

Awareness of primary spontaneous hemorrhagic angiosarcoma of the breast associated with Kasabach–Merritt syndrome in a pregnant woman by enhanced magnetic resonance imaging

A CARE-compliant case report

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Abstract

Introduction: Primary breast angiosarcoma with spontaneous intratumoral bleeding associating with Kasabach-Merritt Syndrome is rarely reported.

Case Findings/Patient Concerns: We herein present such a case in a 30-year-old pregnant woman who was initially diagnosed to hemangioma at her early gestation. However, the sudden rapid tumor growth was aware of the attention and intended for receiving the breast enhanced magnetic resonance imaging.

Diagnoses and Interventions: The dynamic MRI enhancement showed inhomogenous enhancement at the periphery of the lobulated tumor on both early and delayed scans, otherwise a large hematoma was revealed at the center. Surgical resection was performed after baby delivery by Caesarean section, and histopathologic study confirmed breast angiosarcoma.

Conclusion: Despite its rarity, clinicians should recognize the association of breast angiosarcoma with Kasabach-Merritt Syndrome with suggestive finding of enhanced MRI in order to decide the surgical approach.

Abbreviation: MRI = magnetic resonance imaging.

Keywords: breast angiosarcoma, breast tumor, intratumoral bleeding, Kasabach–Merritt syndrome, magnetic resonance imaging

1. Introduction

Primary angiosarcoma of the breast is a rare and rapidly progressive malignant tumor, accounting for only 0.04% of primary malignant breast tumors and 8% of breast sarcomas.^[1,2] We herein present such an unusual case of primary breast angiosarcoma with sudden enlargement secondary to spontaneous intratumoral bleeding in a pregnant woman. Spontaneous bleeding of a primary breast angiosarcoma with

Kasabach–Merritt syndrome (secondary to the consumption coagulopathy) is a rare complication. Despite its rarity, clinicians should recognize the association of breast angiosarcoma with Kasabach–Merritt syndrome, possibility of rapid increase in tumor size secondary to spontaneous intra-tumoral hemorrhage. The enhanced magnetic resonance imaging (MRI) could demonstrate further more features for management decision.

2. Case presentation

The institutional review board approved this study (Chang Gung Medical Foundation) and waived the need for the patient's inform consent. A 30-year-old primigravida pregnant woman at 33 weeks gestation was referred to our hospital with a complaint of rapid growing left breast mass. She had a palpable lump at the upper outer quadrant of her left breast at the beginning of pregnancy. However, the sonography showed a 2.5-cm hypo-echoic nodule with smooth outline and the core needle biopsy subsequently diagnosed to hemangioma at that time. The lesion remained stable with slow tumor growing during the course of pregnancy. Unfortunately, the fast tumor growth and aggravating pain were noted in the recent 2 weeks at 33 weeks gestation. Physical examination showed an 18 cm hard painful left breast mass with engorged vessels and reddish-purple discoloration. Laboratory tests showed abnormal blood count with low hemoglobin level (8.9 g/dL), platelets count ($93 \times 10^3/\mu\text{L}$). A vascular tumor with aggravate growth was suspicious; however, the actual etiology could not definitely diagnosed.

A series of imaging examination was thus arranged. Breast sonography revealed a huge and heterochoic mass with poor demonstration of tumor border. The color Doppler did not

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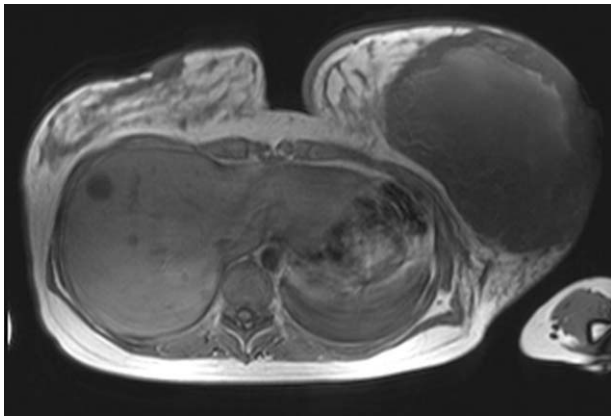


Figure 1. Axial T1-weighted image of MRI showing a large left breast mass with hyperintensity at the center and hypointensity at periphery, which can be due to central intra-tumoral hematoma. MRI = magnetic resonance imaging.

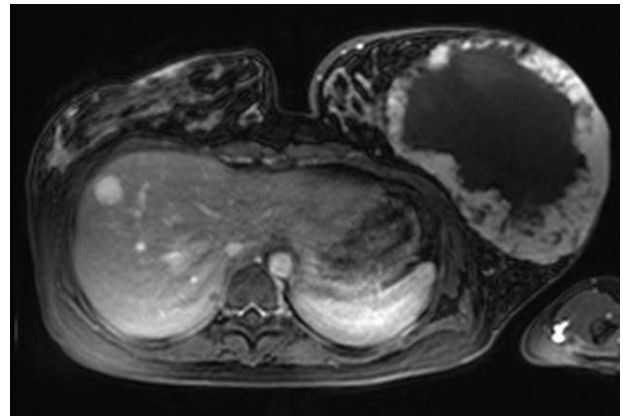


Figure 3. Axial enhanced T1-weighted image with fat suppression at early phase enhancement (1 minute after gadolinium injection) showing remarkable inhomogeneous enhancement at the peripheral portion of tumor with nonenhancement at the center.

display any color flow within the tumor. The dynamic enhanced MRI was thus decided to evaluate the intratumoral features. Precontrast MRI revealed a $19 \times 15 \times 13 \text{ cm}^3$ lobular, circumscribed soft tissue mass at the upper region of the left breast. A large hematoma located at the center of tumor (Figs. 1 and 2). The enhanced MRI showed the tumor was inhomogeneously good enhanced in periphery on both early and delayed scans (Figs. 3 and 4). There was no feature of centripetal enhancement or cotton wool enhancement over the breast tumor. An unusual angiosarcoma with intratumoral bleeding was thus suspected instead of a hemangioma.

The patient decided to receive tumor resection after baby delivery by caesarean section immediately. Histopathologic examination confirmed angiosarcoma with microscopic findings of large area of bridging anaestomotic vascular spaces at the periphery of the tumor with multiple foci of malignant cells and a large hematoma at the center (Fig. 5). After the operation, all the blood count returned to normal and the patient was then arranged for the following treatment.

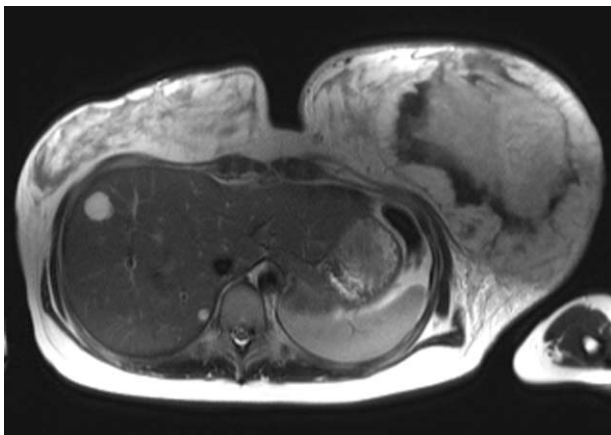


Figure 2. Axial T2-weighted image of MRI showing hyperintensity at the center and hypointensity at periphery, suggesting peripheral tumor location with central intratumoral hematoma. MRI = magnetic resonance imaging.

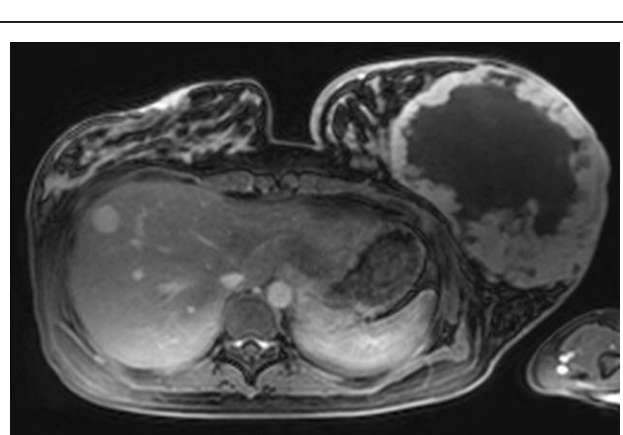


Figure 4. Axial enhanced T1-weighted image with fat suppression at delayed phase enhancement (5 minute after gadolinium injection) showing persistent pattern of tumor enhancement.

3. Discussion

Angiosarcoma is a malignant vascular tumor with endothelial lining originating from any organs in the body. As opposed to the high breast cancer prevalence in elderly patients, primary breast angiosarcoma were more often found in the younger women at 3rd or 4th decades.^[3] Even though the etiology is unclear, hormonal trigger may play a role in the etiopathogenesis of such vascular tumor, in which the presence of estrogen and glucocorticoid receptor in angiosarcoma have been reported.^[4] In addition, sudden and rapid enlargement of breast angiosarcomas in pregnant women had also been described.^[2] Both increased tumoral growth from hormonal influence and the subsequent intra-tumoral hemorrhage contributed to the rapid tumor size increase in our patient during pregnancy. With the initial biopsy diagnosis of hemangioma, it was hard to decide whether surgery with abortion or not.

Consumption coagulopathy coexisting with a rapid enlarging hemangioma in a newborn was first described by Kasabach and Merritt. Since then, only a few cases of breast angiosarcoma associating with spontaneous intra-tumoral hemorrhage and Kasabach–Merritt syndrome had been reported.^[2,5,6] Coagulopathy secondary to the trapping of platelets within the large

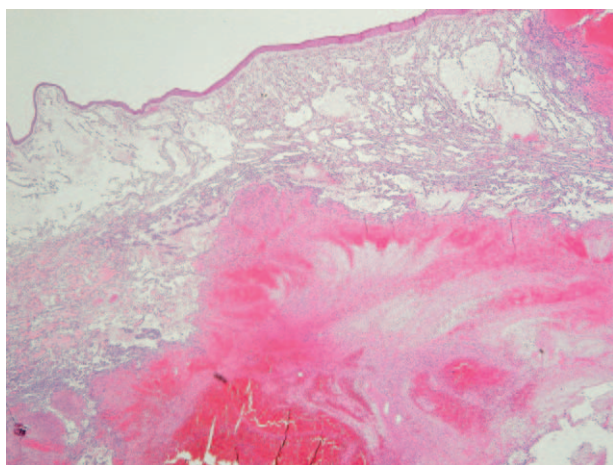


Figure 5. Histopathologic stud with microphotograph (2×) of excised angiosarcoma showing numerous anaestomatic vascular spaces and foci of malignant cells at the periphery of the tumor and hematoma at the center correlated the MRI features. MRI = magnetic resonance imaging.

vascularized tumor had been proposed as the cause of intratumoral hemorrhage, and which can explain the presence of both anemia and thrombocytopenia in our case. The clinical presentations include the disseminating intravascular coagulation and purple discoloration of overlying skin with apparent bruising, as seen in our patient.

The imaging features of breast angiosarcoma are nonspecific on conventional sonography and mammography. About 33% of angiosarcomas were not detectable mammographically.^[3] Nineteen percent of patients in the study by Yang et al^[7] had tumors that were not visible mammographically but were visible with sonography and MRI. Sonography and MRI are useful in characterizing breast lesions, but there is no distinctive feature to angiosarcomas. The mass may be hypoechoic or hyperechoic on ultrasonography.^[3] A larger series of dynamic enhanced MRI on 9 breast angiosarcomas concluded that breast angiosarcomas usually appears as large, lobular, intensely, and heterogeneously rapid enhancement with the washout characteristic of a malignant tumor.^[7] In our case, the dynamic enhanced MRI findings of hypervascular tumor with rapid enhancement, but without significant washout phenomenon, correlated to the histopathologic findings of large amount of bridging anaestomatic vascular spaces at the tumor periphery, which allows the trapping of gadolinium contrast medium within the tumor. The large amount of vascular spaces might histologically mislead to

the initial diagnosis of hemangioma. However, rapid and dynamically centripetal enhancement with cotton wool enhancement might reveal to hemangiomas rather than angiosarcomas. Occasionally, phleboliths may be a suspicious finding to distinguish the hemangioma from angiosarcom.^[8] However, the diagnosis of breast angiosarcoma virtually depends on the presence of malignant cells on histopathologic study. As for the differentiation from typical breast cancer, our presented case corresponds with the young age group as previous reported, along with the presence of thrombocytopenia and anemia due to the Kasabach–Merritt syndrome. The MRI presentation was also unlikely for typical breast cancer, which usually had rapid enhancement with washout pattern. Collectively, this case will not be favored to be breast cancer reoperatively.

4. Conclusion

Despite the atypical imaging presentation and core needle biopsy revealing hemangioma, our presented case may help to increase awareness from clinicians when confronting with rapid tumor progression from possible hormonal influence, which is unlikely for hemangioma, and surgical excision may be the definitive diagnosis. The association of Kasabach–Merritt syndrome with angiosarcoma, although rarely seen, must be taken into consideration in case of anemia and thrombocytopenia. The enhanced MRI findings might provide detailed architectural evaluation to aware the angiosarcoma; that was facilitating for instant consideration.

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