A Case of Idiopathic Fibrosis Involving Head and Neck Musculature

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Abstract

Objectives: Describe a case of idiopathic fibrosis of head and neck musculature, not characteristic of any identifiable neuromuscular or connective tissue disorder.

Study Design: Case report.

Methods: Chart review, PubMed literature review.

Results: A 24 year old Moroccan female presented with right pharyngeal and submental swelling, requiring awake fiberoptic intubation. She was treated with IV antibiotics and steroids with subsequent extubation. She returned 4 months later with progressive dysphagia, dyspnea and neck pain. Her workup showed no evidence of abscess or acute infection, but a CT demonstrated nonspecific thickening of hypopharyngeal soft tissues. She was taken to the operating room for direct laryngoscopy with biopsy. She required emergent tracheostomy due to the inability to access the airway due to severe oral and pharyngeal fibrosis. A biopsy from the tonsillar pillar demonstrated benign fibroconnective and lymphoid tissue. Lab and tissue testing for disease processes including lymphoma, polymyositis and CREST were negative. Additionally, four extremity electromyography was normal. MRI revealed enhancement of the hypopharyngeal soft tissues and paraspinal musculature consistent with inflammatory changes. Repeat biopsy of pharynx and strap musculature demonstrated extensive fibrosis and atrophy. She was started on mycophenolate mofetil 7 months after her initial presentation which has resulted in mild improvement of her symptoms.

Conclusions: We present the case of progressive idiopathic pharyngeal and neck fibrosis with a negative rheumatologic workup. She is currently being treated with mycophenolate mofetil, with some improvement in her symptoms, including improved ability to move neck and eat. She has been successfully decannulated. The pathogenesis of her disease remains unknown.

Introduction

We describe a case report of idiopathic fibrosis of head and neck musculature in a 24-year-old Moroccan female. Clinical findings have shown her case to be an entity that is distinct from known neuromuscular conditions.

Results

A 24 year old Moroccan female initially presented to the emergency department January 20, 2015 with right pharyngeal and submental swelling and pain. A computed tomography (CT) scan demonstrated right parapharyngeal swelling and flexible fiberoptic laryngoscopy revealed edema of the epiglottis and base of tongue. She was taken to the operating room for awake fiberoptic intubation. After treatment with intravenous antibiotics and steroids, she was able to undergo successful extubation four days later.

Outpatient workup included an esophagram which showed thickening of the prevertebral soft tissues at C2-C3 and intermittent aspiration. A modified barium swallow was ordered and showed intermittent silent aspiration with thin liquids. A CT scan was repeated on May 27, 2015 and showed little interval change of her right hypopharyngeal soft tissue thickening.

Results (continued)

She represented to the emergency department on May 28, 2015 with progressive dysphagia, dyspnea and neck pain. Her workup showed no evidence of abscess or acute infection, She was taken to the operating room June 4, 2015 for direct laryngoscopy with biopsy. She required emergent tracheostomy due to the inability to access the airway due to severe oral and pharyngeal fibrosis. A biopsy from the tonsillar pillar demonstrated benign fibroconnective and lymphoid tissue.

Referral to rheumatology was made. Lab and tissue testing for disease processes including lymphoma, polymyositis, lupus and CREST syndrome were negative. Extensive serologic testing for viral and bacterial illnesses including human immunodeficiency virus (HIV), Epstein Barr Virus (EBV) and syphilis was negative. Additionally, four extremity electromyography was normal. MRI was obtained and revealed enhancement of the hypopharyngeal soft tissues and paraspinal musculature consistent with inflammatory changes. She ultimately required gastrostomy tube placement June 16, 2015 due to failure to thrive and inadequate oral intake. She was taken back to the operating room June 18, 2015 for repeat biopsy of pharynx and strap musculature, Pathology again demonstrated extensive fibrosis and atrophy. She was started on mycophenolate mofetil roughly 7 months after her initial presentation which has resulted in mild improvement of her symptoms. She was decannulated on September 14, 2015.

Since abstract submission, the patient has additionally developed areas of fibrosis on her chest wall and upper abdominal wall. She underwent biopsy of a left sided chest wall lesion which demonstrated a reactive myofibroblastic proliferation with irregular involvement of surrounding skeletal muscle without atypia or concern for malignancy. No characteristics of established sclerosing or neoplastic disorders were noted.

Figure 1

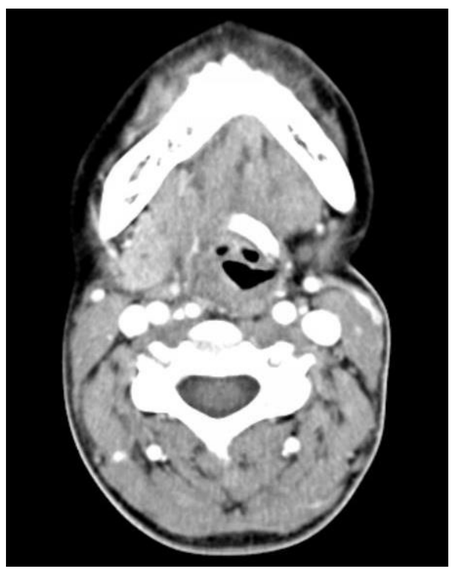

Figure 2

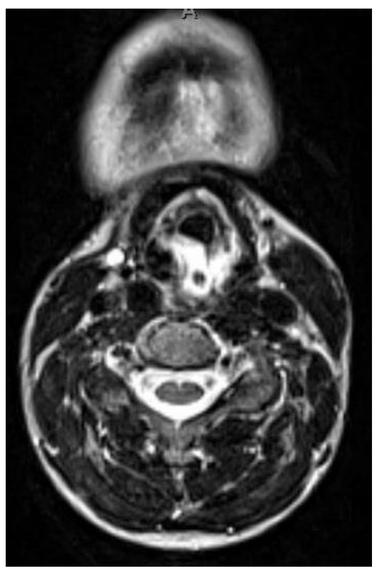

Figure 1: An axial CT image demonstrating thickening of the right hypopharyngeal soft tissues.

Figure 2: An axial MRI image demonstrating prominent T2 hyperintensity in this same region.

Discussion

We present a case of progressive idiopathic pharyngeal and neck fibrosis with a thus far negative rheumatologic, infections and neoplastic workup. A PubMed literature search shows there are no reported similar cases. Our patient experienced some symptomatic improvement with mycophenolate mofetil, including improved ability to move her neck and eat.