Secondary breast angiosarcoma: A case report and Review of literature

Abstract

Angiosarcoma a very aggressive tumor of the vascular endothelium with a different clinical presentation that have been shown between primary and secondary patterns, but both represente a diagnostics challenge because of the nonspecific imaging finding in disease.

Secondary angiosarcoma which is still a rare neoplasm presented in female patient as a palpable unilateral mass after long term exposure to radiation therapy for breast cancer, final diagnosis needs a pathological report which is obtained post-surgical that's why preoperative diagnosis by using imaging modalities and biopsy is important to avoid misdiagnosis to this rare neoplasm.

We reported a special case of 61 female patient with secondary angiosarcoma arises near the scar of previous breast surgery due to invasive ductal carcinoma, patient was exposed to radiation therapy, chemotherapy and have positive family history.

Keywords

Angiosarcoma, Breast cancer, secondary breast angiosarcoma , prognosis, case report, breast, angiosarcoma.

Introduction

Worldwide, breast cancer is the most common cancer and the leading cause of cancer-related deaths in women.cancers caused by genetic mutations as BRACA1 and BRACA2 are the minority of cases,whileThe majority of breast cancers occur occasionally and are linked to several risk factors, such as femalegender, olderage, family history, radiation exposure and exposure to exogenous hormones.

Despite increasing incidence of breast cancer, survival rates have been increasing due to increased awareness about screening and prevention, developed intervention strategies as surgical methods, in addition to adjuvant therapy.

Breast malignancies have many types that differ in pattern of presentation, histopathological characteristics and management. Less than 1% of all breast malignancy is sarcoma which divided into three subgroup, malignant phyllodes tumor , primary and secondary sarcoma.[1]

Among the uncommon breast conditions is angiosarcoma, a rare highly malignant soft tissue tumor originating from the lining endothelial cells of vesicular or lymphatic channels . [1], [2], divided into denovo from the mesenchymal tissue as primary neoplasm with unknown predisposing factors or secondary (radiation – induced).

Breast angiosarcoma account for 0.04% of all breast malignancies . [3] it affects around 0.5% of patients undergone radiotherapy following breast conservation therapy (Stewart Treves Syndrome) as a part of breast cancer management, which commonly occur at mean onset of 10-years. [4]

The purpose of reporting this new case of angiosarcoma was to focus on the impact of prior exposure to radiation therapy ;to describe the clinico pathological features of secondary angiosarcoma.

Case presentation

A 61-year-old female patient brought to the clinic with a complaint about new modifications at

right breast, after we took a full history, did a physical examination we noticed that there was a reddish-brown lesion attached to the ulcerated skin with diffuse skin thickening in the upper inner quadrant of the right breast away from a previous scar 2.0 cm and the lesion was gradually increasing in size, the left breast was normal.

She-had previously invasive ductal carcinoma at the same side that was diagnosed by fine needles aspiration (ERpositive , PR negative , HER2/neu positive, N0 , T2 , , grade II)

AJCC (T2N0Mx). operated 4 years before presentation with wide local excision and axillary dissection followed by 4 cycles of chemotherapy and 20 cycles of radiotherapy.

DISCUSSION

On right side consist of an ellipse of skin measuring 20 *12 cm with underlying breast tissue measuring 25*17 cm, bearing retracted nipple measuring 0.7*0.7 cm, there is skin lesions dark brown in color irregular border measuring 2.7*2.7 cm located in upper inner quadrant, another small black lesion measuring 0.2*0.2 cm away from the previous lesion 1.0 cm.

A biopsy was taken, showing a high-grade tumor measuring 4 cm in greatest dimension involving skin, underlying breast parenchyma as well as the nipple in deep dermal area, it consists of solid areas and anastomosing vascular channels lined by endothelial cells with atypia,also, frequent mitotic figure, necrosis and blood lakes was identified. On immunostaining the neoplastic cells are positive for ERG and negative for PanCK.

The patient agreed to the risks of surgery after they were explained to her and expressed a desire to have it done as soon as possible.sheunderwent a right simple mastectomy in August2023. In the mastectomy specimen the tumor was ill defined with a focal area of necrosis and hemorrhage.

Post operative pathological examination:

Tumor was high grade, showing marked endothelial cell atypia, endothelial tufting present, focal papillary formation, minimal spindle cell Foci, numerous mitotic figures, blood lakes and necrosis also seen.

By immunohistochemistry the tumor cell is positive for factor VIII which supporting the diagnosis of secondary angiosarcoma. All surgical margins and skin margins free from tumor, breast tissue showing fibrosis and cystic changes.

Review of literature

In recent years, patients with secondary breast angiosarcoma increase in order to increase the quantity of patients receiving adjuvant radiation and breast-conserving surgery.[5]butThe risk of developing angiosarcoma after radiation therapy doesn't exceed the benefit of use radiation in treatment .[6] . there are some factors that may increase the risk of secondary breast angiosarcoma as Patients with previous breast cancer have higher risk to develop angiosarcoma at the same side of the breast .[7] also A previous study found that There is a connection between BRCA1 &BRCA2 mutations and breast angiosarcoma [5] , as we know that BRCA1 &BRCA2 genes involved in protection against radiation induced DNA damage .[6]

Clinical presentation:

Primary and secondary angiosarcoma are clinically and pathologically different. Secondary angiosarcoma present in older women with a mean age of 60 with skin discoloration,ulceration, erythematous area, Edema and cutaneous changes in opposite to primary angiosarcoma which affect young age and present as palpable mass with rapid growth pattern.

[5] , [6] , [3] .

A multi centre retrospective study over a period of 25 years on patients with primary and secondary angiosarcoma found that ;71.4% of secondary breast angiosarcoma patients presented with cutaneous mass , 90.9% of Primary breast angiosarcoma presented with parenchymal mass .[8]

Imaging and diagnosis:

Angiosarcomas have a nonspecific imaging finding , It has a variable appearance on mammogram it may appear irregular or well circumscribed ,while in one-third of cases of primary angiosarcoma mammogram was completely normal especially in low grade tumor [3] , [9] . in ultrasound it has nonspecific features with different ecogenesities, [3]. MRI appear to be the best modality(or preferred) to discriminate angiosarcoma from normal breast tissue weather low or high grade tumor . [9] So

MRI is useful in secondary angiosarcoma but Despite a high sensitivity of MRI (92%) It can give a false negative result so biopsy confirmation is necessary. [10] .

Confirming diagnosis of angiosarcoma is challenging; because in histopathological description after biopsy it may misdiagnosed with benign lesion as hemangioma, this is attributed to that both lesions on histopathology composed of thin-walled capillaries lined by cytology bland\low-nuclear grade endothelial cells [9]

A study revealed that 93% was the sensitivity of incisional biopsy to detect radiation associated angiosarcoma and 84% for punch biopsy compared with the sensitivity in fine-needle aspiration cytology which was (0%) and core needle biopsy was (18%).[11]

Treatment and prognosis:

The best surgical treatment of angiosarcoma is still unclear or (depends on patient situation) either to do total mastectomy or mastectomy with axillary dissection (despite uncommon nodal metastasis)or local excision for cases who already undergone previous mastectomy.[10] .

The benefit of adjuvant therapy after surgical treatment differ according to patient health status as well as type of chemotherapeutic agent , Certain studies indicate improvements in overall survival as well as disease-free survival. [12] , [13] ,[14]

While other studies revealed that there is no effect of chemotherapy on recurrence or survival [15] . for radiotherapy the aim is to improve both regional control after surgery and survival .some studies show improvement post radiotherapy [16] , while other study show no correlation [15] .

we have to take into account that the decision of starting radiotherapy is limited especially in patients with secondary angiosarcoma who had already received the maximum dose of radiotherapy [17].

prognosis of angiosarcoma depends highly on tumor grade, tumor size and margin status. Compared to primary type ,secondary angiosarcoma have poor prognosis ,low survival and high recurrence rate [10] .

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