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(CASE REPORT)

Rosai–Dorfman disease of the thyroid presenting as a rapidly progressive airway-compromising mass mimicking anaplastic thyroid carcinoma: A case report

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Abstract

Background: Rosai–Dorfman–Destombes disease (RDD) is a rare histiocytic disorder with heterogeneous nodal and extranodal manifestations. Thyroid involvement is exceptional and may closely simulate aggressive thyroid malignancy. Case presentation: A 75-year-old woman presented with a rapidly enlarging, painful anterior cervical mass and dysphonia. Cervical ultrasonography demonstrated a multinodular goiter with a 7-cm right thyroid nodule classified as EU-TIRADS 4. Fine-needle aspiration was categorized as Bethesda V. Four days later, the patient developed acute upper-airway compromise; computed tomography showed approximately 80% tracheal luminal narrowing and pulmonary foci initially considered suspicious for secondary disease. Emergency surgical exploration was undertaken. Complete thyroidectomy was precluded by apparent common carotid artery involvement, and tumor debulking with tracheostomy was performed. Frozen-section examination showed eosinophilic tumor-like cells with malignant-appearing features but did not establish anaplastic thyroid carcinoma. Definitive histopathological examination and immunohistochemical assessment supported a non-Langerhans histiocytosis consistent with RDD. Corticosteroid therapy was initiated and the patient was referred for oncologic evaluation; she was subsequently lost to follow-up. Conclusion: Thyroid RDD may present as a rapidly progressive, locally aggressive, airway-threatening mass and can be cytologically and intraoperatively indistinguishable from anaplastic thyroid carcinoma. Definitive tissue assessment is essential before disease-specific oncologic treatment is instituted.

Keywords: Rosai–Dorfman Disease; Thyroid; Non-Langerhans Cell Histiocytosis; Anaplastic Thyroid Carcinoma; Airway Obstruction; Tracheostomy; Case Report

1. Introduction

Rosai–Dorfman–Destombes disease (RDD), historically termed sinus histiocytosis with massive lymphadenopathy, is a rare histiocytic disorder characterized by the accumulation of distinctive histiocytes within nodal or extranodal tissues [1,2]. The contemporary Histiocyte Society classification places RDD within the "R group" of histiocytoses, reflecting advances in its clinicopathological and molecular characterization [2]. Although the classical phenotype consists of massive, painless cervical lymphadenopathy, extranodal disease is well recognized and may involve the skin, upper respiratory tract, bone, orbit, and central nervous system [3,4].

Thyroid involvement remains exceptionally uncommon. Published thyroid cases have emphasized a recurrent diagnostic problem: RDD can mimic thyroid carcinoma, anaplastic thyroid carcinoma, or primary thyroid lymphoma on clinical, radiological, and cytological grounds [5–8]. In rapidly enlarging thyroid masses, this distinction is critical because the anticipated prognosis and therapeutic strategy differ substantially. We report an unusual airway-threatening thyroid RDD in a 75-year-old woman, initially managed as a highly aggressive thyroid malignancy. The case

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is notable for its rapid clinical deterioration, severe tracheal compression, apparent vascular involvement, and misleading preoperative and frozen-section findings.

2. Case Presentation

A 75-year-old woman presented to our otorhinolaryngology and head and neck surgery department with a painful, progressively enlarging midline anterior cervical swelling associated with dysphonia. Physical examination revealed an approximately 8-cm hard, multilobulated cervical mass with irregular contours and heterogeneous consistency. The lesion was tender on palpation.

Initial flexible nasofibroscopy demonstrated leftward deviation of the laryngeal structures without vocal-fold paresis or paralysis. Cervical ultrasonography showed a heterogeneous multinodular goiter. The dominant lesion was a 7-cm nodule occupying the right thyroid lobe and was classified as EU-TIRADS 4. Fine-needle aspiration cytology was subsequently categorized as Bethesda V, suspicious for malignancy. The patient was clinically and biochemically euthyroid, and serum calcitonin was negative. In view of the cytological suspicion of malignancy, total thyroidectomy with intraoperative frozen-section assessment was planned.

Four days after fine-needle aspiration, the clinical course changed abruptly. The patient developed laryngeal dyspnea. Computed tomography demonstrated severe tracheal compression with approximately 80% reduction of the tracheal lumen. Pulmonary foci were also identified and were initially considered suspicious for secondary involvement. Given the impending airway compromise and the strong suspicion of an aggressive thyroid malignancy, urgent surgical management was undertaken.

At surgical exploration, the thyroid mass appeared locally invasive, with apparent involvement of the common carotid artery. A complete thyroidectomy could therefore not be safely achieved. Tumor debulking was performed to reduce mass effect, together with tracheostomy for airway protection. Intraoperative frozen-section examination identified tumor-like cells with eosinophilic cytoplasm and malignant-appearing features but did not confirm anaplastic thyroid carcinoma.

Definitive histopathological examination of the surgical specimen, supplemented by immunohistochemical analysis, supported a diagnosis of non-Langerhans cell histiocytosis consistent with Rosai–Dorfman disease. The diagnosis fundamentally revised the initial oncologic hypothesis. Corticosteroid therapy was initiated, and the patient was referred to oncology for multidisciplinary assessment and further management. Unfortunately, she was subsequently lost to follow-up, precluding evaluation of long-term response, recurrence, and the evolution of the pulmonary findings.

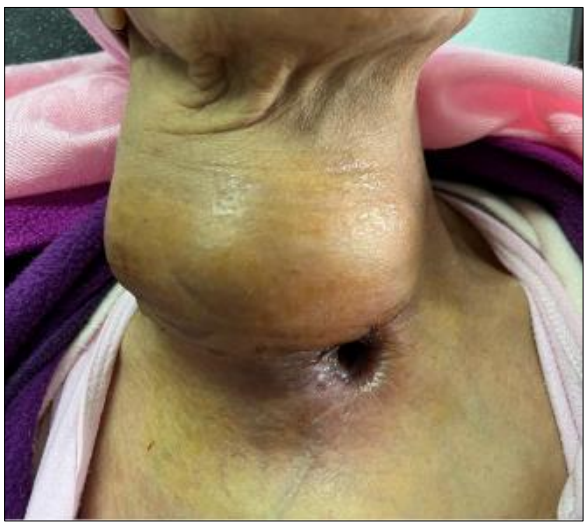

Figure 1 Clinical appearance of the anterior cervical mass after tracheostomy.

3. Discussion

RDD is a rare and biologically heterogeneous histiocytic disorder. Its histological hallmark is an infiltrate of large histiocytes, classically showing emperipolesis, in which intact lymphocytes or other inflammatory cells are present

within the histiocytic cytoplasm [1,3]. The typical immunophenotype includes expression of S100 and macrophage-associated markers such as CD68 and CD163, with absence of CD1a expression; however, diagnosis requires integration of morphology, immunophenotype, and clinical context [3,4]. This distinction is particularly important in extranodal disease, where fibrosis, limited sampling, and less conspicuous emperipolesis may complicate interpretation [3].

Thyroid RDD is exceedingly rare. Early reports by Powell et al. and Lee et al. illustrated that thyroid involvement may be mistaken for anaplastic carcinoma and may lead to an aggressive initial management strategy [5,6]. Chhabra et al. further demonstrated the diagnostic contribution of fine-needle aspiration in nodal and thyroid disease, while also emphasizing the rarity of thyroid localization [7]. More recently, Kim et al. described the substantial histopathological overlap encountered when distinguishing anaplastic thyroid carcinoma, primary thyroid lymphoma, and RDD in a thyroid mass [8]. The present case belongs to this clinically important mimicry spectrum.

Several features made malignancy the dominant initial diagnosis in our patient: advanced age, rapid enlargement of a large thyroid mass, dysphonia, Bethesda V cytology, severe tracheal compression, radiologically suspicious pulmonary foci, and apparent common carotid artery involvement at surgery. The clinical phenotype was therefore highly compatible with anaplastic thyroid carcinoma. Moreover, the acute deterioration with approximately 80% tracheal narrowing created an airway emergency in which immediate decompression and tracheostomy took precedence over prolonged diagnostic work-up. In this setting, the surgical decision was driven by airway rescue rather than by definitive oncologic resection.

The discordance between the preoperative hypothesis and definitive pathology is a central teaching point. Fine-needle aspiration can suggest RDD when characteristic histiocytes and emperipolesis are adequately sampled, but cytological diagnosis is not uniformly straightforward [7]. Likewise, frozen-section interpretation may be challenging in an inflamed, histiocyte-rich thyroid lesion. In our patient, eosinophilic malignant-appearing cells raised concern for aggressive neoplasia, yet anaplastic carcinoma could not be confirmed intraoperatively. The final diagnosis required definitive histopathological and immunohistochemical evaluation. Accordingly, when the pathology of a rapidly enlarging thyroid mass is atypical or internally discordant, RDD should be considered before committing the patient to irreversible disease-specific oncologic treatment.

The clinical behavior of RDD is variable. A substantial proportion of patients may have an indolent or self-limited course, whereas others develop symptomatic, recurrent, or organ-threatening disease [3,4,9]. In a contemporary clinicopathological series, extranodal and multisystem manifestations contributed to marked heterogeneity in presentation, treatment, and outcome [9]. Our case demonstrates that the conventional description of RDD as a benign disease should not obscure its potential for major local morbidity. Severe airway compression represents organ-threatening disease regardless of the non-metastatic biological classification of the lesion.

Management should be individualized according to symptoms, anatomical site, resectability, and organ compromise. Consensus recommendations support observation in selected uncomplicated cases, surgery for accessible symptomatic or potentially curable localized disease, and systemic therapy for multifocal, refractory, or organ-threatening manifestations [3]. Corticosteroids are frequently used, although responses may be variable. Other systemic strategies have included cladribine, methotrexate-based regimens, immunomodulatory agents, and targeted inhibition of the MAPK pathway in selected molecularly characterized cases [3,9,10]. The recognition of recurrent MAPK-pathway alterations in a subset of RDD has further challenged the historical view of the disease as purely reactive and has opened the possibility of genotype-directed therapy [10].

In our patient, complete thyroidectomy was not feasible because of apparent carotid involvement. Debulking and tracheostomy were appropriate for immediate control of life-threatening airway obstruction. Corticosteroids were initiated after the pathological diagnosis, consistent with a systemic therapeutic approach for symptomatic disease. Nevertheless, the absence of longitudinal follow-up is an important limitation. We cannot determine the durability of airway control, the response to corticosteroids, the behavior of the residual cervical disease, or whether the pulmonary foci represented RDD, an unrelated process, or another pathology. A complete staging evaluation and multidisciplinary follow-up would have been particularly important in this patient.

This case expands the clinical spectrum of thyroid RDD by illustrating an unusually aggressive local presentation with abrupt airway deterioration and apparent vascular invasion. For head and neck surgeons, endocrinologists, radiologists, and pathologists, the practical message is not that RDD should supersede malignancy in the initial differential diagnosis of a rapidly enlarging thyroid mass, but that it should remain a recognized diagnostic mimic when cytological, intraoperative, and definitive pathological findings are discordant.

4. Conclusion

Rosai–Dorfman disease of the thyroid is an exceptional diagnostic mimic of aggressive thyroid malignancy. In the present case, a rapidly enlarging thyroid mass with Bethesda V cytology, severe tracheal compression, suspicious pulmonary findings, and apparent carotid involvement closely simulated anaplastic thyroid carcinoma and culminated in an airway emergency. Definitive histopathological and immunohistochemical assessment was decisive. Thyroid RDD should therefore be considered in the differential diagnosis of rapidly progressive thyroid masses when pathological findings are atypical or discordant. Early tissue confirmation and multidisciplinary assessment are essential to avoid inappropriate oncologic treatment while ensuring timely management of organ-threatening complications.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no competing interests.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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Data availability

All data relevant to this case are included in the article. Additional clinical data are unavailable because the patient was lost to follow-up.

References

[1] Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. Arch Pathol. 1969;87(1):63–70.

[2] 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(22):2672–2681. doi:10.1182/blood-2016-01-690636.

[3] 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(26):2877–2890. doi:10.1182/blood-2018-03-839753.

[4] Bruce-Brand C, Schneider JW, Schubert P. Rosai-Dorfman disease: an overview. J Clin Pathol. 2020;73(11):697–705. doi:10.1136/jclinpath-2020-206733.

[5] Powell JG, Goellner JR, Nowak LE, McIver B. Rosai-Dorfman disease of the thyroid masquerading as anaplastic carcinoma. Thyroid. 2003;13(2):217–221. doi:10.1089/105072503321319512.

[6] Lee FY, Jan YJ, Chou G, Wang J, Wang CC. Thyroid involvement in Rosai-Dorfman disease. Thyroid. 2007;17(5):471–476. doi:10.1089/thy.2006.0323.

[7] Chhabra S, Agarwal R, Garg S, Singh H, Singh S. Rosai-Dorfman disease: a case report with extranodal thyroid involvement. Diagn Cytopathol. 2012;40(5):447–449. doi:10.1002/dc.21618.

[8] Kim S, Gray AL, Lao WP, Perez MN, Liu Y, Lee SC. Is it anaplastic thyroid cancer, primary thyroid lymphoma, or Rosai Dorfman disease? An elusive histopathologic diagnosis of a thyroid mass. Head Neck Pathol. 2022;16(2):507–512. doi:10.1007/s12105-021-01392-8.

[9] Goyal G, Ravindran A, Young JR, Shah MV, Bennani NN, Patnaik MM, et al. Clinicopathological features, treatment approaches, and outcomes in Rosai-Dorfman disease. Haematologica. 2020;105(2):348–357. doi:10.3324/haematol.2019.219626.

[10] Garces S, Medeiros LJ, Patel KP, Li S, Pina-Oviedo S, Li J, et al. Mutually exclusive recurrent KRAS and MAP2K1 mutations in Rosai-Dorfman disease. Mod Pathol. 2017;30(10):1367–1377. doi:10.1038/modpathol.2017.55.