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Subglottic Tumor Revealing Rosai-Dorfman Disease

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Abstract **Case Report**

Introduction: Rosai-Dorfman disease (RDD) is a rare atypical proliferation of histiocytes. RDD revealed by laryngeal involvement, as in the present case, is even less common. The objective of this presentation is to illustrate the clinical, para-clinical and therapeutic aspects of this rare localization through this clinical case and a review of the literature. ***Case report:*** A 40 years old woman is presented with a dysphonia of 4 year of evolution, aggravated by laryngeal dyspnea requiring emergency tracheotomy. Physical examination showed multiple cervical adenopathies, with a subglottic lesion covered with healthy mucosa. Consistent with the CT scan findings, the lesion caused a 90% obstruction. Histopathologic findings of the lesion biopsy revealed the diagnosis of RDD confirmed by immunohistochemical complement (positivity of PS100 and negativity of CD1a, and of CD207). The patient was put on chemotherapy based on vincristine. The evolution was marked by a clear regression of the subglottic obstacle; hence the decision to remove the tracheotomy cannula was taken. With a decline of 6 months, no recurrence was noted. ***Conclusion:*** The diagnosis of RDD should be kept in mind when evaluating sub-glottic tumor with cervical lymphadenopathy. A high degree of suspicion and a thorough pathological review are necessary to diagnose this rare clinical entity.

Keywords: Rosai-Dorfman disease, Laryngeal tumors, Histiocytosis.

INTRODUCTION

Rosai-Dorfman disease (RDD) or Sinus histiocytosis with massive lymphadenopathy was 1st described by Rosi and Dorfman in 1969 [1]. RDD is a rare, benign, idiopathic, histiocytic, proliferative, self-limiting disease that seriously affects healthy young adults and adolescents. This disease process is characterized by dilated lymph node sinuses that are filled with massive lymphocytes, plasma cells, and histiocytes that are typically accompanied by fever, polyclonal gammopathy, and leukocytosis with neutrophilia [2]. RDD with laryngeal involvement -as in the present case- is extremely rare, and less than 30 such cases have been reported in the English literature so far, with most being reports of isolated cases [3]. The diagnosis of RDD is made using histopathological and immunohistochemical methods [4]. The objective of this presentation is to illustrate the clinical, para-clinical and therapeutic aspects of this rare localization through this clinical case and a review of the literature.

CASE REPORT

A 40 years old woman, with no history of alcohol, smoking poisoning, tuberculosis or laryngeal trauma, is presented with a dysphonia of 4 years of evolution with no associated symptoms, evolving in a context of apyrexia and conservation of the general state. This symptomatology was aggravated with laryngeal dyspnea which was resistant to medical treatment (Nebulization, corticosteroids).

An emergency tracheotomy was performed due to the respiratory instability of the patient. Physical examination showed multiple bilateral insensitive cervical adenopathies which were mobile in relation to both planes, in the sub-mental and sub-mandibular sectors (Fig. 1). Nasofibroscopy revealed no abnormalities in the nasal cavity, cavum, and oropharynx, whereas the larynx could not be examined for salivary stasis due to the presence of the tracheostomy tube. The rest of the clinical examination was unremarkable.

Neck and chest computed tomography (CT) showed a subglottic tissue thickening, enhanced by intravenous contrast, with multiple adenopathies (Fig.2). We further performed a laryngeal endoscopy and observed a subglottic lesion covered with healthy mucosa (Fig.3). Consistent with the CT scan findings, the lesion caused a 90% obstruction. A biopsy for pathologic analysis was performed. Histopathologic findings revealed that the mucosa was lined by a regular squamous epithelioma; chorion was the bed of a very dense inflammatory infiltrate arranged in layers, made of small lymphocytes.These lymphocytes were mixed with many macrophages that phagocytes lymphocytes and which contained nuclear debris in their cytoplasm (Fig.4).This aspect was in favor of a haemophagocytic non-langerhansian histiocytosis compatible with Roasai Dorfman's disease. The immunohistochemical complement confirmed the diagnosis by highlighting a positivity of PS100 and negativity of CD1a, and of CD207. Based on these clinical, radiological, and histological criteria; the diagnosis of a laryngeal-sounding RosaiDorfman disease was confirmed.

Consequently, the patient was referred to the department of internal medicine, where she was initially put on corticosteroid (prednisolone 1 mg / kg / day) for 2 months. Due to the lack of improvement of the functional symptomatology and the persistence of the same finding in laryngoscopic control, chemotherapy was instituted based on vincristine. The evolution under chemotherapy was marked by a clear regression of the subglottic obstacle, which led to the decision to remove the tracheotomy cannula. No recurrence was noted with a decline of 6 months.

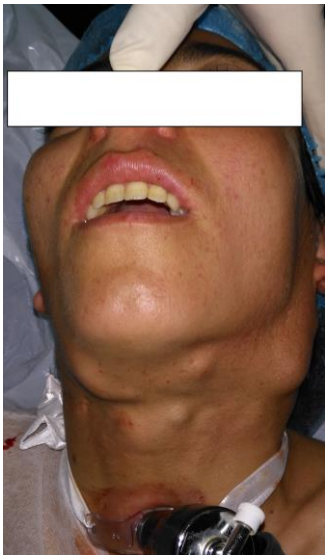

Fig-1: Photo showing submental and submandibular adenopathies

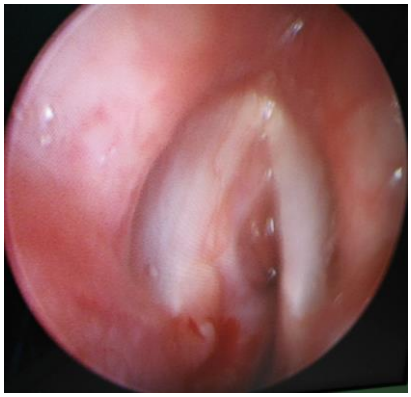

Fig-2: Endoscopic aspect showing an obstructing subglottic process

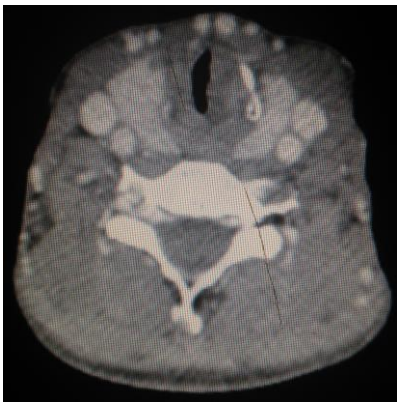

Fig-3: Axial CT section showing a tissular subglottic obstructing process

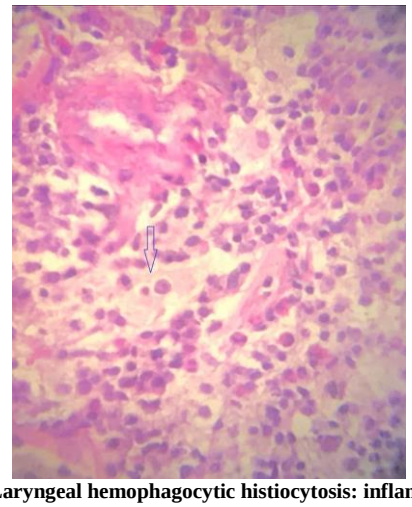

Fig-4: Laryngeal hemophagocytic histiocytosis: inflammatory infiltration of the chorion, rich in macrophages phagocytinglymphocytes (arrow) (HES x200)

DISCUSSION

RDD disease is a rare entity. Despite several articles printed, most of which are either case reports or review articles, the diagnosis and the treatment of the disease are still difficult [5]. The characteristic features of RDD are massive, painless cervical lymphadenopathy, fever, raised erythrocyte sedimentation rate, and raised white cell count. The disease has a male: female ratio of approximately 5:1and an extranodal involvement rate of below 50% or

approximately 25–43% [6]. The most common sites of extranodal involvement include the skin, soft tissue, central nervous system, and upper respiratory tract [3,6]. RDD with laryngeal involvement is extremely rare, and less than 30 such cases have been reported in the English literature so far, with most being reports of isolated cases [3]. Patients with masses of the upper respiratory tract may exhibit a variety of symptoms, such as foreign body sensation, dysphagia, dyspnea, voice changes, cough, and stridor [2]. Airway involvement may cause emergency respiratory distress as seen in the case presented here; and requires surgical intervention. The imaging features of extranodal RDD are non-specific and definitive diagnosis almost always requires pathologic confirmation [2]. Typical histologic features, such as diffuse lymphoplasmacytic infiltrate, Russell bodies, foamy histiocytes, and histiocytes with emperipolesis, confirm a diagnosis of RDD. The cytoplasm of histiocytes contains well-preserved lymphocytes; this is referred to as lymphagocytosis or emperipolesis. The histiocytes were immunohistochemically positive for S-100 protein (a phagocytic marker) and macrophage-specific antibodies, but markers for Langerhans (CD1a) or dendritic cells (DRC, CD23, and CNA42) were absent [7]. Extra nodal RD disease exhibits more fibrosis and lesser degree of Rosai—Dorfman cells [5]. Thus, the pathological diagnosis may be very difficult in case of extra nodal involvement as in the case presented in this article.

The cause of this disease is still unknown, although a viral etiology is suspected. Molecular studies have found no evidence of clonal rearrangement, implying that this disease is a reactive rather than neoplastic condition [7]. The differential diagnosis of RDD includes lymphoreticular malignancies, AIDS, and idiopathic thrombocytopenic purpura, which have similar histopathological features[4].

Due to its low incidence, no optimal or standard treatment has been defined [8].The disease is often self-limiting with a very good outcome; nevertheless, 5–11% of patients die from this disease. Multiple strategies of therapy that should be tailored to the individual lesion or patient include radiation therapy, chemotherapy, steroids, and surgery, which have been used with varying success [6]. Most patients (80%) had spontaneous resolution; extensive treatment is not required for most patients with RDD. A successful short course of high-dose steroid (20 mg dexamethasone daily for 3 days) was reported. Chemotherapeutic agents such as etoposide or 6-mercaptopurine with low-dose methotrexate have also been used.Treatment providers should be aware of a possible life-threatening condition caused by stricture of the subglottic space. Tracheostomy and CO2 laser operation are suggested if the airway is compromised [7].Therefore, it is recommended that patients with RDD undergo a careful laryngoscopy to exclude laryngeal involvement [3].

CONCLUSION

The diagnosis of RDD should be kept in mind when evaluating sub-glottic tumor with cervical lymphadenopathy. A high degree of suspicion and a thorough pathological review are necessary to diagnose this rare clinical entity. Accurate diagnosis is important to avoid unnecessary and often mutilating surgery.

Authors' disclosures of potential conflicts of interest

The authors indicated no potential conflicts of interest.

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