIGURE 365-8 Pathology of inclusion body myositis. ***A.*** Scattered muscle fibers with rimmed vacuoles and rare fibers with eosinophilic inclusions (*arrow*), hematoxylin and eosin stain. ***B.*** Cytochrome oxidase stain demonstrates an increased number of pale-staining or COX-negative muscle fibers. ***C.*** Cytoplasmic inclusions stain positive with p62 within a muscle fiber (*thick arrow*). ***D.*** Electromicroscopy reveals 15- to 21-nm tubulofilamentous inclusions within a myonucleus.

The pathogenesis of IBM is poorly understood. The marked adaptive immune system abnormalities related to T-cell inflammation and the presence of a relatively specific autoantibody against a muscle protein indicate an autoimmune attack on muscle. The chronic and highly inflammatory environment within muscles in IBM may alter protein synthesis and degradation pathways in part via aberrant immunoproteasome expression. Additional histologic features, typically referred to as “degenerative,” include aggregation of various proteins including markers of endoplasmic reticulum (ER)