

Inclusion Body Myositis Associated with Progressive Dysphagia: Treatment with Cricopharyngeal Myotomy

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ABSTRACT: A 68-year-old man known to have inclusion body myositis underwent a cricopharyngeal myotomy in an attempt to improve his progressive dysphagia. Morphological studies from tissues obtained during this procedure showed the diagnostic features typical of this chronic inflammatory myopathy. To our knowledge this is the first pathological demonstration of inclusion body myositis involving the pharyngeal skeletal musculature.

RÉSUMÉ: Myosite à corps d'inclusion associée à une dysphagie progressive: traitement par myotomie cricopharyngienne Un homme de 68 ans connu comme étant porteur d'une myosite à corps d'inclusion a subi une myotomie cricopharyngienne dans le but d'améliorer sa dysphagie progressive. Les études morphologiques faites sur les tissus obtenus lors de l'intervention ont mis en évidence des éléments caractéristiques de cette myopathie inflammatoire chronique. À notre connaissance, c'est la première démonstration anatomo-pathologique d'une atteinte de la musculature du pharynx par une myosite à corps d'inclusion.

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Inclusion body myositis (IBM) is an adult onset, chronic and painless inflammatory myopathy, usually resistant to immunosuppression.^{1,2} In addition to the well-described involvement of limb musculature, a few cases with swallowing difficulties have been reported,^{1,3} but none had pathological studies to pharyngeal muscles. We now present a patient with severe progressive limb weakness and dysphagia in which pharyngeal muscles showed the typical light and electron microscopic features of IBM.

CASE REPORT

This man was known to have chronic progressive diffuse limb weakness of 10 years' duration, and at age 62 was diagnosed to have typical clinical, electromyographic and morphological features of IBM.² In the past two years, he also developed severe progressive dysphagia. He had difficulty tolerating liquids and started experiencing episodes of nasal regurgitation. An oropharyngeal swallow study showed diminished peristalsis in the pharynx and upper esophagus suggestive of a motility disorder. In an effort to improve his dysphagia, a cricopharyngeal myotomy was performed. Biopsies from omohyoid and cricopharyngeal muscles were processed for routine histochemistry and electron microscopy (EM).⁴

Post-operatively his dysphagia gradually, but significantly improved. Two months after the procedure, the patient was able to swallow his secretions and no longer had coughing spasms or aspiration

during sleep. A repeat oropharyngeal swallow study at that time showed some improvement in the patient's deglutition. Retention of masticated material still occurred in the pyriform sinus but to a lesser degree than pre-operatively. Ingestion of a paste material resulted in only mild pyriform hesitation, compared to the severe stasis seen in the earlier study. In the last year of his life he became totally flaccid and quadriplegic. His swallowing difficulties returned to his pre-operative state. He died at age 71 from respiratory complications. An autopsy was not performed.

RESULTS

Light microscopy: Cryostat sections from the cricopharyngeal muscle showed numerous small, rounded fibers, varying in size. There were many necrotic fibers undergoing phagocytosis and extensive mononuclear infiltrates, both perivascular and endomysial in distribution. Rimmed vacuoles were scant, but present. Some fibers showed an excess of internal nuclei (Figure 1). There was a marked increase in endomysial connective tissue and some replacement with fat. Many intramuscular nerve bundles were observed and appeared to be normal. With epoxy-resin histology, rimmed vacuoles were more abundant. They contained dark, rounded, granules and other debris.

Some fibers had extensive Z-disc streaming, while others contained dark gray areas compatible with an increase in mito-

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chondria, which were occasionally intermingled with lipofuscin granules.

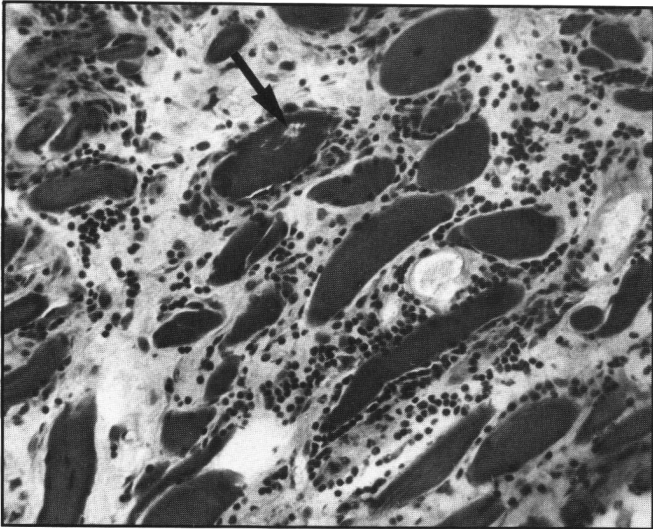


Figure 1 — Cryostat section from the cricopharyngeal muscle showing extensive mononuclear cell infiltration both endomyisial and perivascular in distribution. One fiber also shows a rimmed vacuole (arrow). H and E X60.

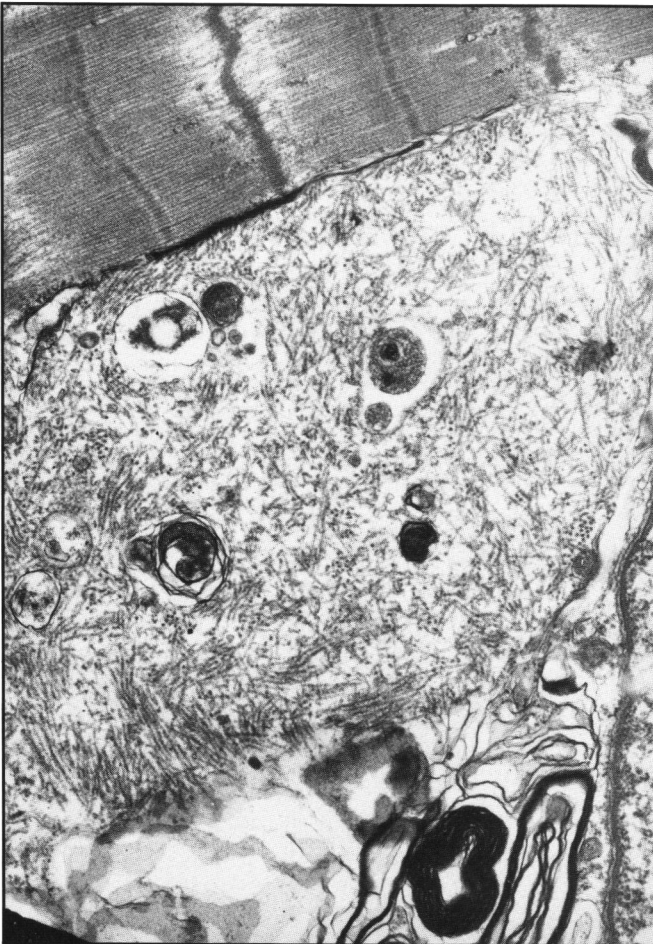


Figure 2 — This electron micrograph of cricopharyngeal muscle shows masses of filaments measuring 14 to 18 nm. in diameter. X 33,000.

Sections from the omohyoid muscle showed similar but less extensive abnormalities than those in the cricopharyngeal muscle. There was no increase in connective tissue and fat. Intranuclear inclusions were not seen in either muscle.

Electron Microscopy: Ultrastructural studies performed on many blocks from both muscles showed rimmed vacuoles containing masses of 14 to 18 nm filaments, cytomembranous whorls and some glycogen particles (Figure 2). The cricopharyngeal muscle also showed intranuclear filaments (Figure 3). Some muscle fibers displayed abnormal accumulations of mitochondria with unusual crystal arrangements and a few contained paracrystalline inclusions. Other scattered muscle fibers had extensive areas of Z-disc streaming and a few nemaline rods.

COMMENT

Although large series of patients with IBM have shown that prominent dysphagia and facial weakness is rare,¹⁻³ in a recent report of 19 patients, Ringel et al⁵ noted dysphagia in 6, and facial weakness in 10. A few patients with IBM have been noted to respond to immunosuppressive therapy but these were reported in abstracted form, and to our knowledge have never been fully described.^{5,6} Of our 17 patients including the present one, none showed improvement with various immunosuppressive

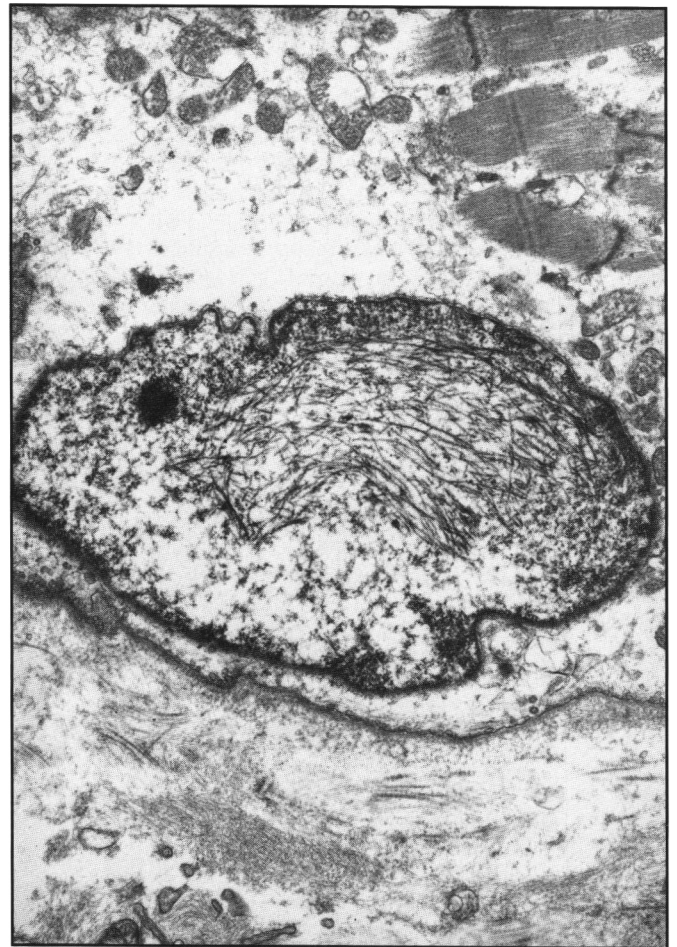


Figure 3 — This electron micrograph shows similar filaments in a nucleus. X 19,250.

strategies. Thus, in an attempt to improve this patient's severe dysphagia, a cricopharyngeal myotomy was performed. During the dissection, conspicuous hypertrophy of the cricopharyngeus muscle was observed. Post-operatively, the patient's swallowing gradually but significantly improved and remained fairly stable until it started to deteriorate again, one year prior to his death.

The normal mechanism for deglutition in the pharynx and upper esophagus involves a complex series of coordinated muscle movements associated with contraction of the pharyngeal musculature and cricopharyngeal relaxation. Dysphagia that is not secondary to a foreign body or neoplasm can occur because of improper contraction of the pharyngeal musculature or failure of the cricopharyngeus muscle to relax resulting in a functional obstruction; thus, myotomy of this muscle in selected patients may improve dysphagia.^{7,8} Cricopharyngeal myotomy is often performed for the surgical treatment of pharyngoesophageal (Zenker's) diverticulum,⁷ in rehabilitation of the functioning pharynx after head and neck surgery⁹ and for voice restoration after laryngectomy.¹⁰ This procedure has also been used in diseases of the central and the peripheral nervous systems, associated with incapacitating dysphagia, e.g. Parkinsonism, amyotrophic lateral sclerosis, bulbar poliomyelitis, oculopharyngeal muscular dystrophy and in idiopathic "cricopharyngeal dysphagia".¹¹⁻¹⁶ The procedure is usually performed through a Mosher type neck incision with division of the omohyoid muscle and lateral mobilization of the thyroid gland. The lower segment of the pharyngeal constrictor is divided from the posterior edge of the thyroid cartilage and the cricopharyngeal portion is divided posterior to the recurrent laryngeal nerve.^{17,18}

Cricopharyngeal myotomy has been shown to be a relatively safe and useful procedure that alleviates dysphagia as a result of improper relaxation of the cricopharyngeus muscle regardless of its etiology. It may also be helpful in patients with chronic neuromuscular diseases such as IBM for which there is no known medical treatment. Determination of the long term benefits of this procedure, however, still needs study of the results in a larger group of patients.

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