Radiotherapy for primary angiosarcoma of the breast: a case report

Yoke Surpri Marlina1, Rima Novirianthy2*, Teuku Muhammad Yus2

ABSTRACT

Background: Primary breast angiosarcomas are very rare type malignant breast tumor. It may have an insidious clinical onset, presenting as a painless, often discrete palpable mass that grows rapidly. The ranged age of disease is 20-50 years.

Case description: We report a case of a young woman with palpable and painless mass of her left breast. Based on radiological findings with mammography and MRI examination reported as malignant mass of the left breast suspected angiosarcoma. Pathological confirmation post-surgery reported a well-differentiated angiosarcoma that arose primarily in the left breast. Patient treated with simple mastectomy followed by whole breast radiotherapy. External beam radiotherapy delivered using 3DRT with FIF technique, prescribed dose 60 Gy in 30 fractions within 6 weeks.

Conclusion: There is no established standard treatment for primary angiosarcoma of the breast. Mastectomy remains the mainstay of treatment. Adjuvant radiotherapy appears to improve local control.

Keywords: breast, angiosarcoma, adjuvant radiotherapy.


INTRODUCTION

Soft tissue sarcomas of the breast is an infrequent site.1 Angiosarcoma affecting the breast is divided into 2 main categories: primary angiosarcoma of the breast and cutaneous angiosarcoma associated with prior radiation therapy. Primary angiosarcoma of the breast is extremely rare and is observed in less than 0.05% of primary breast tumor cases.2,3 These angiosarcomas usually occur in young and premenopausal women, aged 20-50 years.3,5

Angiosarcoma may have an insidious clinical onset, usually presenting as a painless often discrete palpable mass that grows rapidly.4 The typical patient with primary breast angiosarcoma is a young woman with dense breast parenchyma.6 Nascimento et al. indicate higher prevalence of primary angiosarcoma in the right mamma.3 Primary angiosarcoma of the breast tends to metastasize hematogenous to lungs, liver, bones, skin and the cuteralateral breast (from the other breast).4,7

The prognosis of the angiosarcoma of the breast is generally poor and based primarily on tumor size, histologic type of sarcoma, tumor grade, completeness of surgery/the resection margin status, and performance status. However, the factors related to the prognosis of primary breast angiosarcoma have still controversial.2,6

Here we report a case of primary angiosarcoma of the breast received radiation therapy.

CASE REPORT

A 21-year-old female presented with a painful rapidly growing mass in her left breast for a month. The mass was associated with dull, continuous pain, which was not relieved on medications and was not preceded by a history of trauma. Physical examination revealed an irregular, tender mass measuring 15 x 10 cm almost the entire left breast. The mass was firm in consistency, not fixed to the overlying skin or underlying chest wall. Nipple retraction, discharge, or axillary node enlargement are generally absent. No axillary lymph nodes were palpable. She had no personal or family history of breast or ovarian cancer, nor any history of prior radiation exposure and breast surgery.

Mammograph showed dense breast, developing density with bigger sized of left breast is suspicious for malignancy, BIRAD-IV. Furthermore, breast MRI presented a large parenchymal mass of the left breast with multiple nodular cutaneous-intraparenchymal foci predominantly at lateral quadrant, with the largest nodule seen in the other quadrant size ± 2.4 x 2.7 x 1.7 cm. After contrast administration showed inhomogeneous contrast enhancement with various early and prolonged nodular enhancement patterns. There were no distant metastasis and nodal involvement detected from FDG of PET-CT. No sign of malignancy in right breast.

An incisional biopsy showed well-differentiated angiosarcoma. She underwent a simple mastectomy with histology showed a large hemorrhagic tumor measuring +/- 11 x 13.3 x 4.8 cm and diagnosed as a well-differentiated angiosarcoma. As surgical result showed positive margin, the patient was advised to undergo adjuvant chemotherapy but was not keen because of the attending side effects. Thereafter, she received adjuvant radiation therapy.

She had whole breast radiotherapy using 3D conformal radiotherapy technique with FIF, prescribed dose 60 Gy in 30 fractions in 6 weeks. The fractionation scheme is conventional fractionation. During the radiotherapy...
course, pigmentation change and grade two acute skin reactions Radiation therapy Oncology Group (RTOG) were present by the end of treatment. The reactions resolved with supportive medical care at 2 weeks after radiotherapy completion.

**Patient positioning, image acquisition and image co-registration**

The patient was immobilized with breast board in supine position, both arms elevated above the head. A CT simulation was acquired on with 2.25 mm slice intervals. CT images were obtained of the area extending from the underside of the chin to the upper abdomen, including bilateral lungs and heart without intravenous contrast. After CT simulation, DICOM (Digital Imaging and Communications in Medicine) images were transferred to Monaco Treatment Planning for treatment planning. All pretreatment imaging data was imported into the treatment planning system and rigidly registered with the CT simulation dataset.

Whole breast irradiation was planned by Field in Field technique (FIF) using 6MV photons beam, on Monaco treatment planning system (Electa Medical Systems). An anisotropic analytical algorithm with 1.25 cm grid size was used for dose calculation for all plans. The isocentre was placed exactly at the centre of target volume. Two tangential open beams and multiple sub fields were placed in the same beam direction. The open beam was extended to 1.5 cm anterior chest region to create the flash margin in air. Further beam placement in anterior and posterior oblique direction was avoided, which helped in optimal target coverage to breast tissue and minimal dose to organ at risk. The gantry angle for right side breast was 300°±5° for medial tangent field and 120°±5° for lateral tangent field. Treatment was delivered on Sinergy (Electa Medical Systems) with high definition 120 leaf multi-leaf collimator. Daily imaging verification prior to delivery was performed to ensure accurate delivery. Translational errors exceeding 1mm were corrected on a four-degree of freedom treatment couch.

![Image 1](image.png) **Figure 1.** Radiation Planning by Field in Field technique treatment planning images. Axial and coronal dose colour wash (with lower limit set to 95% of 60 Gy) demonstrating adequate coverage of the planning target volume. Beam Eye View Radiation Planning by 130° and 305°.

**DISCUSSION**

Primary breast angiosarcoma is a rare tumor tends to occur at young premenopausal women, between 20 and 50 years, with median age is 43 years. The age at diagnosis of our case is 21 years. This finding correlates to primary angiosarcoma, which occurs in younger patients than secondary angiosarcoma. Primary breast angiosarcoma often presents a large painless mass, rapidly growing, even though some authors reported painful mass.

The radiologic initial finding of primary breast angiosarcoma might exhibit nonspecific result. They often appear as well to ill-defined border mass, hypoechogenic or mixed echogenicity on ultrasound, and high-density mass on mammography. Magnetic resonance imaging (MRI) is useful in identifying breast angiosarcoma, will show low intensity on T1W and high intensity on T2W, hypervascularity feature and heterogeneous mass.

Biopsy and surgical histology consistently showed well-differentiated angiosarcoma. The histopathological of angiosarcoma described in three main patterns as proposed by Donnell et al. 1: (a) grade I, well differentiated, tumors consist of anastomosing vascular channels that invade the surrounding breast tissue, (b) grade II, moderately differentiated, tumors have more solid neoplastic vascular growth and an increased mitotic rate, and (c) grade III, poorly differentiated, tumors have obvious sarcomatous areas, and areas of necrosis, hemorrhage, and infarction.

As angiosarcomas of the breast are very rare, there is no established standard treatment. Mastectomy is the mainstay of treatment. Although some individuals seem to benefit from chemotherapy, it is of minimal benefit for patients with disseminated disease. The gold-standard treatment in patients affected by primary angiosarcoma of the breast is surgery consisting in modified radical mastectomy combined, when necessary, with axillary node dissection. Patients with smaller tumors may benefit of breast-conserving surgery.

Adjuvant radiotherapy on this case was considered as positive surgical margin after simple mastectomy. Postoperative irradiation can be used to improve local control, especially in patients with tumors larger than 5 cm and positive or surgical margins. Microscopic tumoricidal dose (50 Gy) to the whole breast, and at least 60 Gy to the tumor bed, is usually used. A meta-analysis indicates that adjuvant radiation might improve recurrence-free survival.
The role of radiation treatment radiotherapy is limited to the residual microscopic disease after surgery of the primary tumor. The potential impact of radiotherapy on local control and survival of patients with soft tissue sarcomas of other sites has been well documented. Adjuvant radiotherapy after surgery may benefit breast sarcomas, particularly for patients with microscopically positive margins.

Rosen et al. used adjuvant radiotherapy in combination with chemotherapy in 17% of their patients and only radiation in 6% of their patients without improving disease-free survival and overall survival. In the study by Sher et al. 68% of patients received radiotherapy with a documented recurrence-free survival of 47% and 44% at 5 and 10 years. This rate shows an improvement in recurrence-free survival compared with patients who did not receive radiotherapy (33% and 25% at 5 and 10 years respectively). Even though these results are encouraging, there is not a statically significant correlation between adjuvant radiotherapy and improved survival due to the small number of included patients.

CONCLUSIONS

Primary angiosarcoma of the breast is a rare malignancy that is best managed with a surgery as first approach, with due importance given to the patient’s perspective on their disease and choices for adjuvant treatment. Due to paucity of data, decisions in addition to adjuvant radiotherapy need to be made in the multidisciplinary clinic.

The probability of controlling local primary angiosarcoma breast with surgery and adjuvant radiotherapy is high. In our case patient was treated with surgery and radiotherapy remained locally controlled after treatment.

Adjuvant radiotherapy in patients with primary angiosarcoma breast that have been resected with positive margins and in whom salvage surgery would likely be ineffective. Radiotherapy alone for gross disease is employed only by default if the disease is unresectable. Indications for radiotherapy include close or positive margins, multiple positive nodes, extracapsular invasion, and perineural invasion. Radiotherapy doses range from 50 to 60 Gy using once-daily fractionation.

There is little data in the literature on the efficacy of radiotherapy alone. It appears that radiotherapy alone probably controls approximately half of those treated. Radiotherapy is to be considered, more data are needed to understand its effectiveness. It is important to keep the patient under long term follow-up.

CONFLICT OF INTEREST

All author declares there is no conflict of interest regarding publication of current report.

FUNDING

None.

ETHICAL CONSIDERATION

The patient had received signed written informed consent regarding publication of medical data in medical journals with confidentiality towards personal information.

AUTHOR CONTRIBUTION

All authors had contributed in manuscript writing and agreed for the final version of manuscript for publication.

REFERENCES


This work is licensed under a Creative Commons Attribution License