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# Inflammatory Myofibroblastic Tumor in the Larynx

## Larinkste İnflamatuar Myofibroblastik Tümör

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**ABSTRACT**

Inflammatory myofibroblastic tumor is a benign pseudoneoplastic proliferation, that usually seen in the lungs. In the head and neck region paranasal sinuses and orbit are commonly involved, but laryngeal involvement is extremely rare. We present an inflammatory myofibroblastic tumor originating from the true vocal cord of the larynx and we discuss the clinical and microscopic features and the treatment modalities of this rare entity.

***Keywords***
*Laryngeal diseases; laryngeal neoplasms; granuloma, laryngeal; granuloma, plasma cell*

**ÖZET**

İnflamatuar myofibroblastik tümör, en sık yerleşim yeri akciğerler olan benign bir pseudoneoplastik proliferasyondur. Baş-boyun bölgesinde paranazal sinüsler ve orbita daha sık tutulurken larinks yerleşimi oldukça nadirdir. Larinkste vokal kordlardan köken alan bir inflamatuar myofibroblastik tümör vakası sunularak, nadir görülen bu hastalığın klinik ve mikroskopik özellikleri ve tedavi seçenekleri tartışılmıştır.

***Anahtar Sözcükler***
*Laringeal hastalıklar; laringeal neoplaziler; laringeal granüloma; plazma hücreli granüloma*

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## INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a newly described entity. This term includes a variety of synonyms: inflammatory pseudotumor, plasma cell granuloma, plasma cell pseudotumor, and pseudosarcomatous lesion/tumor.[1] It is usually seen in the lungs. In the head and neck region, paranasal sinuses and orbit are commonly involved, but laryngeal involvement is extremely rare.[2] It should be kept in mind that the aim of the therapy must be the organ preservation and to avoid unnecessary agressive treatment.

## CASE REPORT

A 63-year-old male patient presented with hoarseness lasting for 10 months. An accompanying progressive dyspnea was also reported. He had been smoking 20 cigarettes per day for 40 years. He had no history of a previous laryngeal disease or trauma. Otorhinolaryngologic examination revealed a polypoid mass originating from the anterior 1/3 of the right true vocal cord with intact cord mobility. Palpation of the neck revealed no neck mass or lymphadenopathies. CT scan showed an expansive mass with contrast enhancement narrowing the glottic rim (Figure 1). The mass was completely resected via direct laryngoscopy. Histopathologic evaluation of the specimen revealed a 1.8x1.5x1.3 cm solid lesion with superficial ulceration of the mucosa. The mass was mainly composed of proliferating spindle cells (myofibroblasts) in a hyalinized stroma consisting of plasma cell infiltration especially in perivascular regions (Figure 2). Immunohistochemical staining with desmin, CD34, ALK were negative, while mesenchimal cells were positively stained with actin. The diagnosis was reported as IMT. The patient was free of the disease after a two years of follow-up period.

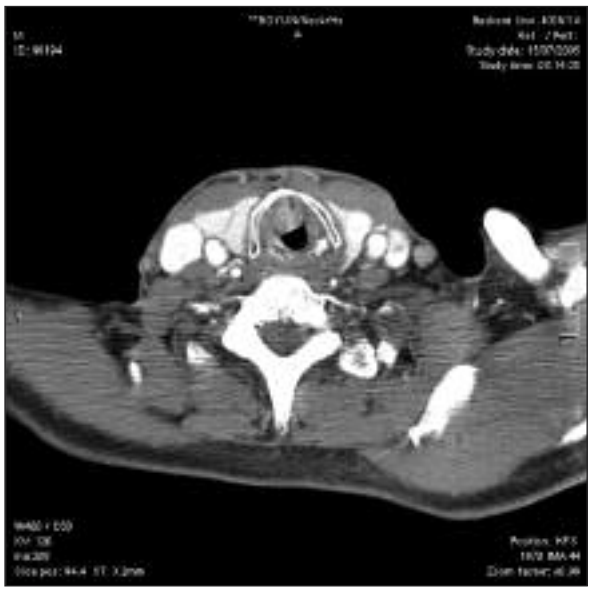

**Figure 1.** CT scan showing an expansive mass with contrast enhancement narrowing the glottic rim.

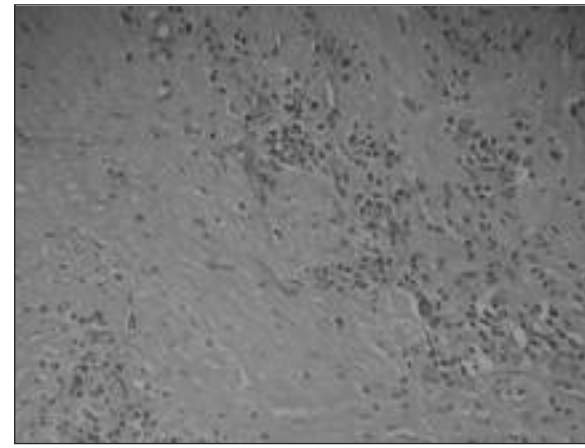

**Figure 2.** Proliferating spindle cells (myofibroblasts) with a mostly hyalinized stroma consisting of plasm cell infiltration especially in perivascular regions (H&E, x40).

## DISCUSSION

IMT is a recently described lesion with the lungs being the site of predilection.[3] This entity was previously categorised in plasma cell granulomas until Wenig et al.[4] reported eight cases including the same histopathologic features and designated them as IMT. Idrees et al[5] reported that the main histologic features of IMT were a chronic inflammatory background containing plenty of fibroblasts with myofibroblastic proliferation. With the plasma cells being the predominant ones, inflammatory infiltrate also consists of lymphocytes, eosinophils, and histiocytes. The degree of pleomorphism is known to be within the benign range, and the mitotic rate is usually less than two mitoses per 10 high-power field.[5] Immunohistochemical investigations could reveal an expression of vimentin in 99%, actin in 92%; as in our case, focally desmin in 69%, cytokeratins in 36%, CD68 in 24%, and CD30 in 6%.[6] Anaplastic lymphoma

kinase (ALK) gene expression is typical, but only one half of IMT cases including children and young adults under 40 years of age are positive.[7] Being 63-year-old, no ALK staining was observed in our case.

True vocal cords are usually the site of origin in laryngeal involvement.[4] However, subglottic, ventricular, and pyriform sinus involvement are also reported.[4,8] The etiology of IMT is still not fully understood. Laryngeal trauma seems to be the most acceptable pathophysiological mechanism.[9] Traumatic intubation, smoking, and gastric acid reflux might be initiating factor for posttraumatic exaggerated inflammation leading to IMT. Alaani et al.[9] reported a case of IMT localised in the subglottic larynx as a result of airbag injury. The lesion was found to progress to myositis ossificans in the histopathological sections of revision surgery.

Differential diagnosis includes benign and malignant spindle-cell neoplasms. In this wide range of diseases, low-grade myofibroblastic sarcoma must be specially taken into consideration as this tumor is often located in the head and neck region.[2] Differing from the IMT, this entity is locally infiltrative with the probability of distant metastasis.

Surgery is the first choice in the treatment. However, the lesion is usually unencapsulated, and this causes difficulties in estimating the extent of excision during the operation. It should be kept in mind that the lesion is benign and the primary aim must be organ preservation.[5,9,10] Laser excision[4] radiotherapy,[11] and steroid therapy[12,13] are reported as other management modalities. Wenig et al.[4] reported eight cases, six of whom treated with laser excision only and reported with no evidence of disease after a 12-24 months of follow-up period. Seider et al.[11] reported use of irradiation in a case of IMT located in the nasal cavity which was initially resected but recurred within one month. After radiotherapy, local control was achieved in a 27-month of follow-up period. However, complete resection is primarily recommended whenever possible and irradiation should be kept in mind in recurrent or inoperable local disease. Little is known about the efficacy of steroid therapy but Suh et al[13] reported an IMT of larynx, treated with systemic steroids after the laryngoscopic biopsy with no recurrence after a four years of follow-up.

The prognosis of laryngeal IMT is excellent. The reported recurrence rate was 21% and most of them occured within 12 months after initial surgery. This may be due to the lack of a line of demarcation surrounding the lesion that cause incomplete resection leading to recurrences.[6,14] Since the lesion is benign, the preferred treatment should be complete resection with organ preservation and it is critical to diagnose this rare entity to avoid unnecessary agresive treatment.

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