Egyptian Journal of Radiology and Nuclear Medicine

CASE REPORT Open Access

The false alarm: Rosai–Dorfman disease of breast: case report

Sanjivanee Ingole, V. Pavithra, Sandhya Sundaram, Leena Dennis Joseph, Syed Altaf Hussain and Bhawna Dev*

Abstract

Introduction: Rosai–Dorfman disease is a rare disorder caused by benign proliferation of histiocytes and thus also called sinus histiocytosis with massive lymphadenopathy. It is typically known to involve lymph nodes but can also affect extra nodal sites with breast involvement being extremely rare. This disease is known to mimic malignancy clinically and on imaging. The final verdict is through histopathological evaluation.

Case presentation: We present a case of a 21-year-old female who presented with a palpable suspicious lump in right breast. Ultrasound, mammogram and MRI breast were done followed by fine needle aspiration, which showed lymphocytes and occasional histiocytes, which is suggestive of a lymphoid process. Surgical excision was done due to high suspicion of low-grade angiosarcoma in spite of a negative cytology. Diagnosis of Rosai–Dorfman disease was concluded after histopathological evaluation and was further confirmed by immunohistochemical staining by immunohistochemical markers S-100 and CD68.

Conclusion: Rosai–Dorfman disease can present as low-grade angiosarcoma clinically as well as on imaging. There is a need for high level of suspicion to rule out benign mimics of malignancy to avoid radical surgery. Final diagnosis is by histopathology along with immunohistochemical staining.

Keywords: Rosai–Dorfman disease, Sinus histiocytosis, Breast, Benign mimic of malignancy

Introduction

Rosai–Dorfman disease is a rare disorder caused by benign proliferation of histiocytes and thus also called sinus histiocytosis with massive lymphadenopathy [1]. It is typically known to involve lymph nodes but can also involve extra nodal sites in up to 23% patients [2].

Breast involvement is extremely rare with only 17 cases reported in English literature [1]. Most cases have been discovered on screening mammograms [3, 4]. Rosai–Dorfman disease of breast mimics malignancy on mammogram and ultrasound [3, 4], thus leading to surgical excision in most cases [5]. As in this case, in spite of a diagnosis of benign lymphoid process on fine-needle aspiration, wide local excision was done on clinical and radiological suspicion of malignancy.

Case presentation

A 21-year-old female came with complaints of lump in right breast for 2 years with recent progressive increase in size. No history of trauma or lactation was present. No family history of breast cancer was noted. On examination, palpable irregular superficial lump in upper outer quadrant with associated bluish red discoloration of skin and ecchymosis (Fig. 1a) was noted.

Patient was advised breast ultrasound and referred to a breast surgeon.

Ultrasound of the lump showed a non-circumscribed irregular heteroechoic lesion in upper outer quadrant at 10–11 o'clock in subcutaneous plane (Fig. 2a). This lesion was seen abutting the skin with overlying significant skin thickening. Doppler ultrasound showed avid

*Correspondence: bhawnadev@gmail.com
Sri Ramachandra Institute of Higher Education and Research (Deemed To Be University), Sri Ramachandra Nagar, Porur, Chennai, Tamil Nadu 600116, India

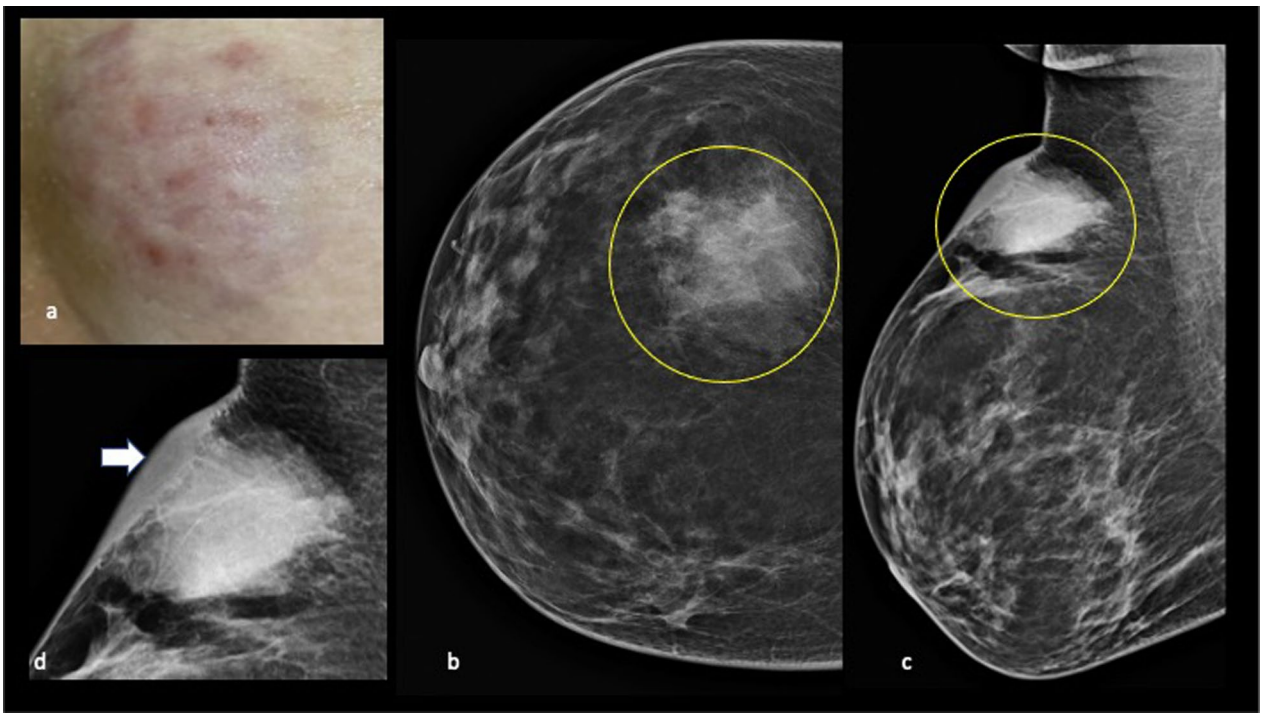

Fig. 1 **a** The clinical image of the lesion shows a superficial lump with bluish red discoloration. **b** Right craniocaudal view and **c** right mediolateral view show a non-circumscribed irregular high-density lesion (yellow circle) with associated overlaying skin thickening (white arrow). **d** Zoomed image of the lesion shows skin thickening overlaying the lesion

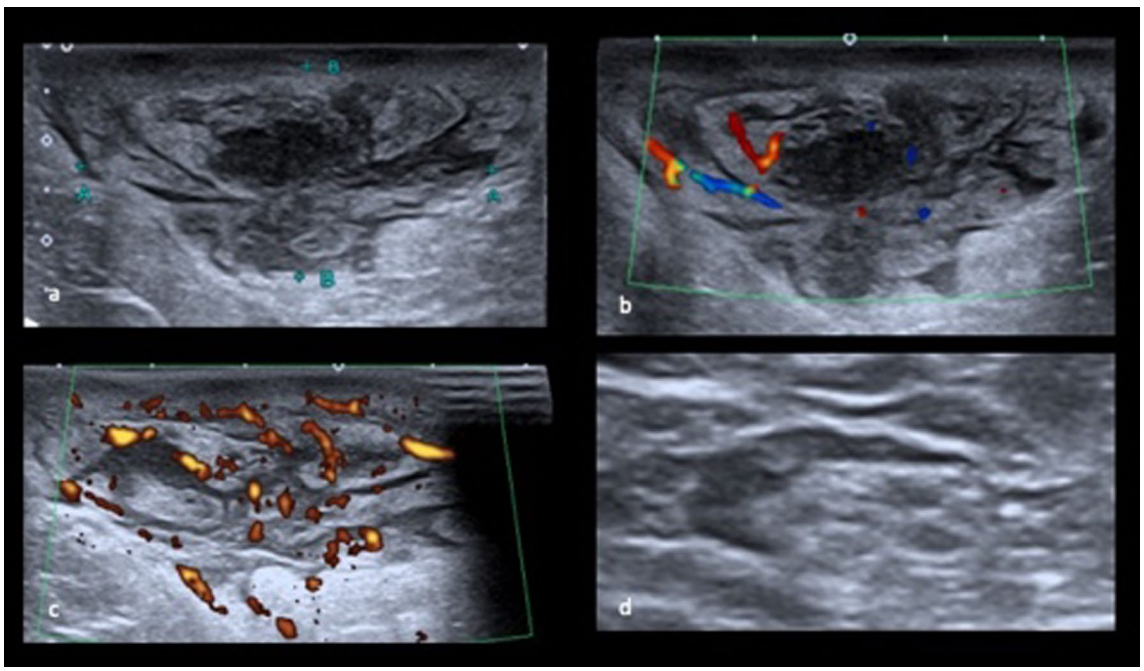

Fig. 2 **a** Grey scale ultrasound of the of the breast shows a non-circumscribed irregular heteroechoic lesion in subcutaneous plane. The lesion closely abuts the skin with overlying significant skin thickening. **b**, **c** Color flow and Doppler ultrasound show avid internal vascularity. **d** Right axilla ipsilateral axillary lymph nodes with preserved fatty hilum

internal vascularity with feeding vessel traced from a branch of internal mammary artery (Fig. 2b,c). Right axilla showed few lymph nodes with preserved fatty hilum (Fig. 2d).

On the suspicion of malignancy digital mammography of the right breast was done. Right craniocaudal view (Fig. 1b) and right mediolateral view (Fig. 1c) showed a non-circumscribed irregular high-density lesion with associated overlying skin thickening (Fig. 1d) (white arrow). This was followed with MRI that showed a circumscribed irregular T1 hypointense (Fig. 3a), T2/STIR hyperintense (Fig. 3b) lesion in upper outer quadrant at 10–11 o'clock position, closely abutting the skin (white arrows) with associated overlaying skin thickening in right breast. Lesion showed DWI restriction (Fig. 3c). On maximum-intensity projection images (Fig. 3d), large feeding vessels arising (white arrowhead) from internal mammary artery (white arrow) and lateral thoracic artery were seen. On T1C, the lesion showed rapid contrast uptake which progresses to intense heterogeneous enhancement (Fig. 3e–h). Considering the above characteristics, the possibility of low-grade angiosarcoma was considered more likely with hemangioma as other possibility.

Due to highly vascular nature of the lesion, fine-needle aspiration of the lesion was done, which showed lymphocytes and occasional histiocytes, suggestive of lymphoid process (Fig. 4a,b).As were equivocal findings and high clinical as well as radiological suspicion, surgical wide local excision (Fig. 4c) was performed and sent for histopathological analysis.

On gross examination, the specimen revealed a firm, ill-circumscribed grey tan lesion (Fig. 4d). Histologically, the lesion had characteristic accumulation of histiocytes with enlarged round to oval hyperchromatic nuclei and abundant cytoplasm (Fig. 5a,b). Few histiocytes displayed a phenomenon of emperipolesis, which was seen engulfing the inflammatory cells and it is characteristic of the said disease (Fig. 5b). On immunohistochemistry, these histiocytes showed positivity for S-100 (Fig. 5c) and CD 68 (Fig. 5d) confirming the diagnosis of Rosai–Dorfman disease.

Final diagnosis was reached only after the characteristic histopathological and immunohistochemical findings.

Wide local excision was done on suspicion of malignancy clinically and on imaging. Whole body examination along with abdominal, neck and bilateral axillary ultrasound was performed. It did not reveal any other

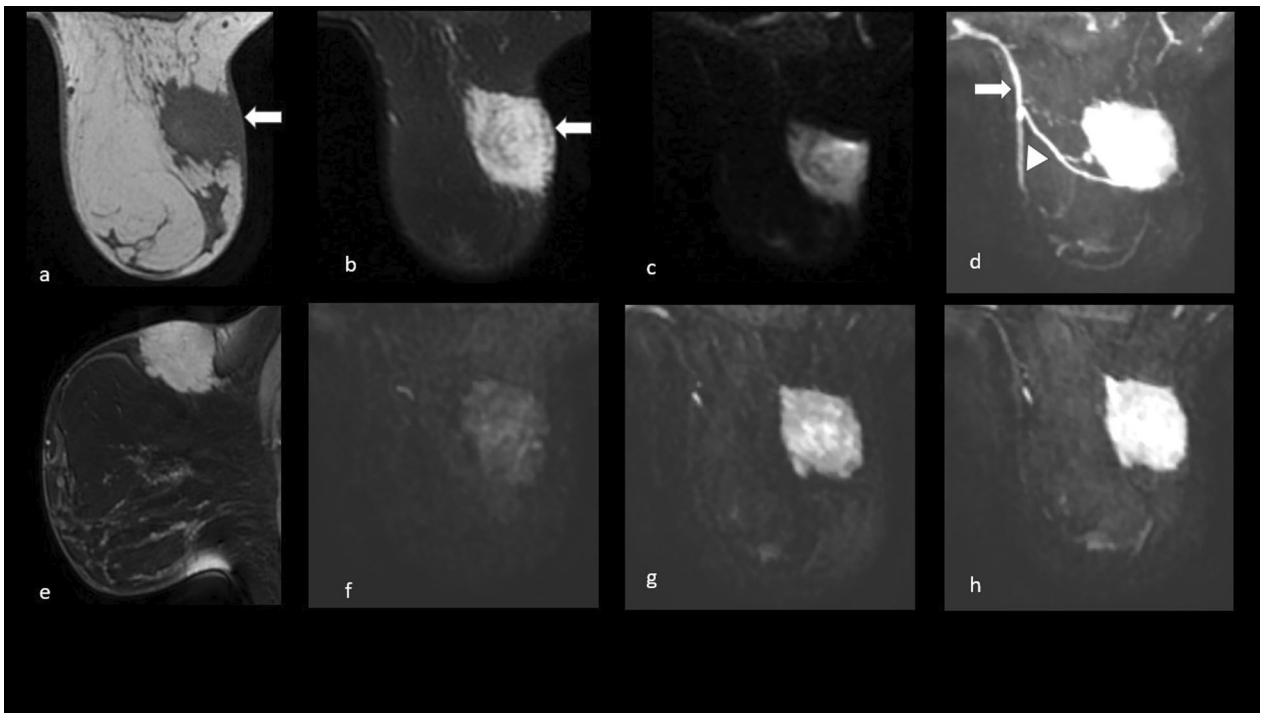

Fig. 3 MRI of breast showed **a** a circumscribed irregular T1 hypointense, **b** T2/STIR hyperintense lesion in upper outer quadrant at 10–11 o'clock position, closely abutting the skin (white arrows) with associated overlaying skin thickening. **c**Lesion shows DWI restriction. **d** On maximum-intensity projection images, large feeding vessels arise (white arrow head) from internal mammary artery (white arrow). **e** T1 sagittal contrast shows homogenously enhancing lesion in the upper quadrant. **f** T1 contrast early phase shows contrast uptake. **g**, **h** Enhancement progresses to intense heterogeneous enhancement in later phases

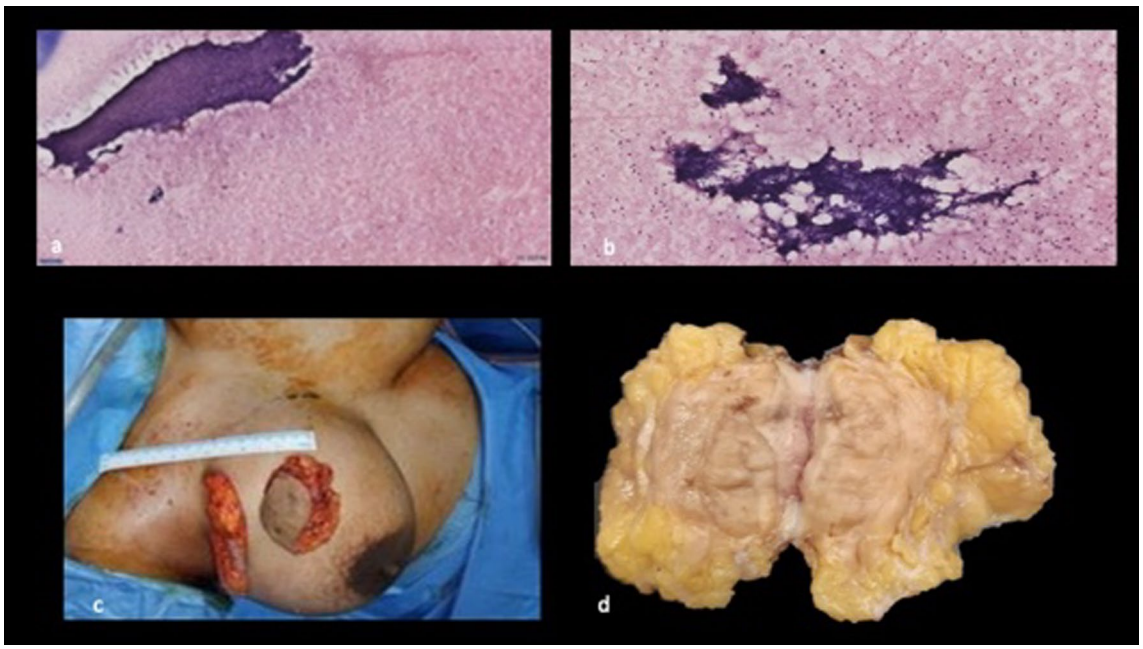

Fig. 4 **a** Cytology image showing fibrocollagenous tissue in a background of lymphocytes (H&E ×200). **b** Cytology image showing reactive lymphocytes and histiocytes in the background (H&E ×400). **c** Intraoperative picture shows a linear incision in the lateral mammary fold and excised specimen placed on the breast. **d** On gross examination, the specimen was firm, ill-circumscribed grey tan lesion

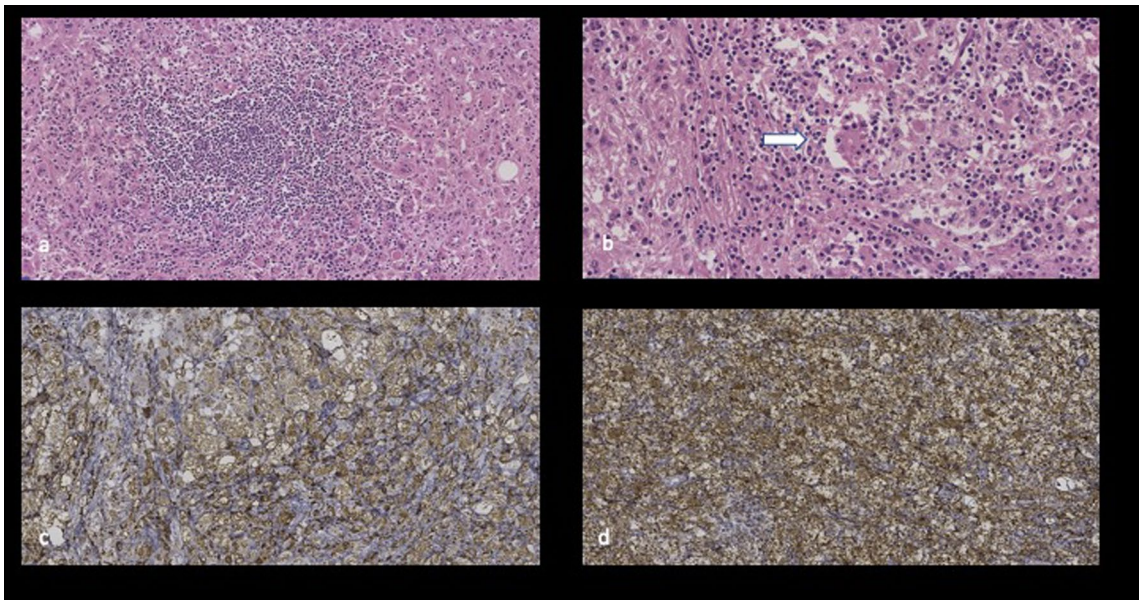

Fig. 5 **a** Histiocytes with enlarged, round-to-oval hypochromatic nuclei and abundant eosinophilic cytoplasm admixed with lymphocytes and a prominent lymphoid collection (H&E ×400). **b** Histiocytes engulfing lymphocytes and red blood cells (emperipolesis) (H&E ×400). **c** CD 68 positivity in the cytoplasm of the histiocytes (IHC ×400). **d** S100 positivity in the cytoplasm of the histiocytes (IHC ×400)

similar lesions. Computed tomography was avoided in view of excessive radiation and young age of the patient.

Patient is advised to be kept on close yearly USG breast, abdomen and neck along with 2 yearly CT surveillance in view of young age of the patient.

Discussion

Rosai–Dorfman disease is a rare disorder caused by benign proliferation of histiocytes and thus also called sinus histiocytosis with massive lymphadenopathy [1]. It is typically known to involve lymph nodes but can also involve extra nodal sites in up to 23% patients, with breast involvement being extremely rare [2].

Most patients present as benign lump in breast, but many cases have been diagnosed in screening mammograms in asymptomatic women [2–4]. Radiological features are variable. It may present as a benign mass or fat necrosis and is known mimic malignancy on ultrasound and mammogram [2, 6]. Some cases may have skin thickening and synchronous or metachronous axillary lymphadenopathy [2].

The final diagnosis is through histopathological evaluation with the hallmark finding of emperipolesis along with S100 immunohistochemical staining that gives a definite diagnosis of Rosai–Dorfman disease. In addition, CD68 immunohistochemical staining confirms the inflammatory nature of the process involving macrophages and monocytes [6, 7]. In our case, the lesion had characteristic accumulation of histiocytes with few displaying emperipolesis with immunohistochemistry showing positivity for S-100 (Fig. 4e) and CD 68.

The disease usually spontaneously resolves and follows a benign course. In few cases, the disease may be progressive with solid organ involvement. These cases carry a bad prognosis especially with the involvement of kidney, liver and lower respiratory tract. In extremely rare cases, Rosai–Dorfman disease can be fatal [2, 8].

Till date no studies could be found to determine the best imaging modality to follow up Rosai–Dorfman disease outside breast. Some studies have shown 18F-fluorodeoxyglucose-positron emission tomography/CT for diagnosis, staging and follow-up [9, 10].

Considering the clinical presentation of a breast lump with skin discoloration and the imaging findings of a superficial, non-circumscribed, highly vascular lesion possibilities of low-grade primary angiosarcoma and hemangioma were considered among the differentials.

Primary angiosarcoma of breast is seen in young females typically in 3rd or 4th decade of life. It presents as a rapidly growing mass with bluish discoloration. Mammography shows non-circumscribed lesions which are heterogeneous on ultrasound with high vascularity on color Doppler. On MRI, they appear T2 hyperintense and T1 hypointense with high-intensity lakes of hemorrhage. Progressive enhancement is seen in low-grade tumors [11].

Breast hemangiomas are superficial subcutaneous or subdermal lesions which appear nonspecific circumscribed oval or lobular on mammography. On ultrasound, large lesions are heterogeneous and highly vascular with serpiginous vessels, often displaying a draining vessel. Limited literature on MRI appearance has shown benign morphological features with avid early enhancement [11, 12].

Conclusion

The present case illustrates that Rosai–Dorfman disease can present as low-grade angiosarcoma clinically as well as on imaging thus a need for high level of suspicion to rule out benign mimics of malignancy to avoid radical surgery is necessary. However, final diagnosis is by histopathology along with immunohistochemical staining. Close surveillance should be maintained as the disease can be progressive and rarely fatal.

Abbreviations

CD 68: Cluster of differentiation 68; CT: Computed tomography; MR: Magnetic resonance Imaging; RDS: Rosai–Dorfman syndrome; USG: Ultrasonography.

Acknowledgements

Not applicable.

Authors' contributions

BD performed conceptualization and validation. SI did data curation. SI, VP, SS, and LD investigated the study. BD done project administration. BD and SAH provided resources. BD and LD supervised the study. SI and VP did writing—original draft. BD and LDJ were involved in writing—review and editing. All authors read and approved the final manuscript.

Funding

This research received no specific grant from any funding agency in public, commercial or nonprofit sectors.

Availability of data and materials

Not Applicable.

Declarations

Ethics approval and consent to participate

Ethical approval was obtained from Institutional Ethical Research committee.

Consent for publication

This is to certify that I Miss Samarpana S, after having undergone treatment at Sri Ramachandra Medical Centre, give consent to use my clinical, surgical and radiological images for academic purpose to be published in a medical journal. I understand that the images will be used for purely academic purpose. I have been assured by my consultant Doctor that my identity as regards name, face, date of birth, residence and any other personal details will be kept anonymous.

Competing interests

The authors whose names are listed immediately below certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or

other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

Received: 24 January 2022 Accepted: 20 February 2022
Published online: 05 April 2022

References

1. Morkowski JJ, Nguyen CV, Lin P, Farr M, Abraham SC, Gilcrease MZ et al (2010) Rosai-Dorfman disease confined to the breast. Ann Diagn Pathol 14(2):81–87
2. Delaney EE, Larkin A, MacMaster S, Sakhdari A, DeBenedectis CM (2017) Rosai–Dorfman disease of the breast. Cureus 9(4):e1153
3. Azari-Yaam A, Abdolsalehi MR, Vasei M, Safavi M, Mehdizadeh M (2020) Rosai–Dorfman disease: a rare clinicopathological presentation and review of the literature. Head Neck Pathol 15(1):352–360
4. Ng SB, Tan LH, Tan PH (2000) Rosai-Dorfman disease of the breast: a mimic of breast malignancy. Pathology (Phila) 32(1):10–15
5. Tenny SO, McGinness M, Zhang D, Damjanov I, Fan F (2011) Rosai–Dorfman disease presenting as a breast mass and enlarged axillary lymph node mimicking malignancy: a case report and review of the literature. Breast J 17(5):516–520
6. Ding D, Qi C, Nasreen AV, Jan W, Zsuzsanna PT, Karlene H-L et al (2019) Rosai–Dorfman disease presenting as a breast mass. Arch Pathol Clin Res 3(1):008–014
7. Chistiakov DA, Killingsworth MC, Myasoedova VA, Orekhov AN, Bobryshev YV (2017) CD68/macrosialin: not just a histochemical marker. Lab Investig J Tech Methods Pathol 97(1):4–13
8. Foucar E, Rosai J, Dorfman R (1990) Sinus histiocytosis with massive lymphadenopathy (Rosai–Dorfman disease): review of the entity. Semin Diagn Pathol 7(1):19–73
9. Albano D, Bosio G, Bertagna F (2015) 18F-FDG PET/CT follow-up of Rosai–Dorfman disease. Clin Nucl Med 40(8):e420-422
10. Fu L, Liu M, Song Z, Xu B, Tian J (2012) 18F-fluoro-deoxyglucose positron emission tomography/computed tomography scan findings in Rosai–Dorfman disease with IgG4-positive plasma cell infiltration mimicking breast malignancy: a case report and literature review. J Med Case Rep 6(1):411
11. Ameen R, Mandalia U, Marr AA, McKensie P (2012) Breast hemangioma: MR appearance with histopathological correlation. J Clin Imaging Sci 2:53
12. Mesurolle B, Sygal V, Lalonde L, Lisbona A, Dufresne M-P, Gagnon JH et al (2008) Sonographic and mammographic appearances of breast hemangioma. AJR Am J Roentgenol 191(1):W17-22

Publisher's Note