**Introduction**

First reported by Holmes and Panje as a mucosal variant of granuloma fasciale, eosinophilic angiocentric fibrosis (EAF) is a rare, idiopathic condition that predominately affects the sinonasal tract and orbit. The descriptive moniker is derived from its characteristic features: an eosinophilic-predominant infiltrate with angiocentric fibrosis. EAF has recently been classified as an IgG4-related disease (IgG4-RD). Although not malignant, these lesions have potential to become locally destructive.

Of the over 55 cases of EAF reported in the literature, only three have involved the larynx. Here we discuss the pertinent clinical, radiographic and pathologic findings of a case of subglottic EAF, demonstrating the diagnostic and management challenges.

**Case Report**

A 44-year-old female was referred for progressive dyspnea and stridor with minimal exertion. Inspiratory and expiratory limbs on flow volume loops were flattened, indicating airway stenosis. The vocal folds were mobile on flexible endoscopy, but the subglottic airway was significantly narrowed. Computed tomography (CT) demonstrated a homogenous, soft tissue lesion, with evidence of erosion of the posterior cricoid plate. (Fig 1) The subglottic narrowing of the larynx was not as significant as the CT imaging demonstrated.

Serology

Observation of elevated IgG4 serum levels in EAF contributed to its classification as an IgG4-RD, but is not required for diagnosis. Some 30% of patients will have a normal serum IgG4 despite pathologic diagnosis. Levels fluctuate with disease activity and are known to decrease with steroid therapy. Additionally, elevated IgG4 is a nonspecific finding and can be found in other conditions such as autoimmune diseases.

**Management**

Treatment of IgG4-RD is based largely on studies of lymphoplasmacytic sclerosis pancreatitis (LPSP), a histologic subtype of autoimmune pancreatitis (AIP). LPSP is responsive to systemic steroids, and so glucocorticoids have become the mainstay of therapy. Patients will often relapse with cessation of treatment, and glucocorticoids appear to be most effective when initiated early in the disease. The effectiveness of steroids declines in later stages when fibrosis is established. The untoward side-effects of prolonged glucocorticoid use has spurred an interest in steroid-sparing therapy. Rituximab has shown promise as a means for reducing disease progression by reducing the number of B-cells available to become IgG4 plasma cells.

Unlike IgG4-RD, treatment of EAF has proven to be largely refractory to medical therapy. Steroids (intravenous or systemic) may slow disease progression, but have been unable to affect a lasting response. There are case reports describing the use of dapsone, azathioprine, hydroxychloroquine and tofacitinib, but results are equivocal and long-term data is lacking. Most authors agree surgical excision is the preferred treatment, in spite the fact that 70% of patients have persistent or recurrent disease on follow-up.

The favored sub-site for laryngeal EAF is the subglottis, and the lesion must be differentiated from any number of potential mimics in this location. The challenge of treating subglottic EAF is in the management of the airway.

**Conclusion**

EAF is an idiopathic, progressive disorder that belongs to the spectrum of IgG4-related diseases. Most commonly reported in the orbit or sinonasal tract, EAF lesions of the larynx are exceedingly rare and typically present as subglottic stenosis. Management of EAF is largely surgical. Persistent or residual disease is common, creating unique challenges in the management of subglottic disease. In the case of our patient, the integrity of the posterior cricoid plate has been compromised, making cricotracheal resection a less than ideal option. While she may eventually require tracheotomy or even laryngectomy, she is currently being managed with serial excision and dilation. She is being evaluated for adjuvant therapy medical therapy, highlighting the importance of a multidisciplinary approach to this disease.