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Original Contribution

Inclusion body myositis and anesthesia: a case series☆,☆☆

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Abstract

Study Objective: Inclusion body myositis is a painless inflammatory myopathy affecting older adults. It manifests as progressive muscle atrophy and weakness, typically affecting proximal lower extremity muscles initially but insidiously progressing to affect other muscles, including bulbar (oropharyngeal) muscles and the diaphragm, and leading to dysphagia and respiratory insufficiency. This study reviews the perioperative outcomes of patients with inclusion body myositis who received general anesthesia.

Design: Observational retrospective study.

Setting: Academic tertiary referral center.

Patients: Patients with inclusion body myositis from October 1, 2009, to September 30, 2015, undergoing procedures requiring general anesthesia.

Interventions: Perioperative health records were reviewed.

Measurements: Perioperative outcomes and complications were assessed, with emphasis on respiratory complications and unexpected reactions to succinylcholine and nondepolarizing neuromuscular blocking drugs.

Main Results: Sixteen patients with inclusion body myositis underwent 18 procedures requiring general anesthesia. Succinylcholine was used during induction in 6 cases (33.3%) and nondepolarizing neuromuscular blocking drugs in 11 cases (61.1%). For 13 patients (72.2%), the trachea was extubated at the end of surgery, and none had postoperative respiratory complications. The 5 patients who continued to have tracheal intubation were expected to require continuous mechanical ventilation postoperatively. Three patients died within 30 days of surgery. One patient underwent a tracheostomy for planned long-term mechanical ventilation but life support was withdrawn after 22 days, and 2 severely deconditioned patients died at 11 and 15 days following general anesthesia for endoscopic procedures.

Abbreviations: IBM, Inclusion body myositis; NMBD, Neuromuscular blocking drug.

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Conclusions: Our patients with inclusion body myositis had uneventful perioperative outcomes following general anesthesia with depolarizing and nondepolarizing muscle relaxants. The small patient cohort in our series precludes a definitive conclusion regarding the safety of anesthetic agents in this patient population.

1. Introduction

Inclusion body myositis (IBM) is a rare, idiopathic, inflammatory myopathy [1] with clinically distinct histologic features that include rimmed vacuoles with granular material in muscle, atrophic fibers, and eosinophilic cytoplasmic inclusions (Fig. 1). IBM is unresponsive to corticosteroids, which differentiates it from the more common immune-inflammatory myopathies, polymyositis and dermatomyositis [2]. IBM is characterized by progressive, painless myositis with muscle atrophy and weakness [3,4]. It preferentially affects men (male to female ratio, 3:1) with typical onset in middle to late life [5]. Initial presentation typically manifests as proximal muscle weakness, more commonly in the lower extremities [3,4,6]. However, the disease progresses over months to years to affect proximal, as well as distal, and upper and lower extremity muscle groups [3,4,6]. Over time, bulbar (oropharyngeal) muscles are affected in approximately one-half of patients, manifesting as dysphagia and creating a risk of aspiration [7]. Although primary respiratory failure secondary to diaphragmatic weakness has been reported in only 2 case reports [8,9], IBM-associated respiratory failure may be misdiagnosed and may be more common than appreciated [10].

Diagnosis of IBM is based on clinical suspicion and confirmed with muscle biopsy (Fig. 1) and electromyography (Fig. 2) [2]. Although no consistent laboratory findings are characteristic for IBM, the creatine kinase level may be slightly elevated [3,4]. Despite initial promise that immunosuppressive therapy could slow IBM progression [3,4,11–13], larger prospective studies have demonstrated that such treatments, including immunoglobulin, have limited benefit [11,14] and may actually speed the progression of disability [2]. Because of the discouraging results of these studies, in addition to the risks involved with immunosuppressive treatments, therapy is generally supportive and includes physical rehabilitation [7].

Characteristics of IBM have raised concerns regarding the safety of anesthetic management. In general, patients with inflammatory myopathies may be hypersensitive to anesthetic agents, analgesics, and volatile agents. In addition, they may have an atypical response to muscle relaxants. Specifically, patients with decreased muscular physiologic reserve who are exposed to anesthetics and muscle relaxants may have respiratory failure, and poor coordination of oropharyngeal muscles may result in aspiration. However, the literature of anesthetic management for these patients is sparse because IBM is rare.

The purpose of the present study was to review the clinical course of patients with IBM who had anesthetic management at a large tertiary referral center and to assess outcomes and perioperative complications. To further expand the knowledge of this rare disorder, we performed a systematic review of the literature.

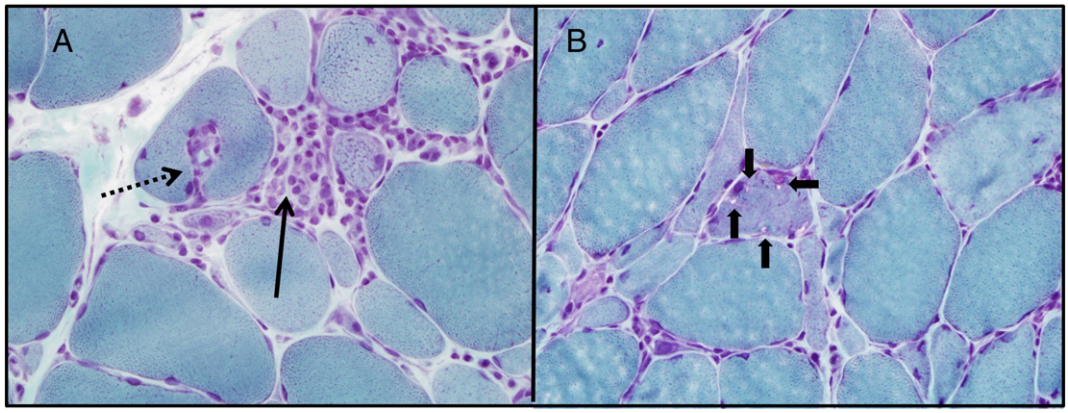

Fig. 1 Characteristic Histopathologic Findings in Inclusion Body Myositis. A, Inflammatory myopathy with mononuclear inflammatory infiltrates (CD8+ T cells and macrophage predominant) (arrow) that also invade a nonnecrotic myofiber (dashed arrow). B, Intracytoplasmic vacuoles (arrows) (Mallory trichrome stain, original magnification ×360). In addition, on Congo-philic staining, amyloid β plaquelike deposits are evident intracytoplasmically (not shown).