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ORIGINAL ARTICLE

Extranodal Rosai-Dorfman disease in the breast: a literature review from 1969 to 2023

Enfermedad de Rosai-Dorfman extraganglionar en la mama: una revisión de la literatura de 1969 a 2023

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Abstract

Objective: *Reviewing available literature regarding extranodal Rosai–Dorfman disease in the breast to explore the clinical characteristics of this disease, the described therapeutic options, and their outcomes.* **Method:** *In January 2024, the PubMed, SpringerOpen, and Scopus databases were searched with the keywords "Rosai," "Dorfman," and "Breast." Forty-two studies were included in the final analysis, obtaining a total of 70 reported cases of extranodal Rosai–Dorfman disease affecting the breast. Patient characteristics, mammogram descriptions, therapeutic management, and outcomes were reviewed for statistical analysis.* **Results:** *The main population consisted of females in their sixth decade of life (93%), presenting with a firm, non-tender nodule (65.7%), generally localized to one breast (72%). About 18.6% of patients had nodal or extranodal disease in other areas. Excisional biopsy was the main treatment strategy (63%) and surgical excision showed a lesser association with recurrence than incisional biopsy (p = 0.049). Most instances of disease recurrence or progression were diagnosed within the first 2 years.* **Conclusions:** *This study revealed that surgical excision showed less association with disease recurrence or progression than expectant management. Follow-up can be conducted with a mammogram and physical examination since recurrence tends to occur locally within 2 years.*

Keywords: *Breast. Extranodal. Rosai–Dorfman disease. Management. Systematic review.*

Resumen

Objetivo: *Revisar la literatura disponible respecto a la enfermedad de Rosai-Dorfman en la mama para explorar sus características clínicas, las opciones terapéuticas descritas y sus desenlaces.* **Método:** *En enero de 2024 se realizó una revisión en las bases de datos PubMed, SpringerOpen y Scopus con las palabras clave "Rosai", "Dorfman" y "Breast". Se incluyeron en el análisis 42 estudios, obteniendo un total de 70 casos reportados de enfermedad de Rosai Dorfman extraganglionar en la mama. Las características de los pacientes, las mastografías, los manejos y los desenlaces se incluyeron en el análisis estadístico.* **Resultados:** *La población principal consistió en mujeres en la sexta década de la vida (93%), debutando con un nódulo firme, no doloroso (65.7%), generalmente localizado en una sola mama (72%). El 18.6% presentaron enfermedad en otros sitios. La biopsia escisional fue el principal manejo (63%), y la escisión quirúrgica mostró menor asociación con recurrencia que la biopsia incisional (p = 0.049). La mayoría de las recurrencias ocurrieron en los primeros 2 años.*

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Date of reception: 01-04-2024
Date of acceptance: 12-05-2024
DOI: 10.24875/CIRUE.M24000797

Cir Cir (Eng). 2024;92(6):725-734
Contents available at PubMed
www.cirugiaycirujanos.com

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Conclusiones: *Este estudio reveló que la escisión quirúrgica se asocia a menor recurrencia que el manejo expectante. El seguimiento debe mantenerse por al menos 2 años con un enfoque en las recurrencias locales.*

Palabras clave: *Extraganglionar. Mama. Enfermedad de Rosai-Dorfman. Manejo. Revisión sistemática.*

Introduction

Rosai–Dorfman disease (RDD), sometimes referred to as "Rosai–Dorfman–Destombes disease," represents a rare, idiopathic, and generally benign pathological entity which most commonly manifests as a massive, bilateral lymphadenopathy in the head and neck region of males between their second and third decade of life[1]. It was first described in 1959 by Pierre-Paul Louis Lucien Destombes but was not recognized as its own pathological entity until pathologists Juan Rosai and Ronald Dorfman's work in 1969[2,3]. Although initially described as a sinus histiocytosis with massive lymphadenopathy, it is currently classified as a type of non-Langerhans cell histiocytosis[4,5]. Less than half of afflicted patients present an extranodal variety of this disease, with some studies citing somewhere between 50 and 90 cases of sole breast affection[5].

Clinically, breast affection is most frequently reported as a firm non-tender nodule in one or both breasts. Confirmatory diagnosis is achieved through histopathological analysis, while core biopsy has been reported to be more reliable in comparison to fine-needle aspiration; excisional biopsy remains the mainstay for definitive diagnosis[6]. Finding proliferative histiocytes with emperipolesis is deemed to be pathognomonic of Rosai–Dorfman, with emperipolesis consisting of the presence of an intact cell within the cytoplasm of another cell[7]. The interiorized cell remains viable and is capable of exiting the other cell at any time without causing structural or functional compromise[8]. Another characteristic that guides diagnosis is its immunohistochemical profile which stains positive for S100 and CD68, and negative for CD1a[9]. While most cases have been associated with a benign prognosis, recurrence, and progression, both at a local and systemic level have been described and data on long-term follow-up is scarce[10]. Despite its low incidence, this pathology tends to mimic breast malignancy on clinical and radiological examination and with no current guidelines regarding a diagnostic and therapeutic gold standard, patients are often managed on a case by case basis[1,5,10].

The following systematic review aims to look at every case currently published in medical literature, gathering information on patient characteristics, presentation, management, and outcome in hopes of acting as a guide for future clinicians faced with this infrequent clinical entity.

Method

For the present systematic review, we selected all published original articles reporting cases of histologically confirmed extranodal Rosai–Dorfman disease with breast affection. We conducted a literature search in January 2024 using the PubMed, SpringerOpen, and Scopus databases with the keywords "Rosai–Dorfman" and "Breast." All articles including the established keywords published since 1969 were initially considered, with most of them corresponding to case reports and literature reviews. Inclusion criteria encompassed all articles detailing patients with a histologically confirmed diagnosis of Rosai–Dorfman in the breast with or without the involvement of other sites. Exclusion criteria included articles which were inaccessible, reports of Rosai–Dorfman outside of the breast, or cases that did not include at least four of the nine items detailed on our checklist. We initially identified 67 possible studies of which only 55 were assessed for eligibility. After a full review of each article, 42 were included in our review with eight being literature reviews and the rest being case reports and case series (Fig. 1). Among the articles included, clinical data were collected including demographic characteristics of patients, use of mammogram, BIRADS, management, length of follow-up, and recurrence. Whenever an article included data on patients previously reported by another author, only new patients were considered toward our final patient count, but any data missing from the previous reports were still compiled. Only articles including previously unreported patients among their series were included in the study. Statistical analysis was conducted with the Statistical Package for the Social Sciences version 25.0 statistical software. Findings were presented as mean ± standard deviation for numerical variables and frequency (%) for categorical

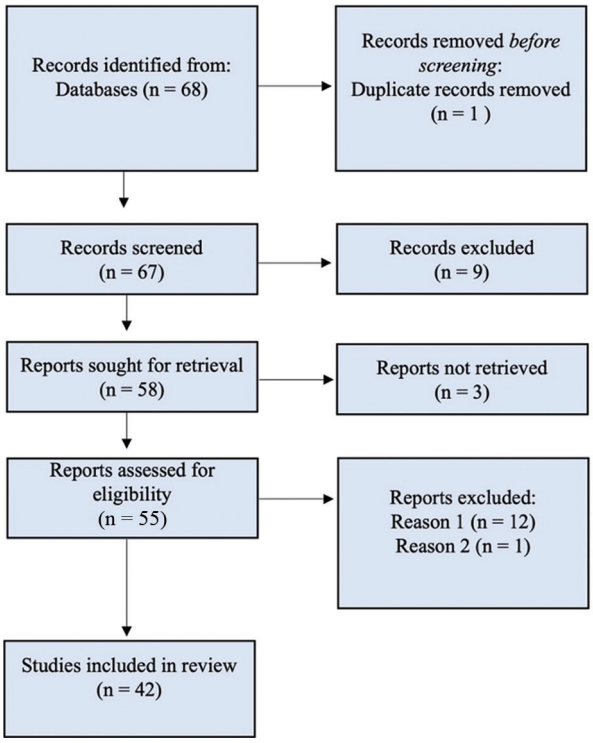

Figure 1. *Algorithm for article selection. Reason 1: Article did not describe a case of extranodal breast Rosai–Dorfman disease. Reason 2: Article described a case of extranodal breast Rosai–Dorfman disease but did not include enough data regarding the patient and management.*

variables. χ^2 and Fisher's exact test were utilized. Statistical significance was deemed as $p < 0.05$.

Results

After a careful review of each of the articles included in our study, we identified a total of 70 cases of extranodal Rosai–Dorfman disease (RDD) of the breast across 42 reports (Table 1). We observed a gender distribution of five males and 65 females (7% vs. 93%), with a generalized age range of 15-84 years and a mean age of 54 (± 15), implying a relatively mature demographic. On gender analysis, a mean age of 54 (± 15) and 44 (± 15) were found among females and males, respectively. Out of 68 patients in whom the initial clinical presentation was reported, the most frequently described scenario consisted of a palpable, non-tender nodule (n = 46, 65.7%). Twelve patients (17.1%) were asymptomatic and had incidental discoveries after a routine imaging study, 4 (5.7%) had bilateral nodules, 3 (4.3%) presented with multiple nodules in one breast, and 3 (4.3%) debuted with mastalgia. Data on clinical presentation were missing on 2 (2.9%) cases. A single breast was affected in 51 cases (72.9%), with bilateral affection in 9 (12.9%) reports. The right breast was most commonly affected (n = 31, 44.2%), while the remaining patients did not have laterality reported (n = 10, 14.2%). Most debuted with a localized breast affection (n = 56, 80%) versus a smaller proportion of patients who had some degree of nodal or extranodal disease in other areas of their bodies (n = 13, 18.6%).

While not all patients underwent radiologic studies, the one most frequently mentioned as part of the diagnostic workup was a mammogram (n = 50, 71.4%). Among these patients, the most commonly reported findings were poorly defined solid nodules classified as BIRADS 4 (n = 40, 80%). The remaining cases were classified as BIRADS 5 (n = 5, 10%), BIRADS 3 (n = 3, 6%), and BIRADS 0 (n = 2, 4%).

In regard to management, the most frequently reported procedure was an excisional biopsy (n = 44, 63%) followed by an incisional biopsy with subsequent expectant management (n = 13, 18.6%). Of the remaining patients, 6 (8.5%) underwent more extensive surgical procedures before diagnosis such as a partial mastectomy (n = 3, 4.3%), quadrantectomy (n = 2, 2.9%), and lumpectomy (n = 1, 1.4%). One of the reports combined an excisional biopsy with oral steroids, while another reported an incidental diagnosis made after a reduction mammoplasty. The remaining reports did not describe management (n = 6, 8.5%).

Follow-up was mentioned in 43 (61.4%) cases. The length of follow-up ranged from 2 months to 14 years, with an average of 20 months. Out of these patients, disease progression or recurrence was shown on 12 (17.1%), presenting as early as 2 months and up to 4 years after initial diagnosis, with an average of 16 months. Roughly half of these occurred locally in the affected breast, with the other half presenting mainly in the subcutaneous tissue of other sites of the body without a particular discernible pattern. Out of the included 70 cases, only one death (1.4%) was reported and directly attributed to disease progression.

On statistical analysis, gender did not appear to impact outcome when comparing recurrence between men and women ($p = 0.82$), debuting with bilateral disease was found to be associated with extranodal disease ($p = 0.007$). In regard to recurrence or progression, extramammary disease at diagnosis showed a stronger association in comparison to isolated

Table 1. Case reports reviewed and findings

Author	Age	Gender	Clinical presentation	Affected Breast	Extension	Mammogram/BIRADS*	Management	Outcome
Rosai and Dorfman[11] (1969)	25	F	-	Bilateral	Regional LN, CNS, soft-tissues, liver	-	Excisional biopsy	Persistent lymphadenopathy
	17	F	-	-	Axillary LN	-	Excisional biopsy	-
Pérez Guillermo et al.[12] (1993)	71	F	Nodule	Bilateral	EBA	LB: Solid well-defined mass (4) RB: Solid poorly defined mass (4)	LB: Excisional biopsy/ RB: Quadrantectomy	-
Hammond et al.[13] (1996)	67	F	Nodule	-	EBA	Solid poorly defined mass (4)	Excisional biopsy	Disease-free at 6 months
Govender and Chetty[14] (1997)	34	F	Nodule	-	EBA	-	-	-
Green et al.[15] (1997)	15	F	Bilateral nodules	Bilateral	Regional LN	(4)	Excisional biopsy	-
	84	F	Asymptomatic	Bilateral	Axillary LN	(4)	Excisional biopsy	Deceased 2 months after surgery due to systemic progression
	56	F	Bilateral nodules	Left	Mediastinal LN	(4)	Excisional biopsy	Disease-free at 5 years
	69	F	Nodule	Right	EBA	(4)	Excisional biopsy	-
	48	F	Nodule	Right	EBA	(4)	Excisional biopsy	Disease-free at 12 months
	54	F	Nodule	Right	EBA	(4)	Excisional biopsy	Disease-free at 12 months
	71	F	Nodule	-	EBA	(4)	Excisional biopsy	-
Soares et al.[16] (1999)	65	F	Multiple bilateral nodules	Bilateral	EBA	Multiple bilateral solid masses (4)	Excisional biopsy	-
Hummel et al.[17] (1999)	52	F	Nodule	Left	EBA	Poorly defined mass (5)	-	-
Ng et al.[18] (2000)	61	F	Nodule	Right	EBA	Not performed	Excisional biopsy	Disease-free at 6 months
	40	F	Mastalgia	Right	EBA	Poorly defined mass (4)	Excisional biopsy	Disease-free at 1 month
Kuzmiak et al.[19] (2003)	30	F	Nodule	Right	EBA	Poorly defined mass (4)	Excisional biopsy	-
Pham et al.[20] (2005)	53	F	Nodule	Left	EBA	Poorly defined mass (4)	Incisional biopsy	Disease-free at publication
Da Silva et al.[21] (2007)	50	F	Nodule	Left	EBA	Poorly defined mass (4)	Excisional biopsy	Disease-free at 5 years
Perera et al.[22] (2007)	23	M	Nodule	Left	EBA	Poorly defined mass (5)	Excisional biopsy	Disease-free at 3 months
Dahlgren et al.[23] (2008)	64	F	Asymptomatic	Bilateral	Parotid and cervical LN	- -	Excisional biopsy	-

(Continues)

Table 1. Case reports reviewed and findings *(continued)*

Author	Age	Gender	Clinical presentation	Affected Breast	Extension	Mammogram/BIRADS*	Management	Outcome
Franco-Odio et al.[24] (2009)	59	F	Nodule	Right	EBA	Poorly defined mass (5)	Excisional biopsy	-
Morkowski et al.[25] (2010)	53	F	Asymptomatic	Left	EBA	Irregular dense mass (4)	Excisional biopsy	Initially presented with a negative incisional biopsy, progressed locally after 4 years, underwent excision, and remains disease-free at 5 years and 10 months
	45	F	Asymptomatic	Left	EBA	Poorly defined mass (4)	Excisional biopsy	Disease-free at 14 months
	54	F	Asymptomatic	Right	EBA	Irregular mass (4)	Excisional biopsy	Disease-free at 4 years
Wu et al.[26] (2010)	33	F	Nodule	Right	EBA	-	Lumpectomy	Disease-free at 2 years
Bansal et al.[27] (2010)	35	M	Nodule	Right	EBA	-	Incisional biopsy	Disease-free at 18 months
Picón-Coronel et al.[28] (2010)	67	F	Nodule	Right	EBA	Poorly defined mass (4)	Excisional biopsy	-
Noordzij et al.[29] (2011)	75	F	Nodule	Right	EBA	Multiple solid masses	Excisional biopsy	-
Gwin et al.[30] (2011)	68	F	Asymptomatic	Bilateral	EBA	Multiple small bilateral masses (4)	Incisional biopsy	-
Tenny et al.[31] (2011)	64	F	Nodule	Right	Axillary LN	Poorly defined mass (4)	Incisional biopsy	Local recurrence at 6 months, contralateral breast mass at 7 months, left flank SCT mass at 11 months (after initial biopsy)
Fu et al.[32] (2012)	78	F	Nodule	Right	EBA	-	Excisional biopsy	-
Cha et al.[33] (2012)	62	F	Nodule	Right	EBA	Poorly defined mass (3)	Incisional biopsy	Disease-free at 10 months
Baladandapani et al.[34] (2012)	59	M	Multiple unilateral nodules	Left	EBA	Poorly defined lobulated mass (5)	Excisional biopsy	-
Parkin et al.[35] (2015)	56	F	Asymptomatic	Left	EBA	Solid well-defined mass (4)	Incisional biopsy	Disease-free at 11 months
Ibáñez et al.[36] (2015)	69	F	Nodule	Right	EBA	Poorly defined mass (4)	Partial mastectomy	Disease-free at 3 years
Mantilla et al.[37] (2016)	59	F	Mastalgia	-	EBA	Non-definitive findings (0)	Incisional biopsy	New nodule in SCT of left thigh at 3 years of follow-up
	67	F	Nodule	-	EBA	-	-	-
Simmons et al.[38] (2016)	41	F	Nodule	Right	EBA	Heterogeneous lobulated mass (4)	Excisional biopsy	Disease-free at 9 months

(Continues)

Table 1. Case reports reviewed and findings *(continued)*

Author	Age	Gender	Clinical presentation	Affected Breast	Extension	Mammogram/BIRADS*	Management	Outcome
Jorns[39] (2016)	-	F	Asymptomatic	-	EBA	Poorly defined mass (4)	Incisional biopsy	Disease-free at publication
Zhou[40] (2016)	71	F	Multiple unilateral nodules	Left	EBA	Poorly defined mass (4)	Excisional biopsy	-
El-Attrache et al.[2] (2017)	55	M	Nodule	Right	EBA	Poorly defined mass (4)	Excisional biopsy	-
Delaney et al.[41] (2017)	63	F	Nodule	Right	EBA	Poorly defined mass (4)	Incisional biopsy	New breast nodule at 7 months of follow-up, no further progression at publication
De Mello et al.[42] (2018)	55	F	Nodule	Left	EBA	Irregular poorly defined nodule (4)	Incisional biopsy	Development of 2 new nodules at 2 months of follow-up (SCT of leg and SCT of biopsy site), underwent excision of all three sites afterward
Araújo et al.[43] (2018)	46	F	Nodule	Left	EBA	Solid nodule (0)	Quadrantectomy	Disease-free at 4 years
Liu et al.[44] (2018)	65	F	Nodule	Right	EBA	- -	Excisional biopsy	Disease-free at 40 months
	78	F	Nodule	Right	EBA	- -	Excisional biopsy	Disease-free at 39 months
	53	F	Nodule	Left	EBA	- -	Excisional biopsy	Disease-free at 13 years
	45	F	Nodule	Left	EBA	- -	Excisional biopsy	Disease-free at 7 years
Goldbach et al.[45] (2018)	44	F	Nodule	Right	SCT of abdomen	Poorly defined mass (4)	Excisional biopsy	Disease-free at publication
Shin et al.[46] (2019)	54	F	Mastalgia	Right	EBA	- -	Excisional biopsy	Local recurrence at 6 months post-surgery
Dai et al.[47] (2019)	42	F	Asymptomatic	Left	EBA	Poorly defined mass (4)	Excisional biopsy	Disease-free at 3 years
Shetty et al.[48] (2020)	52	F	Asymptomatic	Right	EBA	Macromastia (3)	Incidental finding after reduction mammoplasty	Disease-free at 13 months
	54	F	Nodule	Right	EBA	(3)	Partial mastectomy	Disease-free at 14 months
	58	F	Nodule	Right	EBA	- -	Excisional biopsy	-
	63	F	Nodule	Right	Axillary LN	Poorly defined complex cystic masses (4)	Partial mastectomy	Systemic recurrence at 12 months post-surgery

(Continues)

Table 1. Case reports reviewed and findings *(continued)*

Author	Age	Gender	Clinical presentation	Affected Breast	Extension	Mammogram/BIRADS*	Management	Outcome
Elshikh et al.[49] (2020)	64	F	Nodule	-	SCT of breast	- -	-	-
	65	F	Nodule	-	EBA	- -	-	-
	50	F	Nodule	-	SCT of breast and parotid	Solid mass (4)	-	-
Vaidya et al.[50] (2020)	43	F	Nodule	Left	EBA	Spiculated solid mass (5)	Excisional biopsy	-
Battle et al.[51] (2020)	49	F	Multiple unilateral nodules	Right	EBA	Poorly defined mass (4)	Incisional biopsy	Remitting and recurring plaques and papules in affected breast during the 12 months of follow-up
Reddy et al.[8] (2021)	36	F	Nodule	Left	EBA	Spiculated mass (4)	Excisional biopsy	Disease-free at 2 years
	58	F	Asymptomatic	Left	Axillary, cervical and pectoral LN, humerus, sternum, bilateral clavicle, D4 vertebrae, bilateral femur, multiple bilateral pulmonary nodules	Poorly defined mass (4)	Excisional biopsy + Oral steroids	Disease-free at 11 months
Choo et al.[52] (2021)	52	F	Nodule	Left	EBA	Poorly defined mass (4)	Excisional biopsy	Disease-free at 4 years
	37	F	Nodule	Left	EBA	Poorly defined mass (4)	Incisional biopsy	-
Maldonado Marcos et al.[9] (2021)	50	M	Nodule	Right	SCT of breast	- -	Excisional biopsy	Disease-free at 2 years
Iancu et al.[5] (2021)	63	F	Multiple bilateral nodules	Bilateral	EBA	Poorly defined mass (4)	Incisional biopsy	Nodule involution at 6 months of follow-up, disease-free at 12 months
Ingole et al.[10] (2022)	21	F	Nodule	Right	EBA	- -	Excisional biopsy	-
Sumner et al.[53] (2023)	59	F	Asymptomatic	Bilateral	EBA	Solid round mass in RB (4), focal asymmetry in medial LB (4)	Excisional biopsy	-
Díaz-Degollado et al.[1] (2023)	67	F	Nodule	Right	EBA	Poorly defined mass (4)	Excisional biopsy	Disease-free at 5 years

*BIRADS score is described in parentheses; mammogram findings are included when described in their respective reports.
LN: lymph node; CNS: central nervous system; EBA: exclusive breast affection; SCT: subcutaneous tissue; LB: left breast; RB: right breast.

Table 2. Recurrence or progression comparison among patients with exclusive breast affection versus extramammary disease

Disease extension at diagnosis	Outcome		Total (%)
	Recurrence or progression (%)	Disease-free (%)	
Exclusive breast affection	58 (7)	90 (28)	81 (35)
Extramammary disease	42 (5)	10 (3)	19 (8)

p = 0.015612.

Table 3. Recurrence or progression comparison among patients with unilateral versus bilateral breast affection

Disease extension at diagnosis	Outcome		Total (%)
	Recurrence or progression (%)	Disease-free (%)	
Unilateral disease	82 (9)	86 (42)	85 (51)
Bilateral disease	18 (2)	14 (7)	15 (9)

p = 0.743641.

breast affection (p = 0.015) (Table 2), but bilateral disease in comparison to unilateral disease did not seem to show such an association (p = 0.74) (Table 3). BIRADS score did not seem to affect outcome when comparing BIRADS 4 versus other scores (p = 0.63). When comparing management strategies, incisional biopsy with subsequent observation seemed to show a stronger association with recurrence than excisional biopsy, but the analysis did not show statistical significance (p = 0.06) (Table 4); nonetheless, when adjusting for all cases of surgical management against incisional biopsy, a significant association was found (p = 0.049) (Table 5).

Table 4. Recurrence or progression comparison among patients with excisional versus incisional biopsy

Disease extension at diagnosis	Outcome		Total (%)
	Recurrence or progression (%)	Disease-free (%)	
Excisional biopsy	44 (4)	78 (21)	69 (25)
Incisional biopsy	56 (5)	22 (6)	31 (11)

p = 0.060103.

Table 5. Recurrence or progression comparison among patients with surgical versus expectant management

Disease extension at diagnosis	Outcome		Total (%)
	Recurrence or progression (%)	Disease-free (%)	
Surgical management	50 (5)	81 (26)	74 (31)
Incisional biopsy	50 (5)	19 (6)	26 (11)

p = 0.060103.

Discussion

Rosai–Dorfman–Destombes disease (RDD) is a rare, proliferative, and inflammatory idiopathic disorder with a pathogenesis that remains unknown. Breast affection alone is particularly rare, with most reviews citing a prevalence of < 1% of cases[1,2,5,41,44,53]. Most reports consist of women over 50 years of age who present with an asymptomatic non-tender, firm, and palpable nodule which on radiologic examination presents as a poorly defined rounded mass in mammograms and a hypoechoic mass in ultrasound[48]. Due to the affected demography and its imaging characteristics, this presentations tends to elicit a high suspicion of breast malignancy and start the patient on a diagnostic workup which may ultimately guide them to the operating room[1,5,10].

Our review showed demographic findings consistent with those previously described in other articles with a predominance of females in their sixth decade of life. Affected males seemed to debut an average of 10 years before their female counterparts, but no significant impact in outcome was found on statistical analysis. While a variety of clinical presentations were reported, the most common one was a single non-tender, firm, palpable nodule affecting a single breast, a finding previously reported in multiple literature reviews[5,41]. The second most common presentation was an asymptomatic nodule detected through routine screening. In regards to such radiologic studies, the one most frequently mentioned was a mammogram reporting findings deemed as BIRADS 4, but no significant association was found between reported BIRADS score and outcomes in our analysis.

While most patients presented with purely extranodal breast disease without systemic compromise, almost a fifth had some degree of non-breast affection which ranged from local and regional lymph nodes to distant organs and subcutaneous tissue. Cervical and axillary nodes were the most commonly reported sites. Our analysis found an association between those patients who debuted with extramammary

disease and those who presented recurrence during follow-up. When conducting the same analysis on unilateral versus bilateral disease no such association was found, nonetheless, bilateral disease was found to be associated with extramammary disease, therefore indirectly granting a slightly higher risk of disease recurrence.

Regarding management, excisional biopsies were found to be the most frequently described procedure both in previous reviews and in ours[2,44]. The second most frequently reported management option was vigilance after diagnostic confirmation with an incisional biopsy. Most reports of nodal and extranodal RDD describe benign outcomes without intervention, with only a small percentage of patients presenting further progression[5,41]. Our review found that among patients with breast RDD when comparing excisional and incisional biopsies with the available data, no significant association was found with disease progression or recurrence. Nevertheless, when adjusting the parameters to include all patients who received a surgical intervention (excisional biopsy, mastectomy, quadrantectomy, lumpectomy, and a patient who underwent a breast reduction), the statistical analysis produced a significant association which may show a slight decrease in recurrence among those who received some kind of surgical excision.

When dealing with recurrence or progression, half of these presented locally in the affected breast, with the remaining half manifesting either regionally or distantly, such as two patients whose recurrence appeared in the subcutaneous tissue of their leg, and another in whom the new lesion presented in the left flank. These findings show that while follow-up examinations must include the breast, they cannot be restricted to it and should include a general assessment as well.

While lacking, available data on follow-up suggest that disease progression or recurrence tends to take place within the first 2 years and rarely before 6 months. While more data are required to establish an adequate protocol for outpatient observation, a yearly mammogram and physical examination seem to be an adequate option. Mortality is rarely reported as a result of breast RDD in the literature, being mentioned only in 1 of the cases found on review.[11]

Conclusion

Rosai–Dorfman disease is a very infrequent pathology, one with an exquisitely rare disposition to present as a solitary breast nodule. Such presentation steps outside the borders of the population that we normally associate with RDD and seems to favor both a different gender and age group. Among these patients, the clinical presentation of extranodal breast RDD tends to mimic malignancy, making differential diagnosis difficult for those unfamiliar with this pathology. Due to its low incidence, guidelines for management do not currently exist, and more evidence is required to generate appropriate ones; in spite of this, our review lead us to find important aspects of this disease's behavior that we have chosen to sum up in the following points: (1) Imaging studies tend to present findings that can be most frequently reported as BIRADS4. (2) Diagnosis can only be obtained through histopathological analysis of a biopsy sample and requires a strong clinical suspicion. (3) Surgical excision, by excisional biopsy or a larger procedure, seemed to show less association with disease recurrence or progression when compared with expectant management. (4) Excisional biopsy seems to be associated with the same outcomes as more extensive surgical procedures. (5) Follow-up is important as almost a fifth of patients showed some sign of disease progression or recurrence. (6) Breast imaging is an important part of patient monitorization, but not the sole foundation of it since only half of the recorded cases of progression or recurrence occurred locally. (7) Follow-up should be maintained for at least 2 years since most instances of recurrence or progression were detected within the first 16 months.

Funding

The authors declare that they have not received funding.

Conflicts of interest

The authors declare no conflicts of interest.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that no patient data appear in this article. Furthermore, they have acknowledged and followed the recommendations as per the SAGER guidelines depending on the type and nature of the study.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript nor for the creation of images, graphics, tables, or their corresponding captions.

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