EJCRIM
European Journal
of Case Reports
in Internal Medicine

A RARE CASE OF SUBGLOTTIC ROSAI-DORFMAN-DESTOMBES DISEASE CAUSING FIXED UPPER-AIRWAY OBSTRUCTION IN AN ADULT WOMAN

Sheikh Abdullah[1], Camryn Herbert[1], Faryal Shoaib[2], Cole Bourgeois[1], Shiva Jashwanth Gaddam[3]

[1] Department of Internal Medicine, Louisiana State University Health Sciences Center, Shreveport, USA
[2] Department of Pathology, Louisiana State University Health Sciences Center, Shreveport, USA
[3] Department of Hematology and Oncology, Louisiana State University Health Sciences Center, Shreveport, USA

Corresponding author's e-mail: sheikh.abdullah02@lsuhs.edu

Received: 07/04/2026
Accepted: 27/04/2026
Published: 09/07/2026

Conflicts of Interests: The Authors declare that there are no competing interests.
Patient Consent: Written informed consent was obtained from the patient for publication of this case report.
Acknowledgments: The authors thank Feist-Weiller Cancer Center for its support.
This article is licensed under a CC Attribution-NonCommercial-NoDerivatives 4.0 License

How to cite this article: Abdullah S, Herbert C, Shoaib F, Bourgeois C, Gaddam SJ. A rare case of subglottic Rosai-Dorfman-Destombes disease causing fixed upper-airway obstruction in an adult woman. *EJCRIM* 2026;**13**:doi:10.12890/2026_006700

ABSTRACT

Introduction: Rosai–Dorfman–Destombes disease (RDD) is a rare non-Langerhans cell histiocytosis that usually presents with painless cervical lymphadenopathy but may involve extranodal sites. Laryngeal or subglottic involvement is uncommon yet clinically significant because progressive fixed upper airway obstruction may initially resemble more common obstructive airway disease.

Case description: We report a 59-year-old lifelong non-smoker with a history of mediastinal lymphadenopathy who developed several months of progressive exertional dyspnoea and wheeze treated as moderate persistent asthma with escalating inhaled therapy and repeated systemic steroids, without sustained improvement. Pulmonary evaluation demonstrated only mild obstructive physiology with air trapping and reduced diffusing capacity, discordant with the patient's marked exertional limitation. On follow-up, she was noted to have audible biphasic stridor, prompting urgent evaluation for central airway obstruction. Flexible laryngoscopy revealed an exophytic subglottic mass causing approximately 50% fixed airway obstruction. She underwent tracheostomy for airway protection followed by excision of the lesion. Histology demonstrated a lymphoplasmacytic infiltrate with large S100 and CD68 positive histiocytes showing emperipolesis, consistent with extranodal RDD. Cross sectional imaging excluded multifocal disease. She was decannulated and remains asymptomatic under surveillance.

Conclusion: This case underscores that fixed central airway obstruction can masquerade as refractory asthma and that biphasic stridor may be the critical bedside clue prompting reconsideration of the diagnosis. Although subglottic RDD is rare, delayed recognition may lead to life-threatening airway compromise. Compared with the usual nodal presentation and often indolent clinical course of RDD, isolated subglottic disease is unusual and clinically significant because diagnosis depends on biopsy of a structural airway lesion and localized disease may have a favourable prognosis after endoscopic treatment and surveillance.

KEYWORDS

Rosai–Dorfman–Destombes disease, subglottic mass, fixed upper-airway obstruction, biphasic stridor

EFIM
European Federation of Internal Medicine

Publisher
smc media srl

LEARNING POINTS

- Progressive wheeze and dyspnoea with limited response to guideline-directed asthma therapy, particularly when symptom severity is disproportionate to spirometry, should prompt evaluation for non-asthma causes of airflow obstruction.
- Biphasic stridor is a key red flag for fixed central airway obstruction and warrants urgent airway visualization.
- Localized subglottic Rosai–Dorfman–Destombes disease, although rare, can be managed effectively with surgical debulking and temporary airway protection; systemic therapy is reserved for multifocal or organ threatening disease.

INTRODUCTION

Asthma is often empirically diagnosed in adults presenting with wheeze and airflow obstruction; however, fixed upper airway obstruction can produce similar symptoms and may masquerade as "refractory asthma." Recognition of discordance between symptoms and pulmonary function and awareness of red flag features are essential for timely diagnosis.

Rosai–Dorfman–Destombes disease (RDD) is a rare histiocytic disorder classified within the "R group" histiocytoses[1]. Nodular disease classically presents with massive cervical lymphadenopathy; nonetheless, extranodal involvement occurs in up to 50% of patients and may affect the larynx or trachea[2]. These airway manifestations are uncommon yet clinically important because progressive obstruction may be misdiagnosed as asthma and treated inappropriately[3].

We describe a case of subglottic RDD presenting with progressive wheeze and dyspnoea that was initially managed as asthma. The objective of this report is to highlight the importance of recognizing biphasic stridor and re-evaluating an asthma diagnosis when symptoms and spirometry are discordant while emphasizing that the definitive diagnosis was established by biopsy of the airway lesion.

CASE DESCRIPTION

The patient was a 59-year-old African American woman with hypertension, type 2 diabetes mellitus, prior cerebrovascular accident without residual deficits, obesity and obstructive sleep apnoea. She was a lifelong non-smoker and worked as a teacher. Three years before the index presentation, chest imaging performed during stroke evaluation revealed calcified mediastinal and hilar lymph nodes and a right upper lobe nodule, interpreted as healed granulomatous disease due to prior histoplasmosis exposure. Serial computed tomography (CT) scans in 2022 and 2023 showed no progression *(Fig. 1)*.

In October 2024 she developed progressive exertional dyspnoea, wheeze and nocturnal respiratory symptoms. Initial spirometry in November 2024 demonstrated mild obstruction: forced expiratory volume in one second (FEV_1) 1.75 l (86% predicted), forced vital capacity (FVC) 2.66 l (105% predicted) and FEV_1/FVC ratio = 0.66; total lung capacity was normal at 107 % predicted, residual volume/total lung capacity ratio was 46% and diffusing capacity was moderately reduced at 59% predicted *(Fig. 2)*. These findings suggested mild obstructive physiology with air trapping but did not explain her marked functional limitation.

She was treated empirically as having moderate persistent asthma with stepwise escalation of therapy: a twice daily fluticasone 250 µg/salmeterol 50 µg inhaler, then fluticasone plus montelukast 10mg nightly, and eventually budesonide/formoterol 160/4.5 µg two puffs twice daily. Despite adherence, she remained symptomatic and required multiple emergency department visits and hospitalizations during the summer of 2025 for presumed asthma exacerbations treated with systemic corticosteroids, magnesium and nebulized bronchodilators.

At pulmonary follow up in September 2025 she reported severe exertional dyspnoea, walking only 10–20 steps before stopping. On examination she was dyspnoeic at rest with audible biphasic stridor. Auscultation revealed diffuse wheeze without focal crackles. Because her symptom severity was disproportionate to spirometric findings, upper airway pathology was suspected, and she was referred to the emergency department.

On presentation she was hemodynamically stable and well oxygenated on room air. Laboratory tests were unremarkable (haemoglobin 14.0 g/dl, white blood cell



Figure 1. *Computed tomography of the chest (2022), axial and coronal images showing chronic calcified mediastinal/hilar nodes.*

Spirometry

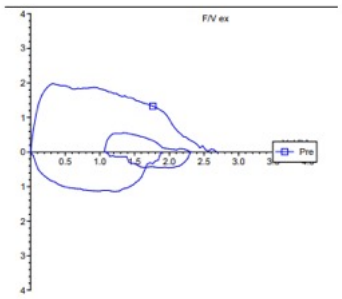
		Ref	LLN	Pre	Pre%Ref
FVC	L	2.54	1.88	2.66	104.7
FEV 1	L	2.03	1.48	1.75	86.3
FEV1/FVC	%	80	68	66	82.1
FEF 25-75%	L/s	1.94	0.85	1.55	79.6
PEF	L/s	5.35	3.47	2.01	37.5
FET 100%	sec			6.49	

Lung Volumes

		Ref	LLN	Pre	Pre%Ref
TLC	L	4.60	3.62	4.91	106.6
VC	L	2.54	1.88	2.66	104.7
FRCpleth	L	2.59	1.76	3.03	117.3
ERV	L	0.79		0.78	98.9
RV	L	1.79	1.22	2.25	125.4
RV/TLC	%	39	29	46	117.5
Raw	cmH2O*s/L	3.06	3.06	8.04	262.9

Diffusing Capacity

		Ref	LLN	Pre	Pre%Ref
DLCO	ml/(min*mmHg)	21.65	15.92	12.72	58.8
DLCO Adj	ml/(min*mmHg)	21.65	15.92		
DLCO/VA	ml/(min*mmHg*L)	4.70	3.10	3.73	79.4
DL/VA Adj	ml/(min*mmHg*L)	4.70	3.10		
VA	L	4.45	4.45	3.41	76.5
IVC Single Breath	L	2.54	1.88	1.85	72.7



Therapist Comments:
meets ats standard for acceptability but not reproducibility

Physician Interpretation:
Mild obstruction on spirometry and flow volume loops
Normal lung volumes with evidence of air trapping

Figure 2. Spirometry and flow–volume loop demonstrating mild obstruction with air trapping and reduced diffusing capacity.

count 8.95 ×10⁹/l, platelets 321 ×10⁹/l, eosinophils 0.3%, creatinine 1.0 mg/dl). Chest radiography showed chronic bilateral lower lobe fibrotic changes without acute abnormalities. She received nebulized racemic epinephrine and bronchodilators with minimal improvement, prompting otolaryngology consultation.

Flexible laryngoscopy revealed a pedunculated exophytic mass arising from the right subglottis obstructing approximately 50% of the airway lumen *(Fig. 3)*. She underwent awake tracheostomy for airway protection followed by direct laryngoscopy, rigid tracheoscopy and endoscopic excision of the lesion. Intraoperatively, a narrow based, pedunculated mass was seen in the right subglottis nearly occluding the airway; no other lesions were identified in the upper aerodigestive tract.

Histopathological examination of the excised lesion revealed a subepithelial lymphoplasmacytic infiltrate with numerous large histiocytes exhibiting emperipolesis *(Fig. 4)*. Immunohistochemistry showed that the histiocytes were positive for S100 and CD68 and negative for CD1a; cyclin D1 and OCT 2 were also expressed *(Fig. 5)*. These biopsy findings were diagnostic of extranodal RDD. Periodic acid–Schiff, Grocott methenamine silver and Ziehl–Neelsen stains were negative for microorganisms. Cross sectional staging with CT scan of the neck, chest, abdomen and pelvis demonstrated calcified mediastinal lymph nodes and patchy ground glass opacities but no additional lesions. Next generation sequencing of the specimen was performed to evaluate for MAPK pathway or other targetable alterations; it demonstrated a low tumour mutational burden (0.8 mutations/Mb), microsatellite stability and no actionable genetic alterations.

The patient recovered well postoperatively. Follow up flexible laryngoscopy showed a widely patent airway with a small smooth residual area below the right arytenoid considered clinically insignificant. She was decannulated and reported complete resolution of dyspnoea and wheeze. She continues to be monitored by otolaryngology and haematology every 6–12 months with endoscopic surveillance or CT imaging. At the time of writing, she remains asymptomatic without recurrence.

DISCUSSION

This case emphasizes the potential for fixed central airway obstruction to masquerade as refractory asthma. The



Figure 3. Flexible laryngoscopy showing an exophytic subglottic mass obstructing approximately 50% of the airway.

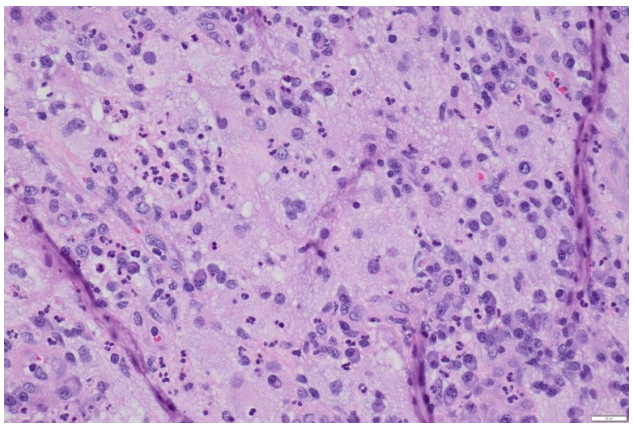


Figure 4. Subglottic lesion demonstrating numerous large histiocytes with abundant pale cytoplasm and prominent emperipolesis, characterized by intact intracytoplasmic lymphocytes and other leukocytes. A mixed inflammatory background is present. Hematoxylin and eosin; original magnification ×40.

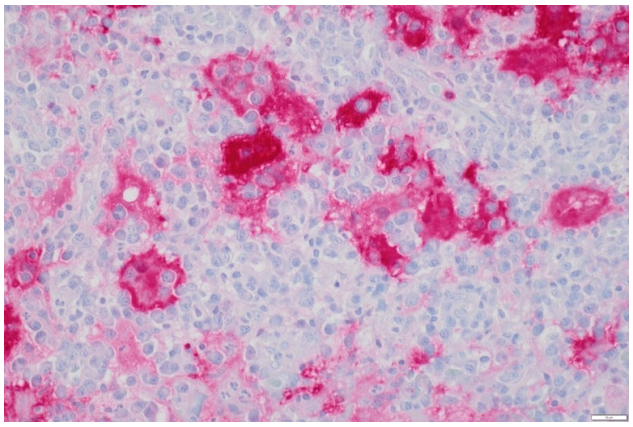


Figure 5. Subglottic lesion demonstrating diffuse cytoplasmic positivity of histiocytes on S100 immunohistochemistry, highlighting large histiocytes with prominent emperipolesis (intracytoplasmic intact lymphocytes/leukocytes). S100 immunohistochemical stain, red chromogen; original magnification ×40.

combination of progressive dyspnoea, wheeze, limited response to escalating inhaled therapy and systemic corticosteroids, and symptom severity out of proportion to spirometry should prompt evaluation for alternative diagnoses. Recognition of biphasic stridor was pivotal in this patient; stridor suggests obstruction in the larynx, subglottis or trachea and warrants urgent airway visualization. Importantly, however, the key diagnostic step was not clinical suspicion of RDD itself, but identification of a structural airway lesion followed by biopsy.

RDD is a rare histiocytic disorder. In its classic form, it most commonly affects children and young adults, presenting with painless bilateral cervical lymphadenopathy and constitutional features such as fever and elevated inflammatory markers[2]. The nodal form usually follows an indolent, self-limited course, with spontaneous remission reported in approximately 50% of cases and an overall favourable long-term prognosis. Extranodal involvement occurs in about 40–50% of patients and most commonly affects the skin, orbit, or central nervous system, with prognosis varying according to the site and extent of disease[2]. In contrast, laryngeal or subglottic involvement is distinctly uncommon but clinically significant because it may present with progressive airway compromise rather than the usual nodal picture. Airway RDD commonly presents with hoarseness, dysphonia, cough, wheeze, or stridor and may be misdiagnosed as asthma or chronic bronchitis prior to identification of fixed obstruction. Unlike the more typical nodal form, airway RDD is less likely to regress spontaneously and may show progressive local growth with risk of life-threatening mechanical obstruction if unrecognized[3-5]. Histologically, RDD is characterized by large pale histiocytes containing intact lymphocytes (emperipolesis) within a mixed inflammatory background. Immunophenotyping shows S100 and CD68 positivity and CD1a negativity, and expression of cyclin D1 and OCT-2 may help confirm the diagnosis in fibrotic or paucicellular lesions[6].

Management of RDD is individualized. Asymptomatic or minimally symptomatic disease can be observed, whereas organ-threatening disease requires active treatment. Localized airway lesions are usually treated with endoscopic excision or debulking, sometimes with adjunctive corticosteroids or radiotherapy; systemic therapy (corticosteroids, cytotoxic agents or targeted therapies directed at MAPK/ERK pathway mutations) is reserved for multifocal, progressive or organ-threatening disease[2]. In our patient, the absence of multifocal disease and complete clinical response to local therapy supported expectant management. One cannot exclude the possibility that the patient's long-standing calcified mediastinal adenopathy represented an earlier or concurrent manifestation of RDD, though the calcified, stable appearance and absence of tissue confirmation make this speculative. Clinicians should avoid diagnostic anchoring on remote imaging findings and should reassess the working diagnosis when new symptoms arise.

Finally, this report underscores the importance for internists and generalists of re-evaluating an asthma diagnosis when the clinical picture does not fit. Severe or progressive symptoms with limited bronchodilator response, discordance between symptom burden and spirometry and the presence of stridor warrant early laryngoscopy or bronchoscopy. Timely recognition of fixed upper-airway obstruction can prevent unnecessary treatments and avert airway emergencies[7]. When a structural airway lesion is identified, biopsy is what ultimately secures the diagnosis and distinguishes unusual entities such as RDD from more common neoplastic or inflammatory causes.

REFERENCES

1. Emile JF, Abla O, Fraitag S, Horne A, Haroche J, Donadieu J, et al. Revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages. *Blood* 2016;**127**:2672-2781. doi: 10.1182/blood-2016-01-690636
2. Abla O, Jacobsen E, Picarsic J, Krenova Z, Jaffe R, Emile JF, et al. Consensus recommendations for the diagnosis and clinical management of Rosai-Dorfman-Destombes disease. *Blood* 2018;**131**:2877-2890. doi: 10.1182/blood-2018-03-839753
3. Niu Y, Li Y, Wang J, Jin X, Yang D, Huo H, et al. Laryngeal Rosai-Dorfman Disease (Sinus Histiocytosis with Massive Lymphadenopathy): A Retrospective Study of 5 Cases. *Biomed Res Int* 2017;**2017**:8521818. doi: 10.1155/2017/8521818
4. Snoussi M, Derbel A, Hentati Y, Mnif Z, Marzouk S, Bahloul Z. Subglottic Masses Revealing Rosai-Dorfman Disease. *Eur J Case Rep Intern Med* 2023;**10**:003389. doi: 10.12890/2023_003389
5. Kokje V, De Vito C, Varela FC, Monnier Y, Dulguerov N, Becker M, et al. Rosai-Dorfman disease presenting as stridor and hoarseness in a young female patient. *Radiol Case Rep* 2022;**18**:591-595. doi: 10.1016/j.radcr.2022.11.018
6. Ravindran A, Rech KL. How I Diagnose Rosai-Dorfman Disease. *Am J Clin Pathol* 2023;**160**:1-10. doi: 10.1093/ajcp/aqad047
7. Mokoka M, Ullah K, Curran DR, O'Connor T. Rare causes of persistent wheeze that mimic poorly controlled asthma. *BMJ Case Rep* 2013;**2013**:bcr2013201100. doi: 10.1136/bcr-2013-201100.