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Novel Pseudoangiomatous Stromal Hyperplasia Mass in a Breast Hamartoma: A Case Report

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Abstract Breast fibroadenolipomas, otherwise known as hamartomas, are a relatively rare finding in regard to the occurrence of breast masses. Breast hamartomas make up approximately 4.8% of benign breast tumors in women. Breast hamartomas are uncommon, and our case presents with a 4.7 cm hamartoma with a focal, irregular mass inside of it which was histologically confirmed as pseudoangiomatous stromal hyperplasia (PASH). PASH is a benign mesenchymal lesion of the breast that is characterized by dense collagenous stroma forming "pseudoangiomatous" capillary-like spaces lined by spindle cells. [1] A rare condition overall, PASH is predominately discovered in premenopausal women, with fewer than 1,500 cases of PASH documented in literature [1]. Therefore, our case presentation describes a combination of two relatively rare phenomena within one palpable breast mass. Due to the absence of symptoms, the patient was scheduled for interval follow up to monitor the breast hamartoma for interval change in size, and definitive treatment by elective surgical excision was offered if accompanying symptoms developed.

Keywords: *breast hamartoma, breast mass, pseudoangiomatous stromal hyperplasia, ultrasound, mammography*

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1. Introduction

PASH presents most commonly in pre- and perimenopausal women, which provides supporting evidence that hormonal factors may play a role in its development. [1] One series of 149 patients with PASH found that the majority of larger, palpable breast masses containing PASH were found in pre-menopausal women, whereas the majority of smaller, non-palpable PASH lesions were diagnosed via screening studies in post-menopausal women. [2] Additionally, more post-menopausal women prescribed hormone replacement therapy were found in the palpable breast mass group than the non-palpable group, suggesting a hormonal etiology of PASH. [2]

Breast fibroadenolipomas, otherwise known as hamartomas, are a relatively rare finding in regard to occurrence of breast masses. Breast hamartomas make up approximately 4.8% of benign breast tumors in women. [3] Breast hamartomas are generally found in women above the age of 35, and are sometimes referred to as "a breast within a breast", as they are composed of fibrous, glandular, and fatty tissue. [4] Mammary hamartomas vary greatly in size, from 17 mm to 170 mm. [4] We report a case of a palpable right breast mass in a 45-year-old female with imaging findings consistent with a breast hamartoma. This case represents an unusual presentation due to the fact that this breast hamartoma contains a focal, irregular, hypoechoic mass within the borders of the hamartoma itself. Atypical features such as these necessitate further workup with biopsy and pathologic correlation, as there are few reported incidences of invasive ductal carcinoma that have arisen from a breast hamartoma. [5]

2. Case Presentation

A 45-year-old woman presented with a large painless right breast mass that was first noticed 6 months prior to presentation. The patient denied any personal and family history of breast cancer.

On physical examination, there was a palpable and nontender breast mass located at the 10:00 o'clock position of the right breast, 7 cm from the nipple. No skin distortion, discoloration, retraction, or axillary lymphadenopathy was noted. Diagnostic mammography revealed a heterogeneously dense breast composition and an oval mass with high density and indistinct borders in the upper outer quadrant of the right breast (Figure 1). These findings were consistent with BI-RADS 4A category – low suspicion for malignancy (2-9%).

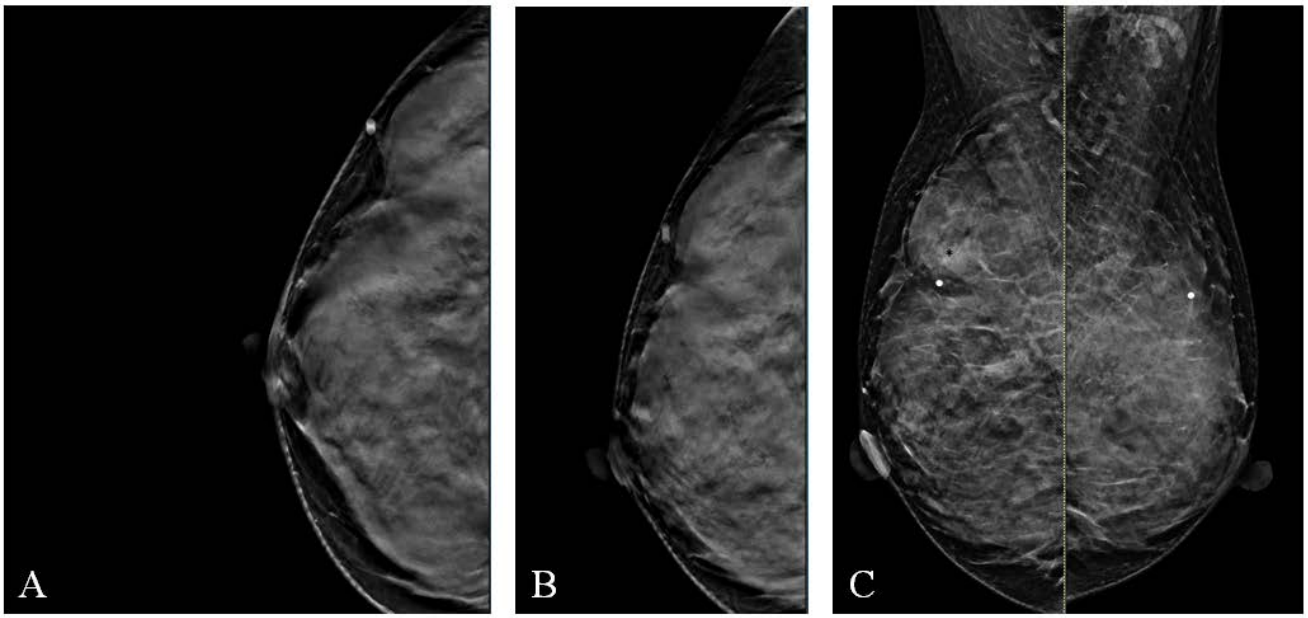

Figure 1. (A) Craniocaudal view of the right breast with palpable mass marker. (B) Exaggerated craniocaudal view of the right breast with palpable mass marker. (C) Bilateral mediolateral oblique views showing comparison of both breasts with markers at palpable breast mass and same relative breast region on the contralateral breast

A breast ultrasound was performed to further characterize the breast mass, which further demonstrated a 4.7 cm oval mass with parallel orientation and no calcifications within the mass. The overall mass had heterogeneous echogenicity and appeared to contain both fibroglandular and fatty tissue (Figure 2). In the mid-anterior portion of the mass, there was a 1.0 cm x 0.7 cm area of hypoechoic asymmetry. This focal asymmetry within the larger mass contained irregular, lobulated margins, and contained mixed posterior enhancement (Figure 3). This focal finding raised suspicion enough to warrant further workup, and ultrasound-guided biopsy was subsequently performed.

Core biopsy of the larger 4.7 cm mass revealed fibrocystic changes with fibrosis, apocrine metaplasia, and usual ductal hyperplasia, consistent with a breast hamartoma. The smaller asymmetry within the hamartoma was also biopsied, revealing pseudoangiomatous stromal hyperplasia – negative for malignancy. Due to the absence of symptoms, the patient was scheduled for interval follow up to monitor the breast hamartoma for interval change in size, and definitive treatment by elective surgical excision was offered if accompanying symptoms developed.

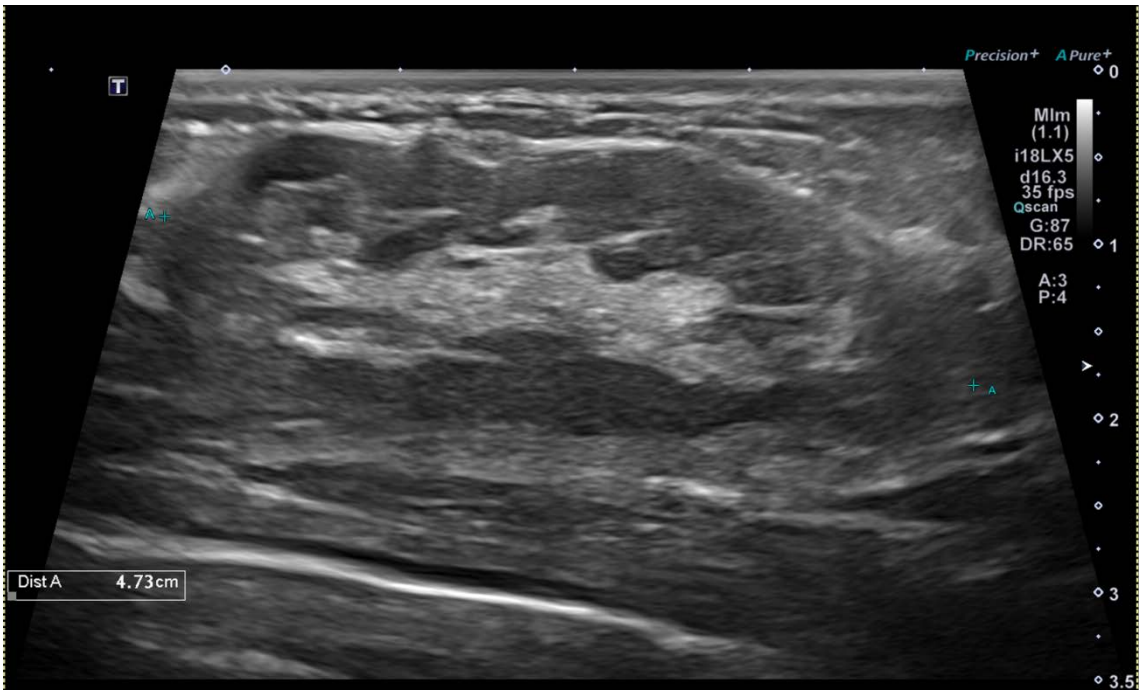

Figure 2. Ultrasound demonstrating size dimensions of the right breast mass (4.73cm). The mass was visualized at 10:00 o'clock of the right breast, 7 cm from the nipple

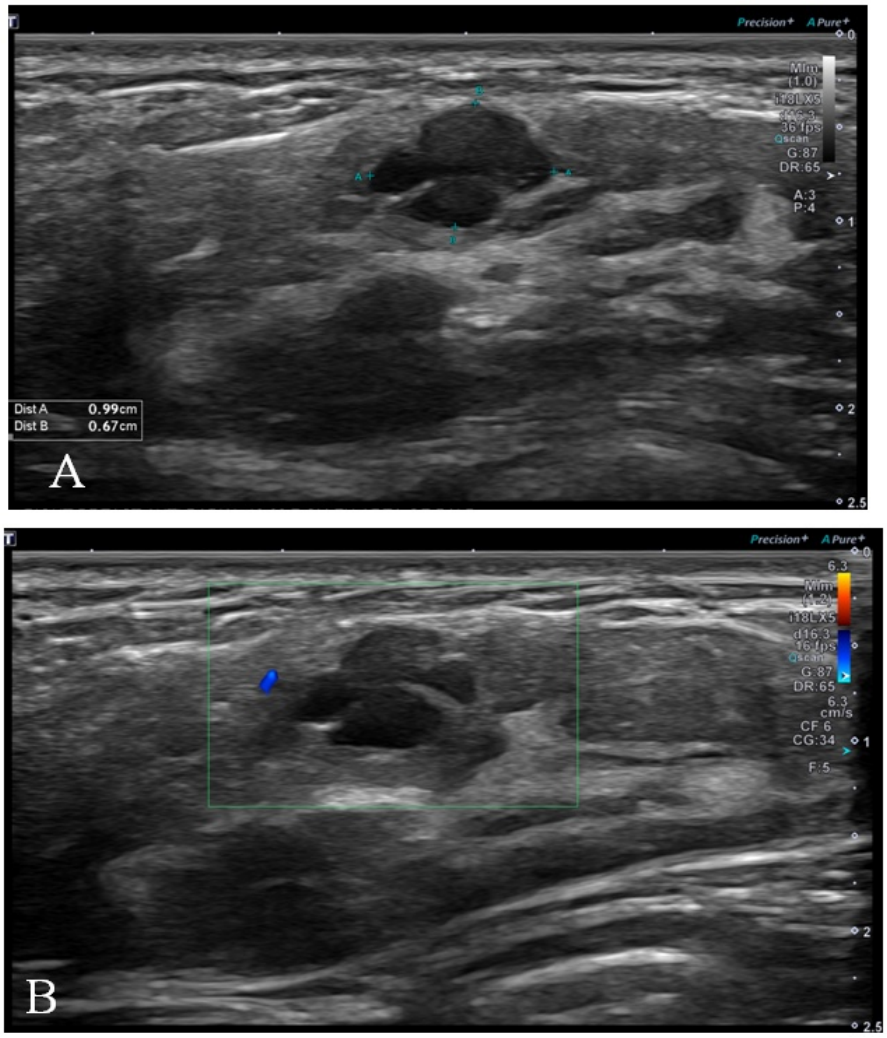

Figure 3. (A) Ultrasound demonstrating the size dimensions of the focal asymmetry within the breast hamartoma. (B) Color doppler ultrasound of the asymmetry within the breast hamartoma demonstrating avascularity

3. Discussion

Since the discovery of pseudoangiomatous hyperplasia in 1986 by Vuitch *et al* to 2007, there have been less than 150 reported cases of tumor-forming pseudoangiomatous hyperplasia. [6,7] The pathogenesis of pseudoangiomatous hyperplasia still remains unclear, however, research is emerging suggesting that hormones may play a significant role. [8] This may provide insight as to why pseudoangiomatous hyperplasia primarily occurs in premenopausal and perimenopausal women. [9] Pseudoangiomatous stromal hyperplasia can be difficult to diagnose because it can present with solely microscopic findings, mammographically evident benign breast masses, or both. [9]

Breast hamartomas are benign masses that are also referred to as lipofibroadenomas, fibroadenolipomas, or adenolipomas. [10] The first description of breast hamartomas in literature is a case series report from 1971 describing masses resembling fibroadenomas. [11] The incidence of breast hamartomas occurs in 4.8% of women with benign masses. [3] However, hamartomas are commonly mistaken for fibroadenomas and the incidence is believed to be much higher than 4.8%. [3] Despite being identified over 50 years ago, the exact pathogenesis of breast hamartomas still remains a mystery to this day.

Although the exact pathogenesis is unknown, it is important to note the relationship between hamartomas and female sex hormones. Two immunohistochemical studies collectively examined over 30 patients and estrogen and progesterone receptors were found in over 90% of the cases. [12,13] The presence of the sex hormone receptors explains increased growth rate of hamartomas during pregnancy. [13]

On physical examination, hamartomas are typically occult. If a mass is discernible, it presents as a large, mobile, soft to firm mass. [14] On mammography, hamartomas are well-circumscribed, round to oval masses with a thin, and radiopaque with a pseudocapsule. On ultrasound, the mass presents as a sharply defined heterogeneous oval mass or as normal glandular tissue. [14]

Breast hamartomas are uncommon, and our case presents with a pseudoangiomatous mass with stromal hyperplasia inside of the hamartoma. A report on 14 breast hamartomas exhibited evidence of myoid differentiation, epithelial ductal hyperplasia, fibrocystic mastopathy, apocrine metaplasia, and cyst formation. [15] The mass identified in our case did have evidence of fibrocystic changes and apocrine metaplasia. Our mass also had pseudoangiomatous stromal hyperplasia present on pathology. Having a pseudoangiomatous mass within a

breast hamartoma is an exceptionally rare lesion, however, it has been reported sparsely in case studies. [16,17]

The research on breast hamartomas is constantly expanding, however, more information is needed to understand pseudoangiomatous stromal hyperplasia within hamartomas. Further research needs to be done to examine the prevalence, incidence, and pretest probability of pseudoangiomatous stromal hyperplasia occurring with breast hamartomas.

4. Conclusion

Breast hamartomas represent a very small percentage of benign breast masses in women. It is important to note that although they are characterized as benign masses, hamartomas are consolidated dysgenic breast tissue, and therefore have a capability of developing malignant features within them. [4] Our case reveals an exceptionally rare presentation of a breast hamartoma, as pseudoangiomatous stromal hyperplasia within the confines of the hamartoma is not well characterized in literature. Pseudoangiomatous stromal hyperplasia must be differentiated from low-grade angiosarcoma and phyllodes tumors, as imaging alone cannot distinguish from these findings. [18] Our case exemplifies the workup necessary in ruling out malignant disease within a breast hamartoma. Research into the prevalence and characterization of pseudoangiomatous stromal hyperplasia can augment our understanding of the presentation of masses found within breast hamartomas.

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Statement of Competing Interests

The authors have no competing interests.

References

[1] Estes, Ashley, et al. “Pseudoangiomatous Stromal Hyperplasia: Overview and Clinical Management.” *Annals of Breast Surgery*, AME Publishing Company, 13 Nov. 2020, https://abs.amegroups.com/article/view/6368/html.

[2] Hargaden GC, Yeh ED, Georgian-Smith D, Moore RH, Rafferty EA, Halpern EF, McKee GT. Analysis of the mammographic and sonographic features of pseudoangiomatous stromal hyperplasia. AJR Am J Roentgenol. 2008 Aug; 191(2): 359-63.

[3] Sevim, Y., Kocaay, A. F., Eker, T., Celasin, H., Karabork, A., Erden, E., & Genc, V. (2014). Breast hamartoma: a clinicopathologic analysis of 27 cases and a literature review. *Clinics (Sao Paulo, Brazil)*, *69*(8), 515-523.

[4] Bhatia, Mukul, et al. “‘Breast within a Breast’ Sign: Mammary Hamartoma.” *Medical Journal, Armed Forces India*, Elsevier, Oct. 2015, https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4646936/.

[5] Lee, E.H., et al. “Invasive Ductal Carcinoma Arising in a Breast Hamartoma: Two Case Reports and a Review of the Literature.” *Clinical Radiology*, W.B. Saunders, 29 Jan. 2003, https://www.sciencedirect.com/science/article/abs/pii/S0009926002004567.

[6] Vuitch, M. F., Rosen, P. P., & Erlandson, R. A. (1986). Pseudoangiomatous hyperplasia of mammary stroma. Human pathology, 17(2), 185-191.

[7] Wieman, S. M., Landercasper, J., Johnson, J. M., Ellis, R. L., Wester, S. M., Lambert, P. J., & Ross, L. A. (2008). Tumoral pseudoangiomatous stromal hyperplasia of the breast. The American surgeon, 74(12), 1211-1214.

[8] Pruthi, S., Reynolds, C., Johnson, R. E., & Gisvold, J. J. (2001). Tamoxifen in the management of pseudoangiomatous stromal hyperplasia. *The breast journal*, *7*(6), 434-439.

[9] Bowman, E., Oprea, G., Okoli, J., Gundry, K., Rizzo, M., Gabram-Mendola, S., Manne, U., Smith, G., Pambuccian, S., & Bumpers, H. L. (2012). Pseudoangiomatous stromal hyperplasia (PASH) of the breast: a series of 24 patients. The breast journal, 18(3), 242–247.

[10] Tse, G. M., Law, B. K., Ma, T. K., Chan, A. B., Pang, L. M., Chu, W. C., & Cheung, H. S. (2002). Hamartoma of the breast: a clinicopathological review. *Journal of clinical pathology*, *55*(12), 951-954.

[11] Arrigoni, M. G., Dockerty, M. B., & Judd, E. S. (1971). The identification and treatment of mammary hamartoma. *Surgery, gynecology & obstetrics*, *133*(4), 577-582.

[12] Chiacchio, R., Panico, L., D'Antonio, A., Delrio, P., Bifano, D., Avallone, M., & Pettinato, G. (1999). Mammary hamartomas: an immunohistochemical study of ten cases. *Pathology, research and practice*, *195*(4), 231-236.

[13] Herbert, M., Sandbank, J., Liokumovich, P., Yanai, O., Pappo, I., Karni, T., & Segal, M. (2002). Breast hamartomas: clinicopathological and immunohistochemical studies of 24 cases. *Histopathology*, *41*(1), 30-34.

[14] Feder, J. M., de Paredes, E. S., Hogge, J. P., & Wilken, J. J. (1999). Unusual breast lesions: radiologic-pathologic correlation. *Radiographics : a review publication of the Radiological Society of North America, Inc*, *19 Spec No*, S11-S260.

[15] R.A. Amir, S.S. Sheikh, Breast hamartoma: A report of 14 cases of an under-recognized and under-reported entity, International Journal of Surgery Case Reports, Volume 22, 2016, Pages 1-4, ISSN 2210-2612.

[16] Baik, H., Kim, J. W., Park, Y. M., Jung, S. J., Lee, A., Yoon, H. K., & Kim, T. H. (2016). Huge Bilateral Breast Hamartoma Accompanied with Pseudoangiomatous Stromal Hyperplasia. *Journal of Breast Disease*, *4*(1), 28-32.

[17] Su, C. C., Chen, C. J., Kuo, S. J., & Chen, D. R. (2015). Myoid hamartoma of the breast with focal chondromyoxid metaplasia and pseudoangiomatous stromal hyperplasia: A case report. *Oncology letters*, *9*(4), 1787-1789.

[18] Bowman, Erin, et al. “Pseudoangiomatous Stromal Hyperplasia (PASH) of the Breast: A Series of 24 Patients.” *The Breast Journal*, U.S. National Library of Medicine, 2012, https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3658111/.