A rarity in breast pathology: A male case of Rosai-Dorfman disease and literature review

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A B S T R A C T

Rosai-Dorfman Disease (RDD) is a rare disease that normally presents with bilateral cervical lymph node enlargement. Systemic symptoms of fever and weight loss may be present but patients are usually asymptomatic. This benign disease tends to regress on its own without treatment but there have been cases that required treatment with steroids or chemotherapeutic regimes. Extranodal disease in the breast is extremely rare, with only three cases identified in the male breast. The patient in this study presented with an asymptomatic right breast lump identified incidentally. After excisional biopsy, a diagnosis of RDD was confirmed on immunochemistry and histopathology.

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1. Introduction

Rosai-Dorfman disease was first described in 1959 by Destombes as a lipid storage disorder that develops after inflammation. In 1969, Rosai and Dorfman had discovered the clinical symptoms of painless, enlarged, bilateral cervical lymphadenopathy to be associated with the pathological features of lymphoproliferation and accumulation of histiocytes [1]. This disease process was subsequently named Sinus Histiocytosis with Massive Lymphadenopathy (SHML). After a thirty case analysis of this disease in 1972, SHML was recognized as a distinct clinicopathologic process. Now known as RDD, it was found to extend beyond cervical lymph nodes in other lymphatic regions and extranodal sites. The axillary, inguinal, and mediastinal lymph nodes can present with disease. Extranodal disease presents 25–40% of the time in different areas such as: the bone, upper respiratory tract, skeletal muscle, skin, subcutaneous tissue, eyes, gastrointestinal tract, heart, thyroid, head/neck, brain, cervix, and breast [2]. RDD of the breast is very rare and there have been only about 20–25 cases reported, with only 3 cases in the male breast. Typically it is identified in a palpable mass, which can be localized with ultrasound, or as an abnormality on mammogram [3].

2. Presentation of case

A 55 year-old male had presented to the Ellen Hermanson Breast Center at Southampton Hospital in March 2015 with a complaint of a painless right breast lump. The mammogram had shown a 3 cm sub-areolar mass at 12:00 position in the right breast with irregular margins. The ultrasound had demonstrated an abnormal echo-texture in an infiltrative pattern without a discrete mass in the right breast [Fig. 1]. A biopsy was performed which had revealed atypical lymphoid tissue.

His past medical history was significant for erectile dysfunction, dyslipidemia, glaucoma, vitamin D deficiency, hypertension, diabetes mellitus type 2, a history of deep vein thrombosis, and a prior inguinal hernia repair. Family cancer history included liver cancer in his father. Physical examination revealed a slightly enlarged right breast with a 3 × 5 cm firm mobile mass in the superior retro-areolar region. There was no evidence of nipple distortion or discharge and the left breast exam was unremarkable.

The patient underwent an excision of the right breast mass. The pathology revealed a mixed histiocytic and lymphoplasmacytic infiltrate with sclerosis and histiocytes exhibiting emperipolesis [Fig. 2]. The findings were consistent with extranodal Rosai-Dorfman disease and a second opinion confirmed the diagnosis.

3. Discussion

RDD is a benign disease that presents in the first few decades of life and has a male predominance with a higher incidence in African Americans than Caucasians. The classic presentation of bilateral non-tender cervical adenopathy occurs in approximately 90% of
Common additional symptoms include fever and weight loss with laboratory elevations of white cells and ESR [1]. It has an idiopathic etiology, although there is suggestive evidence that viruses such as parvovirus B19, human herpesvirus (HHV), and Epstein–Barr virus (EBV) may have a role [4]. The hallmark of RDD is histiocytes with emperipolesis, which is the presence of engulfed and intact inflammatory cells within the cytoplasm of the histiocyte where the engulfed cell remains viable as opposed to phagocytosis where it does not. Histiocyte immunostaining is positive for CD68, CD14, S100, CD33, CD11C, and negative for CD1a [2].

Treatment for RDD is recommended only for those who have symptoms with systemic or vital organ involvement. Relapsing and remitting disease without any intervention occurs in approximately 70% of patients. Twenty percent of patients have regression of the disease without treatment. Localized disease causing symptoms is usually treated with surgical excision with observation post-procedure. Symptomatic systemic disease is treated with steroids as a first line agent. Other therapies with chemotherapy and radiation for systemic disease have varying levels of success [4].

On mammography, a poorly defined mass without evidence of calcifications is typically found. Ultrasound, however, tends to show a hypoechoic mass. A diagnosis can be made by core needle biopsy, fine needle aspiration (FNA), or excisional biopsy [4]. Cutaneous cases of RDD presenting on the breast skin have been identified as well. The first was in 1994 in a 34 year old man with the most recent one in January 2015 on a 72 year old woman’s right breast [5,6].

To our knowledge, there are only three other reported cases of RDD that were found within a male breast mass in 2007, 2010, and 2012. The patients were aged 23, 35, and 59, respectively. The 59 year old presented with the only case of multiple masses in the left breast [7–9]. In addition to the 35 year old’s case, this is the only other one identified that presented in the right breast. The other two cases were in the left breast.

4. Conclusion

It appears that RDD of the breast has a relatively indolent course that presents without systemic symptoms. Given that RDD has a
tendency to regress on its own, treatment for these breast masses beyond diagnosis seems to be close observation. While there have been documented cases of morbidity and mortality with RDD in general, no cases to our knowledge have been reported due to breast involvement. With the extremely rare nature of this disease, diagnosis, especially in an uncommon site like the breast, can prove to be difficult [5].

**Conflicts of interest**

There are no conflicts of interest.

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**Ethical approval**

Approval was not required for this manuscript.

**Author contribution**

1. BenFauzi El-Attrache, DO – writing the paper, data collection and interpretation.
2. Edna Kapenhas, MD – study concept, case supervision, and editing
3. Jack Morgani, MD – interpretation of ultrasound imaging.
4. Tahameena Ahmed, MBBS – diagnosis of disease from pathology specimen.

**Consent**

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

**Registration of research studies**

This is not required for this manuscript.

Guarantor

Edna Kapenhas.

**References**

[1] N. Symns, G. Cugati, M. Vasudevan, R. Ramamurthi, A. Pande, Intracranial Rosai-Dorfman Disease: report of three cases and literature review, Asian J. Neurosurg. 5 (2) (2010) 19–30 (Accessed April 6, 2015) http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3201083.

[2] S. Sharma, S. Bhardwaj, D. Hans, Rosai-Dorfman disease, JKI Sci. 12 (4) (2010) 194–196 (accessed April 6, 2015) http://www.jkscience.org/archive/volume124/Rosai-DorfmanDisease.pdf.

[3] C. Pham, L. Abruzzo, E. Cook, G. Whitman, T. Stephens, Rosai-Dorfman disease of the Breast, Am. J. Roentgenol. 185 (4) (2005) 971–972 (Accessed April 6, 2015) http://www.ajronline.org/article-pdf/10.2214/AJR.05.0224.

[4] D. Samir, E. Sagatsy, S. Lobomir, K. Timothy, Rosai-Dorfman disease: tumor biology, Clinical features, pathology, and treatment, Cancer Control 21 (4) (2014) 322–326 (Accessed April 6, 2015) https://www.mercy.net/sites/default/files/vendor-sources/dalia_et_al_ccj_rosai_dorfman_oct_2014.pdf.

[5] L Green, R. Dorfman, J. Rosai, Breast involvement by extranodal rosai-Dorfman disease: report of seven cases, Am. J. Surg. Pathol. 21 (6) (1997) 664–668 (Accessed April 6, 2015) http://ovidsp.ovid.com.arktos.nyist.edu/sp-3.21.0a/ovidweb.cgi?r6=546&u=10.2214/AJR.05.0224&url=http://www.ajronline.org/doi/pdf/10.2214/AJR.05.0224.

[6] M. Nadal, T. Kervarrec, M. Machet, T. Petrella, L. Machet, Cutaneous Rosai-Dorfman disease located on the Breast: rapid effectiveness of methotrexate after failure of topical corticosteroids, acitretin and thalidomide, Acta Derm. Venereol. 95 (2015) 758–759 (Accessed April 12, 2015) http://www.google.com/url?sa=t&rct=j&q=&esrc=s&source=web&cd=4&cad=rja&uact=8&ved=0CDQQFjAD&url=http://www.ncbi.nlm.nih.gov/pubmed/26757792.

[7] A. Pereira, A. Keleher, M. Nath, Rosai-Dorfman disease presenting as a male breast mass, The American Surgeon 73 (3) (2007) 294–295 (Accessed April 6, 2015) http://www.ncbi.nlm.nih.gov/pubmed/17377592.

[8] P. Bansal, S. Chakraborti, G. Krishnamand, R. Bansal, Rosai-Dorfman disease of the breast in a male: a case report, Acta Cyrol. 54 (3) (2010) 349–352 (Accessed April 6, 2015) http://www.ncbi.nlm.nih.gov/pubmed/20518426.

[9] P. Baladandapani, Y. Hu, K. Kapoor, L. Merriam, P. Fisher, Rosai-Dorfman disease presenting as multiple breast masses in an otherwise asymptomatic male patient, Clin. Radiol. 67 (4) (2012) 393–395 (accessed April 6, 2015) http://www.clinicalradiologyonline.net/article/S0009-9260(11)00456-9/abstract.

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