Systematic Review

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Submitted: 30 September 2023 Revised: 27 November 2023 Accepted: 5 December 2023 Published: 28 December 2023

Abstract

Background: Rosai-Dorfman disease is a rare condition that typically presents as a nodal disease. Cardiac involvement is extremely uncommon, occurring in 0.1-0.2% of cases, which has hindered our understanding. We report a case of Rosai-Dorfman disease (RDD) related cardiac manifestation in a patient without nodal involvement. Further, we conduct a comprehensive review of the literature to consolidate data on how patients with cardiac manifestations of RDD are typically managed and treated. Methods: A systematic review of PubMed, Web of Science, and Embase was conducted to identify cases of RDD with cardiac involvement. Out of 464 studies identified, 42 publications encompassing 43 patients met the criteria and were incorporated in this review. We gathered data on patient demographics, as well as their management and treatment approaches. Additionally, we share our own experience with a patient who presented with a cardiac mass related to RDD. **Results**: Out of the 43 patients, only 20.9% (n = 9) had a documented history of RDD prior to cardiac manifestations. Nodal involvement was reported in 32.6% (n = 14), while extranodal extracardiac involvement was reported in 46.5% (n = 20). Upon presentation, the most prevalent symptoms were dyspnea (48.8%, n = 21), chest discomfort (41.9%, n = 18), and lower extremity edema (16.3%, n = 7). Cardiac manifestations were most frequently found in the right atrium (41.9%, n = 18) and pericardium (18.6%, n = 8). Treatment encompassed systemic medical therapy (34.9%, n = 15) and cardiac surgery (39.5%, n = 17). The median follow-up period was 12 months (with a range of 1 to 36), and 8 patients (18.6%) experienced mortality. Our patient, who had a cardiac mass in the left atrium, underwent resection and has remained symptom-free without any recurrence for the past 5 years. Conclusion: The frequency of cardiac related-RDD manifestations may be greater than initially perceived. These results underscore the significance of identifying RDD and its cardiac-related presentations, facilitating timely diagnosis and treatment for affected individuals.

Keywords

Rosai-Dorfman disease; cardiac mass; myxoma; histiocy-tosis

Introduction

Rosai-Dorfman disease (RDD), also known as sinus histiocytosis with massive lymphadenopathy, is a rare histiocytic disorder of non-neoplastic nature [1]. First described in 1965 [2], this disease entity was characterized by Rosai and Dorfman in 1969 and 1972 [3,4], with further delineation by additional reports [5]. Rosai-Dorfman disease is a rare disease that typically presents as a nodal disease in young children. The etiology of the disease is currently unknown. Although concurrent extranodal involvement of various systems and tissues have been reported, manifestation in the cardiovascular system is extremely uncommon. Moreover, isolated extranodal involvement has been reported in only a handful of cases, limiting our understanding of this disease. We report a case of Rosai-Dorfman disease in the heart, without clinical evidence of nodal involvement. Additionally, we perform a systematic review of patients with cardiac manifestations of RDD which aims to summarize the data regarding patterns of patient presentation, management, and treatment of this patient population.

Materials and Methods

Case Presentation

With the patient's consent, we report a case of a 46year-old otherwise healthy female (body mass index 39.4) who presented with progressively worsening dyspnea on exertion. She otherwise denied fever, chest pain, cough, sputum, recent illness, night sweats, leg swelling, paroxysmal nocturnal dyspnea, or weight loss. Her vital signs were normal. Physical examination, including cardiovascular, respiratory, abdominal, and skin, were unremarkable.

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No head and neck, suprascapular, axillary, or inguinal lymphadenopathy was appreciated.

At this time, complete blood count was only significant for leukocytosis (14,080 cells/ μ L), anemia (hemoglobin count of 9.7 g/dL, and hematocrit of 29.8%), and thrombocytopenia (118,000/ μ L). Other laboratory parameters were unremarkable. Electrocardiogram revealed sinus rhythm with first degree atrioventricular block. Transthoracic echocardiography (TTE) demonstrated a 4.0 \times 3.0 \times 3.0 cm mass in the left atrium (LA) prompting referral for surgical evaluation.

Preoperative transesophageal echocardiography (TEE) revealed a 6.0×3.4 cm mass in the left interatrial septum with projection into the LA appendage and the anterior leaflet of the mitral valve, resulting in reduced mobility of the mitral valve leaflet. TEE also revealed moderate-to-severe mitral regurgitation with no evidence of mitral stenosis and moderate dilation of the LA. The left ventricular ejection fraction was preserved at >55%. The patient was suspected to have an atrial myxoma for which she was advised to undergo surgical resection.

After median sternotomy, central cannulation, and induction of cardiopulmonary bypass, a right atriotomy was performed. A solid pink-to-tan mass was visualized originating from the interatrial septum and protruding bilaterally into both the atrial chambers, extending apically towards the interventricular septum for unknown length. The tip of the right atrial protrusion was in proximity to the inferior vena cava and the left atrial protrusion was riding over the anterior leaflet of the mitral valve. An intraoperative frozen section was obtained, revealing a benign neoplasm with dystrophic calcification. Although the relatively extensive process of the mass prevented a complete resection, a radical septectomy with reconstruction using bovine pericardial patch was performed. The subsequent pathology report of the excised mass revealed RDD.

Follow up TTE at 4 months revealed a normal ejection fraction without any recurrence of the LA mass. The patient has been symptom free with no evidence of recurrence for the past 5 years.

Systematic Review Method

Search Strategy

In accordance with the Preferred Reporting Items for Systematic Review and Meta-Analyses (PRISMA) [6], three databases (PubMed, Embase, and Web of Science) were systematically searched to identify all published cases on cardiac manifestations of RDD from each database's inception until July 30th, 2023 (**Supplementary Table 3**). We used the Covidence Systematic Review Software for article screening and data extraction tool for conducting systematic reviews. Search term "Rosai-Dorfman" was used interchangeably with the terms "heart", "cardiac", "pericardium", "pericardial", "epicardium", "epicardial", "myocardium", "myocardial", "atria", "ventricle", "cardiovascular", or "artery", using the Boolean operator AND. Two authors independently screened the titles and abstracts found in the initial search and extracted data. Following selection, we conducted a full text review for confirmation and data extraction.

Inclusion and Exclusion Criteria

We included all literature describing cardiac manifestations in patients with a confirmed diagnosis of RDD, whether discovered prior to or during the onset of cardiac manifestations, or through autopsy findings. Cardiac diagnostic testing, such as biopsy, was not required for inclusion. The selected publications were assessed for additional references that were not detected in the initial database.

We applied the following exclusion criteria: (1) involvement of the great vessels (e.g., aorta, pulmonary artery) or mediastinal structures, without concurrent pericardial, myocardial, and endocardial involvement, (2) lack of histopathologic diagnostic testing for RDD, (3) review articles and commentaries that did not specifically describe individual cases. Fig. 1 displays the publication selection flow diagram and type of articles included.

Data Extraction

We included all data from the available literature as defined by the publishing authors. The heterogeneity of the data in the identified studies prevented a formal analysis. As a result, we conducted descriptive analyses only.

Using a predesigned collecting table, we extracted the author, year of publication, age, comorbidities, clinical presentation, diagnostic workup, treatment, follow up, and outcome when provided. One foreign language article was translated using Google translate. Pooled data and calculations in this review are derived from studies that provide individualized data.

Results

The systematic search strategy generated 464 citations and the manual full text review resulted in three additional articles. Ultimately 42 studies, including 34 case reports and 8 case abstracts, met inclusion criteria and were extracted, representing a total of 43 patients [5,7–45].

Most patients were female (60.5%) with a median age of 51 years (range 12 to 79 years). 32.6% had nodal involvement and 46.5% had extranodal extracardiac involvement. Not all non-cardiac manifestations of RDD were clinically apparent at the time of presentation and some were detected radiographically and/or during autopsy. Due to the heterogeneity of included studies, we only report cumulative data. More granular data on patient characteristics can

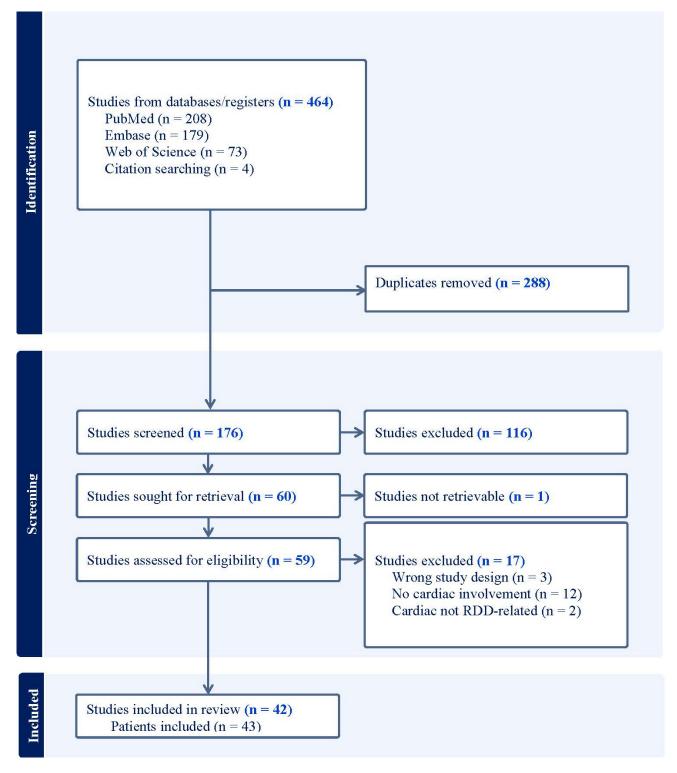


Fig. 1. Flowchart of the systematic review of the literature and study selection, detailing the inclusion and exclusion criteria.

be found in Supplementary Tables 1,2.

At the time of presentation, 20.9% had a known history of RDD, and reported symptoms of dyspnea (48.8%), chest discomfort (41.9%), and lower extremity edema (16.3%). Most patients had an echocardiogram (65.1%), cardiac magnetic resonance imaging (cMRI) (44.2%), and chest computed tomography (CT) scan (39.5%). Diagnosis of RDD was frequently with a biopsy (60.5%) or surgical resection (27.9%) in patients without a prior biopsy or those with inconclusive findings. Overall, 35 (81.4%) out of 43 patients had confirmed RDD-related cardiac manifestations, diagnosed with biopsy, surgical resection, or autopsy. The remaining 8 (18.6%) cases underwent biopsies from alternative sources (e.g., skin, lymph node, or testicle), and it was assumed that their cardiac manifestations were also related to RDD. Table 1 demonstrates presentation and workup of patients with cardiac involvement in RDD.

Cardiac involvement usually involved the right atrium (RA) (41.9%) and the pericardium (18.6%), and less frequently involved other chambers in no particular pattern. Most patients were treated with systemic medial therapy (34.9%) and/or cardiac surgery (39.5%). Table 2 demonstrates characteristics of cardiac involvement, treatment, and outcomes, where reported.

Treatment

Of 43 cases, 8 patients were treated solely with steroids. When follow-up data was provided, 3 out of 8 of these patients died, 3 developed partial/complete resolution of symptoms, and 1 developed minimal worsening of symptoms. Four patients received chemotherapy in addition to steroids. However, follow-up data is only reported for 2 patients who continued to suffer from progressive disease. One of these patients required surgical resection at the 1-year. Two patients received immunotherapy in addition to steroids. One of them required debulking at 2 months due to progressive disease, while the other showed symptom improvement at 6 months. One patient exclusively received immunotherapy but required debulking and pericardiocentesis due to extensive disease with resolution of symptoms at the 2-year follow-up.

Out of 14 patients who underwent surgical resection of their cardiac mass, only 7 provided interval follow-up outcomes, ranging from 1 to 18 months (median 5 months), none of whom experienced recurrent cardiac manifestations. One patient who underwent heart transplant had no recurrent symptoms at 18 months. Overall, there was an 18.6% mortality rate in this patient cohort. Information regarding individual cases and treatment is detailed in **Supplementary Table 2**.

Discussion

RDD is a rare histiocytic disorder that is largely considered idiopathic, with a prevalence of 1:200,000 [46]. The most common variant of RDD is sporadic in nature (e.g., nodal, extranodal, neoplasia-associated, and autoimmuneassociated), while less common forms include familial (e.g., H syndrome and autoimmune lymphoproliferative syndrome) and cutaneous [47]. Roughly 30–50% [48] of sporadic cases have lymph node involvement and up to 40% can have extranodal manifestations [49]. Generally, extranodal disease coincides with lymphadenopathy and can involve virtually all organ systems [49] with skin (10–50%), lung (10–20%), and bone (5–15%) being the most frequently affected [47,48,50]. In up to 23% [51] of instances, the initial disease manifestation of the disease solely involves extranodal symptoms, which is linked to a more aggressive course [49,52,53].

Cardiac manifestations of RDD are exceedingly rare, occurring in 0.1-0.2% of cases [54]. In the absence of extracardiac disease, RDD can be extremely difficult to diagnose. Our systematic search identified 43 patients with cardiac manifestations of RDD, most of whom presented with heart failure symptoms. Our patient's only presenting symptom was dyspnea and her LA mass was initially misdiagnosed as a myxoma, which is a common occurrence in these patients. A retrospective study (n = 38) utilizing position emission tomography-CT (PET-CT) in patients with RDD found cardiac involvement in 13.2% [55], suggesting that cardiac-related RDD manifestations may be more prevalent than previously understood-although not all instances may result in noticeable symptoms. These findings highlight the importance of recognizing RDD and cardiacrelated manifestations.

Clinical Presentation

RDD typically presents in children or young adults, with an average age 20.6 years, primarily among individuals of African American descent [46]. While RDD is generally more prevalent in males, cutaneous manifestations are most common in females [46]. The classic presentation of nodal RDD includes massive painless lymphadenopathy (cervical or generalized), fever, night sweats, and weight loss [47]. In cases of lymphomas or malignancies, constitutional symptoms can also arise. Therefore, in the absence of lymphadenopathy and/or extranodal disease, diagnosing RDD with cardiac involvement can be challenging.

Specifically in patients presenting with cardiac involvement, classical RDD symptoms were rarely present. Therefore, a high index of suspicion and histologic confirmation is required in patients presenting with heart failure symptoms and evidence of intracardiac mass.

The dominant cardiac manifestation of RDD is an intracardiac mass with or without underlying infiltration. Intracardiac masses frequently present with multifocal cardiac involvement, mainly arising from the RA (41.9%), but LA involvement is uncommon (7.0%). Intracardiac RDD is associated with hemodynamic implications such as flow obstruction as well as conduction abnormalities (e.g., AVB and electrical storm). A minority of the patients in this cohort required a permanent pacemaker placement (9.3%) or electrical cardioversion (4.7%). Pericardial involvement is also common (18.6%), which frequently manifests as pericardial effusion with or without cardiac tamponade.

Diagnosis

Given its rarity, RDD is often undiagnosed during its early stages, which may be partially responsible for frequent diagnosis of RDD in late-adulthood, even though the majority of RDD emerge during childhood [56]. Cardiac

Variable	Result
Age, Years	51 (12–79)
Female	26 (60.5)
Known History of RDD	9 (20.9)
Nodal Involvement	14 (32.6)
Extranodal Extracardiac Involvement*	20 (46.5)
Skin	7 (35.0)
Kidney	7 (35.0)
Bone	6 (30.0)
Orbital	5 (25.0)
Lung	4 (20.0)
Presenting Symptom*	
Dyspnea	21 (48.8)
Chest discomfort	18 (41.9)
Lower Extremity Edema	7 (16.3)
Arrhythmia/Palpitations	5 (11.6)
Dizziness/Presyncope	3 (7.0)
Initial Workup	
Echocardiogram	28 (65.1)
Cardiac MRI	19 (44.2)
Chest CT scan	17 (39.5)
Electrocardiogram	16 (37.2)
Chest X-Ray	9 (20.9)
PET-CT scan	9 (20.9)
Diagnostic Workup	
Biopsy†	26 (60.5)
Myocardial/Pericardial	14 (53.8)
Lymph Node	5 (19.2)
Skin	4 (15.3)
Other (adrenal, parotid, pleural)	3 (11.5)
Pericardiocentesis	2 (4.7)
Surgical Resection	12 (27.9)
Autopsy** (in patients without a prior RDD diagnosis)	3 (7.0)

Table 1. Presentation, and management of patients with Rosai-Dorfman disease-related cardiac manifestations (N = 43).

Values are presented as n (%) or median (range). *Indicates five most common variables reported are demonstrated in the table. †Excludes diagnostic studies with inconclusive results. Several cases underwent biopsies from multiple areas, detailed in **Supplementary Table 2**. **Only includes those without prior conclusive diagnostic studies. CT, Computed tomography; RDD, Rosai-Dorfman Disease; MRI, Magnetic resonance imaging; PET-CT, Positron emission tomography computer tomography.

manifestations of RDD most often trigger an investigation into progressing symptoms. Where reported, electrocardiogram (ECG) often revealed a heart block or arrhythmia. Chest X-rays were often non-specific, demonstrating pericardial/pleural effusion or cardiomegaly in some cases. Chest CT often detected an intracardiac mass, prompting additional assessment through echocardiography, cMRI and/or PET-CT scans [56].

When encountering a patient with a cardiac tumor, the range of potential diagnoses is extensive. This is due to the fact that over 99% of cardiac tumors are secondary, with many attributed to metastatic disease (e.g., breast, lung,

melanoma), particularly for right-sided tumors [57,58]. In contrast, primary cardiac tumors are rare, and almost 90% of them are benign—with approximately 80% being myxomas—and the remaining 10% are malignant, of which 95% are sarcomas and 5% are lymphomas/mesotheliomas [58].

In patients with RDD, echocardiography and cMRI features often resemble those of malignant primary tumors, attributed to local invasion into surrounding structures, location in the RA, involvement of the pericardium, and a lack of a stalk, which distinguishes it from myxomas [58]. Nevertheless, in cases where the tumor originates from the LA

Table 2. Characteristic and treatment of Rosai-Dorfman disease-related cardiac manifestations (N = 43)

Variable	Result
Size of Intracardiac Mass, cm*	2.9 (0.8–9.0)
Cardiac Involvement†	
Right Atrium	18 (41.9)
Interatrial septum	4 (9.3)
Interventricular septum	3 (7.0)
Left Atrium	3 (7.0)
Left Ventricle	3 (7.0)
Right Ventricle	3 (7.0)
Pericardium	8 (18.6)
Epicardium	4 (9.3)
Myocardium	3 (7.0)
Treatment	
Systemic Medical Therapy	15 (34.9)
Steroids	8 (53.3)
Steroids + Chemotherapy	4 (26.7)
Steroids + Immunotherapy	2 (13.3)
Immunotherapy	1 (6.7)
Cardiac Surgery	17 (39.5)
Resection	14 (82.4)
Debulking	2 (11.8)
Transplant	1 (5.9)
Permanent Pacemaker Placement	4 (9.3)
Electrical Cardioversion	2 (4.7)
Follow Up, Months**	12 (1-36)
Mortality	8 (18.6)

Values are presented as n (%) or median (range). *Size reported by n = 21 studies; †Only the major site of cardiac involvement is included, unless the patient had two or more separate nodules. **Follow-up interval was reported by n = 22 studies.

and does not infiltrate the surrounding tissue—as seen in our case—the tumor can be easily misdiagnosed as a myxoma. PET-CT scan in RDD is typically characterized by avid fludeoxyglucose-18 (FDG) uptake, likely due to underlying inflammatory process. While PET-CT cannot provide useful information concerning the intracardiac mass, it can rule out metastatic disease and identify a suitable biopsy site, which may be especially valuable for patients for those whom cardiac biopsy is not safe [10,55].

A definitive diagnosis is accomplished through histological assessment and identification of emperipolesis histiocytes containing intact lymphocytes within their cytoplasm [38]. However, the lesion often appears as a nonspecific fibroinflammatory lesion, and emperipolesis can be quite subtle, especially in cases of extranodal RDD [55]. This scenario expands the range of potential differential diagnoses to encompass inflammatory pseudotumor, fibrohistiocytic tumor, non-specific chronic inflammation, and even histiocytic sarcoma in cases displaying atypia [49,52]. This often necessitates multiple biopsies to confirm RDD. When dealing with nodal histiocytic proliferation, the list of differential diagnoses is also extensive, including sinus histiocytosis, Langerhans cell histiocytosis, infectious disorders, and lymphoproliferative disorders [59].

Immunohistochemical staining plays a critical role in distinguishing RDD. Histiocytes in RDD are positive for S100, CD68, and CD163 and negative for CD1a and CD207 [47]. RDD also shares certain morphological features with IgG4-related disease, and it often displayed elevated IgG4 to IgG ratios at both nodal and extranodal sites [47]. Current guidelines recommend incorporating immunohistochemistry for IgG4, especially when numerous plasma-cells are present. Flow Cytometry and cytogenetic testing are used to rule out lymphoproliferative disorder, but their utility in the diagnosis of RDD remains limited [38,46].

Management

The progression of RDD is often unpredictable, marked by episodes of worsening symptoms followed by periods of improvement lasting several years. RDD is often linked to a favorable prognosis, and it can resolve spontaneously in up to 50% of patients with asymptomatic RDD and/or uncomplicated adenopathy [46]. On the other hand, extranodal involvement has a poor prognosis and is fatal in approximately 7–12% of patients due to dysfunction or failure of involved vital organs [46]. Given the diverse array of ways RDD can manifest and unclear underlying etiology, there is no standardized approach to managing patients with RDD.

The approach to treatment varies depending on the extent of RDD, the affected site(s), and the presenting symptoms, necessitating an individualized approach [60]. For cases involving RDD-related cardiac manifestations, the primary approach to managing intracardiac masses and their associated symptoms is surgical resection or debulking. When reported, patients who underwent surgical resection did not experience disease progression during follow-up periods ranging from 1 to 18 months.

In patients with symptomatic RDD, multifocal extranodal disease, and nonresectable cardiac tumors, systemic treatment is required. In patients without RDD-related cardiac manifestations, corticosteroids have demonstrated favorable responses, but the optimal duration of treatment remains unclear [46]. In our cohort, 8 (18.6%) were treated solely with steroids, most of which had extensive multiorgan involvement. Three out of these 8 cases died, and the remaining cases displayed minimal disease progression or had partial/complete resolution of their symptoms. In the literature, corticosteroid therapy has been associated with frequent relapse in patients with extranodal RDD [46]. This dynamic is less clear in the context of cardiac-related RDD.

Even when steroids were used in conjunction chemotherapy (n = 4) or immunotherapy (n = 2), 2 patients required subsequent surgical intervention due to progressive disease. In patients with RDD, in the absence of cardiac manifestations, chemotherapy outcomes have been inconsistent and are typically reserved for cases that are recurrent or unresponsive to other treatments, or as an initial strategy in cases of multifocal and life-threatening disease [46]. Radiotherapy was administered in a single case and is generally reserved as a last resort for providing palliative relief.

While the available data is limited for making definitive recommendations, it seems that surgical intervention is preferred whenever feasible. Nonetheless, for patients with nonresectable cardiac involvement and extensive disease, systemic medical therapy is indicated.

Follow Up

For patients undergoing systemic therapy, generally 6-to-12 months of therapy is recommended [46]. Initial follow-up appointments should be more frequent but can be extended to intervals of 6-to-12 months once the disease has stabilized and entered remission. PET-CT scans are highly sensitive in gauging response to therapy and can be used annually following remission to monitor for potential relapse.

In patients who receive curative surgical resection and lack other extranodal manifestations, annual follow-ups utilizing echocardiogram or cMRI may be more practical [34]. This approach, coupled with comprehensive physical examinations and routine laboratory studies, can avoid unnecessary radiation exposure.

Limitations

This study is constrained by a limited sample size drawn from case reports and abstracts, which may be attributed to the rarity of RDD and the challenges associated with its diagnosis. Some patients with symptomatic RDD may not undergo diagnostic testing and/or autopsy, thus were excluded from this study. RDD also appears to be more prevalent in third world countries, where diagnostic and reporting capabilities may be limited. We did not find any retrospective studies, precluding a comprehensive statistical analysis and generalizability of our findings. Second, it is reasonable to assume a positive outcome bias in most studies. Some of the studies included may have focused only on pertinent findings and work-up, potentially influencing our results. To enhance prompt RDD diagnosis and gather prospective data, we aim to raise awareness about RDD and advocate for patient enrollment in international registries.

Conclusion

RDD is a rare histiocytic disorder that often presents with nodal involvement, but it can affect any organ, of-

ten with a poor prognosis. Cardiac involvement, reported in just 43 cases so far, is particularly uncommon. In this group, most patients lacked a history of RDD or nodal involvement, but approximately half had extra-cardiac manifestations. As a result, diagnosis of RDD is often challenging, especially when presenting with isolated cardiac masses, which can be misdiagnosed. Our patient, with no evidence of concurrent nodal or extracardiac involvement, presented with a LA mass, initially diagnosed as a myxoma based on imaging.

Diagnostic studies, when possible, are crucial for determining etiology and guiding treatment. Due to limited data and considerable heterogeneity in reported literature, the optimal management approach remains unclear. Surgical resection appears to be the most effective, with medical management reserved for unresectable masses and multiorgan involvement. Notably, our patient has remained symptom-free for 5 years after surgical resection, marking the longest reported follow-up to date.

We hope that our findings raise awareness in the medical community to improve accurate diagnosis of RDD. We encourage patient enrollment in international registries to facilitate prospective studies aimed at improving our understanding of how to best manage these patients and optimize their outcomes. The CARE checklist was used when writing this case report (**Supplementary Table 4**).

Abbreviations

cMRI, cardiac magnetic resonance imaging; CT, computed tomography; RA, Right atria; RDD, Rosai-Dorfman disease; TTE, Transthoracic echocardiography; TEE, Transesophageal Echocardiography; LA, Left atria; PET-CT, position emission tomography-computed tomography.

Author Contributions

NST performed data gathering, data analysis, manuscript writing, manuscript revisions, and coordination of the study. MHD conceived the study and reviewed the manuscript. DW and PS helped with data gathering, manuscript writing, and manuscript revisions. SNM and SS helped with data gathering and manuscript writing. All authors contributed to editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiency in the work to take public responsibility for the content and to be accountable for all aspects of the work in ensuring that accuracy and integrity of the study.

Ethics Approval and Consent to Participate

Informed content was obtained from the patient for scientific activity, including publication of this report, thus the case report was deemed exempt by the Institutional Review Board.

Acknowledgment

Not applicable.

Funding

This research received no external funding.

Conflict of Interest

The authors declare no conflict of interest.

Supplementary Material

Supplementary material associated with this article can be found, in the online version, at https://doi.org/10. 59958/hsf.6887.

References

- Lu D, Estalilla OC, Manning JT, Jr, Medeiros LJ. Sinus histiocytosis with massive lymphadenopathy and malignant lymphoma involving the same lymph node: a report of four cases and review of the literature. Modern Pathology. 2000; 13: 414–419.
- [2] Destombes P. Adenitis with lipid excess, in children or young adults, seen in the Antilles and in Mali. (4 cases). Bulletin De La Societe De Pathologie Exotique et De Ses Filiales. 1965; 58: 1169–1175. (In French)
- [3] Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. A newly recognized benign clinicopathological entity. Archives of Pathology. 1969; 87: 63–70.
- [4] Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy: a pseudolymphomatous benign disorder. Analysis of 34 cases. Cancer. 1972; 30: 1174–1188.
- [5] Richter JT, Strange RG Jr, Fisher SI, Miller DV, Delvecchio DM. Extranodal Rosai-Dorfman disease presenting as a cardiac mass in an adult: report of a unique case and lack of relationship to IgG4-related sclerosing lesions. Human Pathology. 2010; 41: 297–301.
- [6] Page MJ, McKenzie JE, Bossuyt PM, Boutron I, Hoffmann TC, Mulrow CD, et al. The PRISMA 2020 statement: an updated guideline for reporting systematic reviews. British Medical Journal. 2021; 372: n71.
- Butt AH, Peterzan M, Shah C, Wright M, Rinaldi CA, Gill J, et al. Interatrial Septal Mass Secondary to Rosai-Dorfman Disease: A Rare Cause of Complete Heart Block. JACC. Case Reports. 2023; 14: 101824.
- [8] Hritani R, Ramsey Z, Lee R, Bates WB, Williams H, Garcia D, et

al. An unusual left atrial Rosai-Dorfman involvement in sickle cell disease patient. Journal of Nuclear Cardiology. 2023; 30: 1286–1289.

- [9] Gill JS, Gill H, Green M, Kapetanakis S. Rare infiltrative cause of impaired cardiac conduction. Journal of the American College of Cardiology. 2023; 81: 3248.
- [10] Yoshida M, Zoshima T, Hara S, Takahashi Y, Nishioka R, Ito K, et al. Case report: Rosai-Dorfman disease with rare extranodal lesions in the pelvis, heart, liver and skin. Frontiers in Oncology. 2023; 12: 1083500.
- [11] Yadav A, Charate RA, Erickson MD, Platzer P, Patel DV, Manyam H. Cardiac Rosai-Dorfman disease: a rare, challenging, infiltrative pathology. Journal of the American College of Cardiology. 2023; 81: 3639.
- [12] Conte E, Brucato A, Petrella F, Passoni E, Lauri G, Bigliardi M, et al. Case Report: Pericardial Effusion Treated With Pericardiectomy Plus Right Atrial Mass Resection: A 2-Year Follow-Up of Cardiac Rosai-Dorfman Disease. Frontiers in Cardiovascular Medicine. 2021; 8: 668031.
- [13] Kondo T, Takahashi M, Yamasaki G, Sugimoto M, Kuse A, Morichika M, *et al.* Autopsy case of Rosai-Dorfman disease presenting as fibrinous pericarditis. Legal Medicine. 2021; 48: 101812.
- [14] Liang S, Huang ML, Wang B, Qiu MR. Pericardial rosaidorfman disease: the role of next generation sequencing. Pathology. 2021; 53: S32.
- [15] Maronese CA, Bonometti A, Moltrasio C, Passoni E, Berti E. Extranodal disseminated rosai-dorfman-destombes disease with cutaneous, articular and cardiac involvement. Pediatric Blood and Cancer. 2021; 68: S13–S14.
- [16] González-Pacheco H, Salas-Villela RA, Carmona-Levario P, Manzur-Sandoval D, Mora-Cervantes R, Palma-Carbajal R, et al. Electrical Storm in a Patient With Rosai-Dorfman Disease With Intracardiac Masses and Myocardial Infiltration. The Canadian Journal of Cardiology. 2020; 36: 1326.e13–1326.e15.
- [17] Tınazlı R, Tunçbilek Özmanevra P, Tuna Yalçınozan E, Erişir F. Cardiac Involvement of Rosai-Dorfman Disease Causing an Aesthetic Problem in a Young Woman. Balkan Medical Journal. 2020; 37: 222–223.
- [18] Hou G, Pan Q, Cui R, Li F, Cheng W. Rosai-Dorfman Disease of the Right Atrium Revealed by FDG PET/CT. Clinical Nuclear Medicine. 2020; 45: 709–710.
- [19] Wang TKM, Mukherjee S, Tan CD, Shepard D, Pettersson G, Desai M. Rosai-Dorfman Disease of the Right Ventricular Outflow Tract: Role of Multimodality Imaging. Circulation. Cardiovascular Imaging. 2020; 13: e010783.
- [20] Desai CK, Hayes SN, Shah MV, Maleszewski JJ, Bleeker J, Steffen K, *et al.* Extranodal Rosai-Dorfman Disease Presenting as a Pericardial Mass and Constrictive Pericarditis. JACC. Case Reports. 2019; 1: 643–647.
- [21] Yaman B, Cerit L, Kemal HS, Usalp S, Duygu H. Unusual involvement of right ventricle in patient with Rosai-Dorfman disease. Anatolian Journal of Cardiology. 2019; 22: 271–272.
- [22] Tsigaridas N, Mantzoukis S, Mpakas K, Troganis E, Patsouras D. A Rare Case of Cardiac Involvement in Rosai-Dorfman Disease. Journal of Cardiovascular Imaging. 2019; 27: 286–287.
- [23] Tarkin JM, Wolstenholme V, Sheaff M, Westwood M, Manisty C. A rare case of Rosai-Dorfman disease presenting with cardiac tamponade. European Heart Journal. Cardiovascular Imaging. 2019; 20: 718.
- [24] Khan PSJ, Sheaff M, Rizvi H. An Unusual Dual Diagnosis of Rosai-Dorfman Disease Presenting Concurrently with IgG4 Related Disease. Journal of Pathology. 2019; 249: S48.
- [25] Laubham MP, Darki A. Rosai-Dorfman disease and left ventricular noncompaction cardiomyopathy: A heart failure conundrum. Journal of Nuclear Cardiology. 2018; 25: 1971–1975.

- [26] Ghosh GC, Rebecca G, Joseph E, Joseph G. Heart failure with multiple cardiac masses: Extensive cardiac involvement in Rosai–Dorfman disease. IHJ Cardiovascular Case Reports (CVCR). 2018; 2: 12–17.
- [27] Khanna SN, Iyer KS, Paul M, Radhakrishnan S, Rathi AK. Rosai-Dorfman Disease of Right Atrium Mimicking Myxoma. The Annals of Thoracic Surgery. 2017; 103: e443–e445.
- [28] Summers MR, Pettersson G, Maalouf JF, Jaber WA. Sinus histiocytosis with massive lymphadenopathy: extra-nodal Rosai-Dorfman disease presenting as a rare aetiology of a large intracardiac mass. European Heart Journal. 2017; 38: 1439–1440.
- [29] Sendrasoa FA, Ranaivo IM, Raharolahy O, Andrianarison M, Razanakoto NH, Ramarozatovo LS, *et al.* Rosai-Dorfman Disease Involving Multiple Organs: An Unusual Case with Poor Prognosis. Case Reports in Medicine. 2016; 2016: 3920516.
- [30] Schaffer LR, Caltharp SA, Milla SS, Kogon BF, Cundiff CA, Dalal A, *et al.* Rare presentation of four primary pediatric cardiac tumors. Cardiovascular Pathology. 2016; 25: 72–77.
- [31] Moon TG, O'Gallagher K, Sado D. Infiltrative cardiomyopathy: A novel aetiology. European Journal of Heart Failure. 2016; 18: 301.
- [32] Lao IW, Dong Y, Wang J. Rosai-Dorfman disease of the pericardium: a case report and review of literature. International Journal of Clinical and Experimental Pathology. 2014; 7: 3408– 3412.
- [33] Ozbudak E, Arıkan AA, Yavuz S, Halıcı U, Berki T. A very rarely seen cardiac mass (Rosai-Dorfman disease). The Anatolian Journal of Cardiology. 2014; 14: 481–483.
- [34] Daruwalla VJ, Parekh K, Tahir H, Collins JD, Carr J. Extranodal Rosai-Dorfman Disease Involving the Left Atrium: Cardiac MRI, CT, and PET Scan Findings. Case Reports in Radiology. 2015; 2015: 753160.
- [35] Sarraj A, Zarra KV, Jimenez Borreguero LJ, Caballero P, Nuche JM. Isolated cardiac involvement of Rosai-Dorfman disease. The Annals of Thoracic Surgery. 2012; 94: 2118–2120.
- [36] Yontz L, Franco A, Sharma S, Lewis K, McDonough C. A case of Rosai-Dorfman disease in a pediatric patient with cardiac involvement. Journal of Radiology Case Reports. 2012; 6: 1–8.
- [37] Aguilar C, Carbajal T. Extranodal Rosai-Dorfman disease of pericardium presenting as large pericardial effusion: case report. Histopathology. 2012; 61: 35–35.
- [38] Ajise OE, Stahl-Herz J, Goozner B, Cassai N, McRae G, Wieczorek R. Extranodal Rosai-Dorfman disease arising in the right atrium: a case report with literature review. International Journal of Surgical Pathology. 2011; 19: 637–642.
- [39] Chen J, Tang H, Li B, Xiu Q. Rosai-Dorfman disease of multiple organs, including the epicardium: An unusual case with poor prognosis. Heart & Lung. 2011; 40: 168–171.
- [40] Maleszewski JJ, Hristov AC, Halushka MK, Miller DV. Extranodal Rosai-Dorfman disease involving the heart: report of two cases. Cardiovascular Pathology. 2010; 19: 380–384.
- [41] Da Xu J, Cao XX, Liu XP, Li QQ, Wang WJ, De Xu Z. Hydropericardium prevented diagnosis of constrictive pericarditis: an unusual case of Rosai-Dorfman disease. BMJ Case Reports. 2009; 2009: bcr0320091688.
- [42] Scheffel H, Vogt P, Alkadhi H. Cardiac manifestations of Rosai-Dorfman disease. Herz. 2006; 31: 715–716.
- [43] Bi Y, Huo Z, Meng Y, Wu H, Yan J, Zhou Y, et al. Extranodal Rosai-Dorfman disease involving the right atrium in a 60-year-

old male. Diagnostic Pathology. 2014; 9: 115.

- [44] Buchino JJ, Byrd RP, Kmetz DR. Disseminated sinus histiocytosis with massive lymphadenopathy: its pathologic aspects. Archives of Pathology & Laboratory Medicine. 1982; 106: 13– 16.
- [45] Chaitanya V, Gupta N. Isolated cardiac involvement of extranodal Rosai-Dorfman disease: an atypical presentation. Journal of Cardiology & Current Research. 2015; 3: 00099.
- [46] 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.
- [47] Bruce-Brand C, Schneider JW, Schubert P. Rosai-Dorfman disease: an overview. Journal of Clinical Pathology. 2020; 73: 697–705.
- [48] Goyal G, Young JR, Koster MJ, Tobin WO, Vassallo R, Ryu JH, et al. The Mayo Clinic Histiocytosis Working Group Consensus Statement for the Diagnosis and Evaluation of Adult Patients With Histiocytic Neoplasms: Erdheim-Chester Disease, Langerhans Cell Histiocytosis, and Rosai-Dorfman Disease. Mayo Clinic Proceedings. 2019; 94: 2054–2071.
- [49] Foucar E, Rosai J, Dorfman R. Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): review of the entity. Seminars in Diagnostic Pathology. 1990; 7: 19–73.
- [50] O'Gallagher K, Dancy L, Sinha A, Sado D. Rosai-Dorfman disease and the heart. Intractable & Rare Diseases Research. 2016; 5: 1–5.
- [51] Vargas D, Richards JC, Ocazionez D, Sirajuddin A, Browne L, Restrepo CS. Cardiothoracic manifestations of primary histiocytoses. The British Journal of Radiology. 2016; 89: 20160347.
- [52] Montgomery EA, Meis JM, Frizzera G. Rosai-Dorfman disease of soft tissue. The American Journal of Surgical Pathology. 1992; 16: 122–129.
- [53] Foucar E, Rosai J, Dorfman RF. Sinus histiocytosis with massive lymphadenopathy. An analysis of 14 deaths occurring in a patient registry. Cancer. 1984; 54: 1834–1840.
- [54] Heidarian A, Anwar A, Haseeb MA, Gupta R. Extranodal Rosai-Dorfman disease arising in the heart: clinical course and review of literature. Cardiovascular Pathology. 2017; 31: 1–4.
- [55] Zhang J, Cui R, Li J, Cao X, Luo Y. Characterizing Rosai-Dorfman disease with [18F] FDG PET/CT: a retrospective analysis of a single-center study. European Radiology. 2023; 33: 6492–6501.
- [56] Menzel C, Hamscho N, Döbert N, Grünwald F, Kovács AF, Wolter M, *et al.* PET imaging of Rosai-Dorfman disease: correlation with histopathology and ex-vivo beta-imaging. Archives of Dermatological Research. 2003; 295: 280–283.
- [57] McAllister BJ. Multi Modality Imaging Features of Cardiac Myxoma. Journal of Cardiovascular Imaging. 2020; 28: 235– 243.
- [58] Mankad R, Herrmann J. Cardiac tumors: echo assessment. Echo Research and Practice. 2016; 3: R65–R77.
- [59] Deodhare SS, Ang LC, Bilbao JM. Isolated intracranial involvement in Rosai-Dorfman disease: a report of two cases and review of the literature. Archives of Pathology & Laboratory Medicine. 1998; 122: 161–165.
- [60] Dalia S, Sagatys E, Sokol L, Kubal T. Rosai-Dorfman disease: tumor biology, clinical features, pathology, and treatment. Cancer Control. 2014; 21: 322–327.