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Clinical report

Clues to imaging diagnosis of primary angiosarcoma of the breast

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Background: Primary angiosarcoma of the breast is rare and there are no defined diagnostic criteria for breast imaging.
Objective: To describe breast imaging of two cases of histologically proven angiosarcoma and identify clues to imaging diagnosis.
Methods: Clinical study of two female patients with breast lumps at King Chulalongkorn Memorial Hospital. The modalities of breast imaging included mammography, ultrasongraphy and magnetic resonance imaging (MRI).
Results: Both female patients were young adults, aged 26 and 22 seen in 2002 and 2004. They manifested huge breast lumps. The first patient had bilateral simultaneous involvement. The left breast was very large and not compressible on mammography. It showed a rapid growth and bleeding ulceration. The second patient suffered from unilateral left breast enlargement with tenderness. Both masses were hyperdense on mammogram and hypoechogenicity with hypervascularity on color Doppler ultrasound. The MRI findings supported vascular masses with internal hemorrhage. An early subacute phase of hemorrhage was displayed by the second patient. Both were operated but succumbed to lung metastatses.
Conclusion: The imaging diagnostic clues consisted of huge masses without pathologic lymph node on the mammograms, hypervascularized masses in color Doppler ultrasound, hemorrhagic areas and strong enhancement demonstrable by MRI. Another clue is presence on imaging of lung metastasis at the time of initial diagnosis.

Keywords: Angiosarcoma, breast, imaging.

Although angiosarcoma of the breast is rare, representing less than 1% of all primary breast malignancies and 3% to 10% of all breast sarcomas [1], it is increasing in prevalence. The increasing use of radiation to conserve breast is now widespread and may be one cause [2]. When the disease occurs *de novo*, it is called primary angiosarcoma of the breast. The preoperative diagnosis is challenging. The clinical clue is a large breast lump with bluish discoloration or bruising of the overlying skin in a young adult [3]. Imaging in most cases shows a low echoic large lump with lobulated structure on sonogram and a solitary uncalcified ill-defined lump on mammograms [4, 5]. We report two cases and emphasize clues to imaging diagnosis that require understanding of the natural course as well as findings on mammograms, ultrasounds and magnetic resonance imaging (MRI).

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Case reports

Case 1

A 26-year-old female presented with a rapidly growing left breast mass with bleeding ulcerations. The main mass was 3 cm in diameter together with a smaller movable mass in her right breast. The left mammogram was not performed due to a non-compressible breast. Mammographically, the right breast showed a well-defined isodense mass (**Fig. 1A**). There were no axillary lymphnodes. Ultrasound revealed one hypoechoic mass in the right and several hypoechoic masses in the left breast. The masses were well-defined and hypervascularized (**Fig. 1B**). Axial T1 weighted MRI imaging (T1WI) demonstrated a huge lump in her left breast consisting of hyposignal intensity with some incorporated areas

of hypersignal intensity (**Fig. 1C**). The whole tumor had a heterogeneous pattern of increased hypersignal intensity on axial T2 weighted imaging (T2WI). In addition, a small well-defined hypersignal intensity mass was disclosed in the inner part of the right breast, which suggestion simultaneous bilateral involvement with tumor (**Fig. 1D**). On post Gadolinium-enhanced dynamic study, there was intense enhancement of the mass containing internal areas of hemorrhage with low signal intensity (**Fig. 1E**). Computerized tomography scan of the chest exhibited evidence of lung metastases. Imaging was suggestive of angiosarcoma. She had a left mastectomy and excisional biopsy of the right breast mass lump. Histopathological studies confirmed the diagnosis of angiosarcoma of both breasts. She was lost to follow-up.

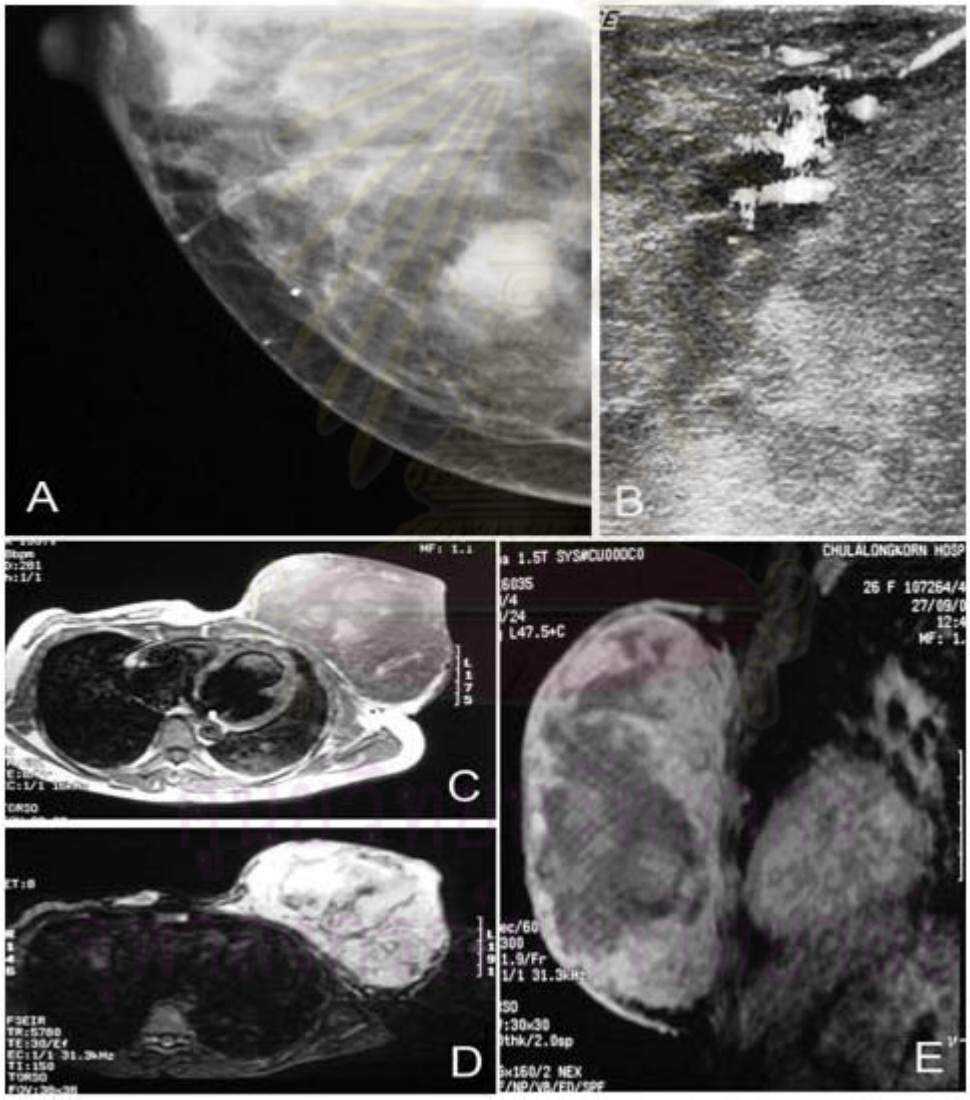

Fig. 1 A: The mammogram showing a partially well-defined hyperdense lesion of the right breast; **B:** A hypoechoic mass with hypervascularity by color Doppler sonogram; **C:** Axial T1 weighted-image of the left breast demonstrating the mass having hyposignal but internal areas of hemorrhage with hypersignal intensity; **D:** The hemorrhage areas and the whole mass exhibiting heterogeneous increased signal intensity on axial T2 weighted-image. Note that the right breast tumor shows hypersignal density; **E:** Sagittal view of post Gadolinium-enhanced dynamic study revealing enhancement of the mass except for the area of internal hemorrhage.

Case 2

A 22-year-old female presented with enlargement, tenderness and bluish discoloration of her left breast. A mammogram showed an enlarged left breast of diffusely increased density (**Fig. 2A**). There were no axillary lymph nodes. Ultrasound demonstrated a big diffusely heterogeneous hypoechoic mass involving almost the entire left breast with some acoustic shadowing and hypervascularity (**Fig. 2B**). MRI revealed the left breast being occupied by a big mass with hyposignal intensity on T1WI, hypersignal intensity on T2WI and enhancement on post Gadolinium administration. The mass also contained areas of early subacute hemorrhage at the central portion. This showed hypersignal intensity on T1WI and hyposignal intensity on T2WI (**Fig. 2 (C, D, E**)). Several vascular channels were evident in the mass. Thickening of the overlying skin was seen. The imaging diagnosis suggested angiosacoma. She underwent left mastectomy. The histological diagnosis verified angiosarcoma. One year after surgery, she succumbed to lung metastases.

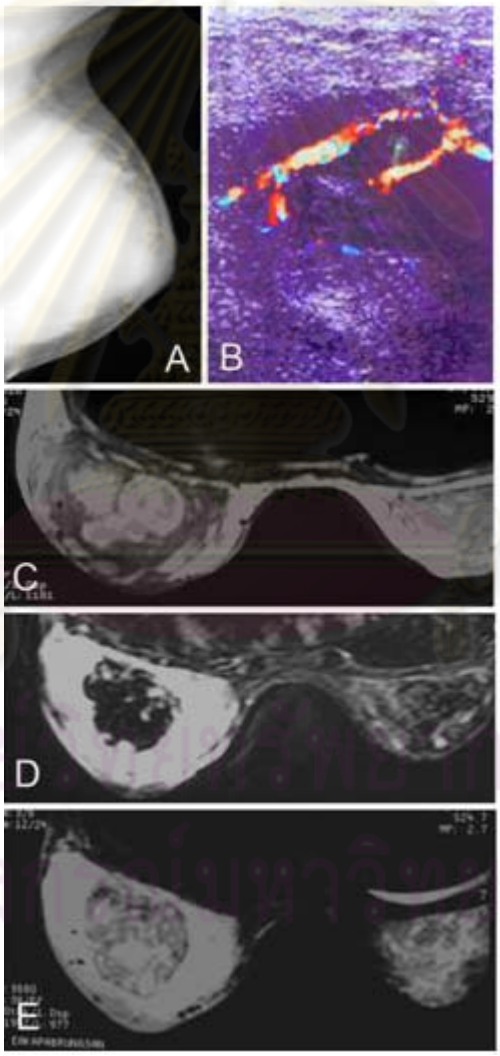

Fig. 2 A: Mammogram MLO view showing diffuse homogeneous high density of the enlarged left breast; **B:** Color Doppler Sonogram showing hypoechoic mass with hypervascularity, red colour indicated vessels penetrating the mass. (Explain what red or blue color indicates); **C:** Axial T1 weighted-image; **D:** Axial T2 weighted-image revealing early subacute hemorrhage within the mass, showing high signal intensity on T1 weighted- image and low signal intensity on T2 weighted-image; **E:** post Gadolinium enhanced dynamic study demonstrating strong enhancement of the big mass with nonenhancing areas of hemorrhage in the central portion.

Discussion

Primary angiosarcoma of the breast occurs most commonly in young women in the third and fourth decades [5]. Our two patients were 26 and 22 years of age. Both manifested large breast lumps. The first patient had simultaneous bilateral breast involvement with the left breast larger and not compressible for mammography. The second case presented also with a mass involving nearly the entire left breast. There were no axillary lymph nodes on mammograms of both patients. This was the first imaging clue that radiologists should keep in mind that it is unlikely for a big carcinoma, not result in nodal metastases. Ultrasonography is helpful since angiosarcomas are hypervascular. The second diagnostic clue was the hypervascularity pattern that appeared with color Doppler and was elicited in both of our cases. Nevertheless, the presence of hypervascularity is not a clue for angiosarcoma since many carcinomas show feeding vessels on color Doppler as well [6]. To discriminate angiosarcoma from hypervascularized carcinoma is the hemorrhage that usually happens in angiosarcoma [7]. It is not a natural event in carcinoma. MRI of angiosarcoma usually shows a mass with low signal intensity on T1WI, but high signal intensity on T2WI. The latter suggests vascular channels containing slow-flowing blood [8, 9]. MRI is a useful imaging modality to demonstrate hemorrhage. Furthermore, it can identify stages of hemorrhage by the characters of signals in T1 and T2 WI. Therefore, hemorrhage of vascular masses in both of our patients represented a third imaging clue. Both also documented masses with hyposignal intensity on T1WI, hypersignal intensity on T2WI and strong enhancement supporting vascular tumors. These hemorrhages caused different signal intensity, dependent on the stage of hemorrhage. The first patient had fresh hemorrhages. It provoked hypersignal intensity on T1WI and T2WI. By contrast, the mass of the second patient elicited internal areas of hypersignal intensity on T1WI but hyposignal intensity on T2WI. The latter finding was compatible with hemorrhage in an early subacute stage. All the hemorrhages gave non-enhancing areas on post Gadolinium administration. Additionally, the detection of lung metastasis at the time of breast imaging is another clue that supports a pre-operative diagnosis of angiosarcoma.

In conclusion, the pre-operative imaging diagnosis of primary anigosarcoma of the breast is possible when the radiologist has the following clues in mind: A huge mass without pathologic lymph node on mammogram, hypervascularized mass on color Doppler ultrasound, hemorrhagic areas and strong enhancement demonstrable by MRI. Another clue, if present, is lung metastases at the time of diagnosis.

References

1. Wong RWY, Rappaport WD, Davis JR, Leong SPL. Primary angiosarcoma of the breast: a case report and review of the literature. Surg Rounds.1996;19:454-8.
2. West JG, Qureshi A, West JE, Chacon M, Sutherland ML, Highighi B, Harrison J. Risk of angiosarcoma following breast conservation: a clinical alert. The Breast Journal 2005;11:115-23.
3. Ohta M, Tokuda Y, Kuge S, Okumura A, Tanaka M, Kubota M et al. A case of angiosarcoma of the breast. Japanese Journal of clinical oncology 1997;27:91-5.
4. Farrokh D, Hashemi J, Zandi B, Iran J Radiol. 2006, 3:81.
5. Liberman L, Dershaw DD, Kanufman RJ, Rosen PP. Angiosarcoma of the breast. Radiology 1992;183: 649-54.
6. Boonjunwetwat D, Prueksadee J, Sampatanukul P, Chatamra K. Does color Doppler ultrasound vascularity predict the response to neoadjuvant chemotherapy in breast cancer? J Med Assoc Thai. 2005;88:1367-72.
7. http://surgpathcriteria.stanford.edu/breast/angiosarcbreast/differentialdiagnosis.html
8. Glazebrook KN, Morton MJ, Reynolds C. Vascular tumors of the breast: Mammographic, sonographic, and MRI appearances. Am J Roentgenol. 2005;184: 331-8.
9. Marchant LK, Orel SG, Perez-Jaffe LA, Reynolds C, Schnall MD. Bilateral angiosarcoma of the breast on MR imaging. Am J Roengenol. 1997; 169: 1009-10.