Cricopharyngeal Muscle Hypertrophy Associated with Florid Myositis

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Abstract. Hypertrophy of the cricopharyngeal muscle is a serious clinical condition that can cause severe dysphagic symptoms, including prolonged deglutition and postdeglutitive aspiration. Although the therapeutic concepts are well established, the pathogenic mechanism of cricopharyngeal hypertrophy remains unclear. We present a patient with a ten-year history of progressive dysphagia. The neurological and MRI findings were normal. However, videofiberendoscopy showed severe hypertrophy of the cricopharyngeal muscle. This condition was first treated by injections of botulinum toxin, which did not alleviate the symptoms. Next, myotomy and muscle biopsy were performed. Histological evaluation disclosed lymphoplasmacellular florid myositis, single-fiber atrophy, and muscle fiber necrosis with phagocytosis. There were no signs of inclusion body myositis or oculopharyngeal muscular dystrophy. Our finding of severe cricopharyngeal muscle hypertrophy associated with myositis has been published previously (n = 34). The study presented here shows cricopharyngeal dysphagia associated with various systemic diseases, including motor neuron disease, general granulomatous disease, dermatomyositis, or inclusion body myositis. Isolated changes of the cricopharyngeal muscle were described in 65% of the cases.

Key words: Hypertrophy — Myositis — Cricopharyngeal muscle — Deglutition — Deglutition disorders.

First described by Valsalva [1], the cricopharyngeal muscle plays an important role in deglutition. Dysfunction of the pharyngeal phase may be due to neurogenic oropharyngeal dysphagia or to oculopharyngeal muscular dystrophy. However, the mechanism by which long-term pharyngeal dysfunction leads to hypertrophy of the cricopharyngeal muscle has not been clarified. We present a patient with severe hypertrophy of the cricopharyngeal muscle associated with the histomorphological diagnosis of florid myositis.

Case Report

In June 1998, a 75-year-old woman presented with a ten-year history of dysphagia. She complained of occasional postprandial aspirations but no episodes of pneumonia. Thyroidectomy was performed in 1972 and radio-iodine therapy in 1994 for Basedow’s disease. The only medication prescribed was thyrone 100 μg/d. Laryngoscopy showed drastic pooling of saliva in both piriform recesses. Neurological examinations were normal. Intracranial white matter lesions resulting from microangiopathy were demonstrated by magnetic resonance tomography. Videofiberendoscopy confirmed severe stenosis of the upper esophageal sphincter caused by cricopharyngeal hypertrophy (Fig. 1). Laboratory tests on thyroid function, WBC, ESR, and CRP were all normal. Creatinphosphokinase was 32 U/l (normal range 10–70). Manometry revealed a hypertonic upper esophageal sphincter. The patient received botulinum toxin injections (Dysport®, Speywood Pharma, Berkshire, UK, 120 units) in the cricopharyngeal muscle in August 1998 as described elsewhere [2]. Following treatment, the maximal pressure of the cricopharyngeal muscle had decreased from 117 to 31 mmHg and the resting pressure had decreased from 68 to 17 mmHg, but the patient’s symptoms were undiminished. To induce cricopharyngeal muscle atrophy, the botulinum toxin was injected again in October 1998, without significant improvement of the patient’s dysphagic symptoms. In June 1999, eight months after the second botulinum toxin injection, a 6-cm myotomy of the inferior pharyngeal constrictor, the cricopharyngeal, and the upper esophageal muscle was performed. There was a small spondylodeviation
clification body myositis or oculopharyngeal muscular dystrophy. Ragged red fibers suggestive of mitochondriopathy were not observed.

Discussion

The basis for cricopharyngeal muscle hypertrophy is still unknown, but it seems likely that multiple factors contribute to the disease. The cricopharyngeal muscle is innervated by the IXth and Xth cranial nerves and the sympathetic cervical plexus, which participate in controlling deglutition and the respiration. The muscle contracts when the Valsava test is performed and prevents air from entering the stomach [3].

Samples from normal cricopharyngeal muscles obtained postmortem showed significantly more oxidative fibers and Type I fibers but fewer Type IIb fibers compared with biopsies from vastus lateralis muscles [4]. In addition, abundant endomysial and perimysial connective tissues were reported in normal cricopharyngeal muscle biopsies taken postmortem [5]. This was confirmed in samples from open surgery, where connective tissue and fatty cell infiltration was observed in a control group [6].

So far, 34 cases of patients with cricopharyngeal muscle hypertrophy have been reported. The literature on the histopathology of cricopharyngeal muscle biopsies suggests that cricopharyngeal muscle hypertrophy is either localized [6–10] or part of a systemic disease [3,5,11–18] (Tables 1 and 2). The systemic diseases reported so far include dermatomyositis [12], polymyositis [13,14], inclusion body myositis [15–18], generalized granulomatous disease [11], and neuromuscular degeneration [3]. Concomitant scleroderma was observed in one case, but there were no histological signs of scleroderma in the cricopharyngeal muscle biopsy [9]. Recently, it was shown that dysphagia occurred in 80% of patients with inclusion body myositis [19]. Histological samples of patients without systemic disease showed inflammatory cell reactions in most cases (n = 13/21) [6–10].

We found marked endomysial inflammation and features consistent with severe florid myositis. The neurological examination was normal and there were no signs of polymyositis or other systemic myopathies. The present case supports the association of florid localized myositis and cricopharyngeal muscle hypertrophy as one of the possible reasons of cricopharyngeal hypertrophy.

Fig. 1. Videocineradiography of the lower pharyngeal and upper esophageal phase (48 frames/s). The anterior wall of the trachea shows contrast medium due to postdeglutitive aspiration. R = retention in the piriform recess. Black arrow = very thin contrast medium line in the center of the hypertrophic cricopharyngeal muscle. * = small spondylyphyte, which was observed during open surgery also.