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Rosai-Dorfman Disease in the Larynx: Report of a Case and Literature Review (Postprint)

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Abstract

Rosai-Dorfman disease is a benign histiocytic proliferative lesion occurring in lymph nodes or extranodal tissues. Extranodal Rosai-Dorfman disease involving the larynx is extremely rare, with inconspicuous lesion features and lacking typical symptoms of fever and lymphadenopathy, making it prone to clinical misdiagnosis as a malignant tumor. Additionally, there are no standardized treatment protocols. This article reports one case of laryngeal Rosai-Dorfman disease treated with minimally invasive surgery combined with corticosteroid therapy, which preserved laryngeal function and achieved favorable outcomes, and discusses the diagnosis and treatment of this rare disease in conjunction with a literature review.

Full Text

Preamble

Case Report: Rosai-Dorfman Disease in the Larynx: Report of a Case and Literature Review

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Abstract

Rosai-Dorfman disease (RDD) is a rare histiocytic disorder characterized by nodal and extranodal manifestations. Currently, no standardized guidelines are available for the management of this disease. We report a case of laryngeal RDD that presented without classical symptoms such as fever or lymphadenopathy and could be easily misdiagnosed as a malignant tumor. The patient underwent

minimally invasive surgeries combined with steroid therapy to preserve laryngeal function as much as possible, achieving a favorable clinical outcome. We reviewed similar cases reported in the literature to better understand the disease course, diagnosis, and treatment of this uncommon condition.

Keywords: Rosai-Dorfman disease; extranodal involvement; surgical treatment

Introduction

In the 1960s, a distinctive lymphadenopathy characterized by fever, polyclonal gammopathy, and neutrophilia was first described. Occurring predominantly in childhood and early adulthood, this disease was defined by Rosai and Dorfman in 1969 as sinus histiocytosis with massive lymphadenopathy (SHML) [1]. In subsequent years, scattered cases reported extranodal involvement by SHML, including the respiratory tract, visceral organs, skin, bone, central nervous system, and genitourinary system. These entities were later collectively termed Rosai-Dorfman disease (RDD) [2]. RDD has a predilection for lymph nodes and usually presents as painless cervical lymphadenopathy [3]. In 1988, Suster described an unusual case of RDD involving only soft tissue without any lymphadenopathy [4], and later reports documented RDD presenting with isolated extranodal involvement [5], which is now estimated to occur in more than half of cases [6].

According to the scope of involvement, RDD can be classified into three types: the lymph node type with only lymph node involvement (often cervical lymph nodes with possible involvement of other regions); the extranodal type involving multiple extranodal organs (frequently the head and neck with possible involvement of limbs and other trunk areas); and the mixed type with involvement of both lymph nodes and extranodal organs [3]. Although massive cervical lymphadenopathy is the hallmark of RDD, the head and neck region is the most common site for isolated extranodal involvement. However, laryngeal involvement remains extremely rare. Of 12 cases retrieved from published literature, only one Chinese case was reported (Table 1). Most patients were middle-aged and lacked characteristic RDD features such as painless cervical lymphadenopathy, fever, or elevated white cell count. In contrast, laryngeal neoplasm presentation with symptoms such as hoarseness, dyspnea, and dysphagia was very common. Clinical diagnosis of RDD is difficult, and definitive diagnosis requires histopathological and immunohistochemical evidence. Currently, no well-defined treatment guidelines are available. Herein, we report a rare case of RDD with isolated laryngeal involvement that required differentiation from malignant laryngeal tumor, and we review similar documented cases to summarize our experience with diagnosis and treatment.

Case Report

A 57-year-old man presented to our hospital with progressive hoarseness for 2 months, mild dyspnea after activities, and an anterior neck mass for 1 month. Physical examination revealed a hard, nontender mass measuring 3 cm × 4 cm on the left side of the thyroid cartilage. The mass was fixed to the thyroid cartilage without skin adhesion. Under laryngoscopy, a cauliflower-like neoplasm was observed on the left side of the glottic area, subglottic area, and piriform fossa, with limited motion of the left vocal fold (Fig. 1A, B). Computed tomography (CT) scan of the neck showed a 3 cm × 4 cm × 5 cm mass arising from the left false vocal fold, extending to the subglottis and left piriform fossa, causing narrowing of the glottis. The mass breached the thyroid cartilage and extended to the cervical strap muscles (Fig. 1C, D). Based on these findings, diagnostic hypotheses of laryngeal tuberculosis or squamous cell carcinoma were considered. Subsequent excisional biopsies under laryngoscopy were performed twice, revealing inflammatory granulation and exudative necrosis without detection of cancer cells.

After multidisciplinary discussion, excision of the tumor was performed under suspension laryngoscopy following tracheotomy. Intraoperative frozen section examination reported an inflammatory polyp. Additional external biopsy of the neck mass was performed, and immunohistochemical analysis of surgical specimens showed a fibroadipose tissue mass with histiocytes exhibiting extensive phagocytosis of lymphocytes, plasma cells, and occasional neutrophils, supporting a diagnosis of laryngeal RDD. In areas of lymphophagocytosis (emperipolesis), immunohistochemistry confirmed positivity for S-100 protein and CD68 and negativity for CD1a in RDD cells (Fig. 2).

Postoperatively, the patient received steroid therapy with prednisone at 50 mg/day for two weeks, then reduced to 20 mg/day for two months. Three months later, the patient reported obvious relief of dyspnea and hoarseness, though the neck mass remained unchanged. He agreed to undergo a second surgery for resection of the neck mass along with necrotic thyroid cartilage without opening the laryngeal cavity. Pathological examination again confirmed the diagnosis of laryngeal RDD. After the second operation, the patient received prednisone at 10 mg/day for two months, with dose gradually tapered until discontinuation over three weeks.

The patient showed satisfactory recovery of respiratory, swallowing, and phonation functions. Laryngoscopic and CT examinations revealed smooth and mobile vocal cords. The patient was closely followed for 2 years, during which no recurrence was detected (Fig. 3).

Discussion

Differential diagnosis between laryngeal RDD and malignant tumor is difficult based solely on symptoms and imaging findings, and definitive diagnosis requires pathological evidence. Laryngoscopic biopsy often fails to obtain a definitive di-

agnosis due to insufficient tissue specimen. In our case, diagnosis was established only after mass excision, as three laryngoscopic biopsies and an intraoperative frozen section all failed to confirm the diagnosis.

Histopathological features of extranodal RDD are roughly consistent with nodal RDD, with heavy infiltration by histiocytes, lymphocytes, plasma cells, and erythrocytes, along with lymphocyte endocytosis by histiocytes (emperipolesis). However, extranodal RDD demonstrates more obvious fibrosis with fewer histiocytes and less distinct emperipolesis. RDD histiocytes are immunohistochemically positive for S-100 protein and CD68 and negative for CD1a, which facilitates differential diagnosis from other diseases [3]. According to statistics, RDD is self-limited in more than 20% of cases and shows spontaneous regression without intervention, while 70% of patients have noticeable symptoms and vital organ involvement requiring treatment [6]. The most common treatment modalities include surgery, steroids, radiation, and chemotherapy. Surgical excision remains the most effective option for solitary lesions or a limited number of lesions. In cases of laryngeal RDD with obstructive or compressive symptoms, surgery should be the primary therapeutic approach. Glucocorticoids are the first-line option for systemic treatment, as most RDD patients, whether classical or extranodal, show favorable response to glucocorticoid therapy [11, 14]. Radiotherapy and chemotherapy are only considered for intravenous indications or cases with failed surgical treatment [12].

Among the 12 cases reported in literature, 6 received laryngomicrosurgery under suspension laryngoscope, 3 underwent laryngofissure, and the remaining cases received monotherapy with observation, chemotherapy, radiotherapy, or steroid therapy as a common treatment for RDD was given in 5 cases. After treatment, the condition was controlled in all 12 cases (Table 1). Therefore, we recommend that microinvasive operation or steroid therapy followed by surgery be the primary treatment choice once RDD diagnosis is established. Microinvasive surgery can quickly relieve symptoms such as hoarseness and dyspnea, while steroid therapy can control disease progression and prevent recurrence. In our case, we performed two microinvasive surgeries (intralaryngeal and extralaryngeal) with postoperative steroid therapy, preserving the laryngeal structure and mucosa well. The patient was satisfied with phonation during follow-up.

Conclusion

RDD with isolated laryngeal involvement is a rare, benign, self-limited histiocytic disorder that mimics laryngeal malignancy. Laryngoscopic biopsy often fails to obtain an accurate diagnosis. In such cases, microinvasive surgery combined with steroid therapy may produce good long-term outcomes. Clinicians should be aware of RDD with isolated laryngeal involvement when performing surgeries to avoid functional damage to the larynx.

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