World Journal of Medical and Surgical Case Reports

Case Report

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Primary Leiomyosarcoma of the Breast with Axillary Nodal Metastasis : A Case Report and Review of Literature

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Introduction

Primary breast sarcomas are rare tumors. They originate from the mesenchymal tissue of the breast. The prevalence of primary breast sarcomas as reported at Mayo Clinic among breast cancers was found to be 0.0006% [1]. Among sarcomas of breast primary leiomyosarcoma is even more rarely seen and less than 50 cases have been reported in the world literature. Most of the reported cases are between the age of fifty and eighty years and occurs usually in postmenopausal women. Most cases are low-grade and are cured by complete excision with wide margins. In the present study, we present a case of primary leiomyosarcoma of the breast which had axillary metastasis, and the first such case reported in literature and we discuss optimal treatment options.

Case Report

A 40-year-old female patient was referred to our hospital because of a big ulcerated mass in the breast (figure 1). She noticed a lump in her right breast 1 year ago and was on some ayurvedic medicine, but recently this lump had grown rapidly and led to the ulceration of the skin . Physical examination revealed 25/25 cm ulcerated lesion in the breast. There was no retraction of the nipple but had palpable axillary nodes 2×2 cm. The contralateral breast was normal. Mammography detected a large mass (BIRADS 5).

Core needle biopsy of the right breast revealed a spindle-cell neoplasm composed of tumor cells with blunt ended nuclei that were strongly positive for smooth muscle actin (SMA) and vimentin, and lacked expression of pan-cytokeratin, CD34, and S-100. This immunophenotype is most consistent with a diagnosis of breast sarcoma.

Microscopically, the tumor was composed of pleomorphic spindle cells showing frequent mitoses and necrosis. Tumor cells demonstrated diffuse immunohistochemical staining with smooth muscle actin (SMA). No staining was observed with desmin, S100, CD117. According to the histopathological and immunohistochemical analysis, the tumor was diagnosed as leiomyosarcoma. FNAC of the axillary nodes showed metastatic sarcoma. Modified radicalmastectomy was done as the tumor was large and had axillary metastasis. The patient is doing well and on regular follow up.

A follow-up thoracoabdominal computed tomography scan and bone scintigraphy

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Submitted: Thursday, April 14, 2016; Accepted: Friday, August 19, 2016; Published Sunday, August 21, 2016

Figure 1: Clinical picture of Leimyosarcoma Breast

performed 1 month after the operation were normal.

Discussion

Primary leiomyosarcoma of the breast is an extremely rare malignant neoplasm of uncertain biological behavior. There are less than 50 well-documented cases reported in the English medical literature [1]. As this tumor is rare more patient data should be presented for the development of a proper treatment strategy.

The majority of these cases present as a well-circumscribed mass in the breast of postmenopausal women. The exact origin of the entity is not clear. The myofibroblasts in the nipple areola complex have been proposed as the origin for the neoplasm [2]. The mainstay treatment is wide margin local excision. Most reported cases have undergone mastectomy with a few exceptions being treated with lumpectomy. Axillary dissection is believed to be unnecessary as the primary leiomyosaroma of the breast does not spread through the lymphatic route, but as in our case it had metastasized to axillary nodes MRM was necessary and ours is first such case reported in literature.

To date, this type of tumor has been observed to affect middle-aged women and manifests as a mass in their breast over a long period of time. The average tumor size is 4.7 cm [3, 4]. The involvement of endocrine factors in this tumor appears to be unlikely as the two reported cases were in males [3]. This tumor tends to show local recurrence and distant metastasis has been observed in 25% of patients [5, 6]. Metastatic spreading usually occurs via the hematogenous route. Tumor size is believed to have no significant correlation with metastasis [5]. The studies published to date show that the tumor presents itself as a long-standing and slow-growing mass. The duration of symptoms varies between 2 weeks and 5 years, confirming those characteristics [5]. Local recurrence and distant metastases can arise even after 15-20 years [7]. Chen *et al.*, [8] and Nielsen [9] lost their patients 16 and 20 years, respectively, after the operation due to hepatic and multiple metastases.

Leiomyosarcoma is characterized by spindle-shaped cells with pleomorphic, hyperchromatic and elongated nuclei; eosinophilic cytoplasm; large nucleoli; and significant mitoses. Definitive diagnosis is established through histological examination, in which positive staining is observed immunohistochemically with desmin, vimentin, and muscle-specific actin, whereas negative staining is seen with cytokeratin, myoglobin, and S-100 [10]. Leiomyosarcoma are generally strongly and uniformly positive for SMA and HHF35 (Figure 2). Smooth muscle actin is more specific for smooth muscle than panactin HHF35. It is usually negative in skeletal muscle tumors in contrast to HHF35, which is frequently positive in these tumors. Desmin positivity varies with leiomyoma being usually at least focally positive, whereas LMS is positive in only 70% to 80% of cases, with less staining in the poorly differentiated examples. Calponin, a cytoskeleton-associated actin-binding protein, is frequently used as a myoepithelial marker but is also very useful in detecting smooth muscle differentiation in STTs. In addition to smooth muscle and myoepithelial cells, it is also expressed in myofibroblasts. It is consistently positive in leiomyoma, and it is

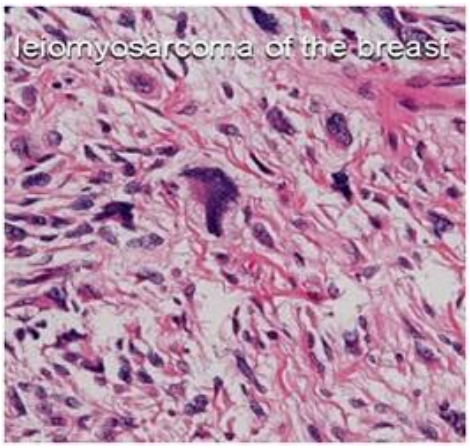

Figure 2: Photomicrograph showing tumor composed of spindle cell: leiomyoma

positive in a high percentage of LMSs (90% in conventional LMS, 70% in pleomorphic LMS). To define a poorly differentiated spindle cell sarcoma as LMS, at least 2 of 3 muscle markers (using for example SMA/desmin/HHF35 or SMA/desmin/calponin) should be positive, and the H&E appearance should be supportive as well. Markers specific for skeletal muscle differentiation (myoglobin, myogenin) are consistently negative in LMS. Keratins and EMA are positive in 10% to 30% of LMSs [11].

Because leiomyosarcoma often invades peripheral tissues, such as the skin and fascia, curative surgery requires a wide resection. Because there has been no reported cases of lymph node metastasis in which dissemination occurred via the hematogenous route, axillary lymph node dissection is unnecessary if leiomyosarcoma diagnosis can be achieved before the operation [10], but our case is the first to show axillary nodal metastasis and thus importance of preoperative FNAC for any palpable axillary lymphadenopathy. Prognosis is determined primarily by the adequacy of surgical resection. Although, there is no definite consensus on the use of adjuvant chemotherapy or radiotherapy, most patients reported till date have done well without any chemotherapy or radiotherapy, at least in the initial few years(Table1) . In the present case, there was axillary lymph node involvement. MRM was performed due to large ulcerated mass with invasion of the pectoralis major muscle by the tumor. The prognosis of leiomyosarcoma is better than that of other breast sarcomas [5]. The prognostic factors are not fully known because of the limited number of studies.

They require long-term follow-up because local recurrence and distant metastasis can occur long after the operation. After surgical resection, late local recurrence and distant hematogenous metastasis to lungs and liver is, however, well-documented and bone metastasis has once been reported [12]. It has a better prognosis than other breast sarcomas. However, there is a need for further studies to determine the prognostic factors.

Conclusion

Leiomyosarcoma of the breast is a rare entity with patients typically being in the 5th−7th decade. Morphologically it can be suspected by the typical histological features of circumscription, high cellularity and being composed of fusiform spindle cells having blunt end nuclei. Confirmation by an immunohistochemical profile of smooth muscle actin, vimentin, and desmin positivity is helpful. The mainstay treatment is wide margin local excision. Axillary dissection is believed to be unnecessary as the primary leiomyosaroma of the breast does not spread through the lymphatic route.

Author's Contribution

SN: Collection of data and involved in editing the article.

GP: Helped in finding the previous research material for the article.

HQ: involved in collection of data and final editing of the article.

BBP: Helped in analyzing the histopathological and Immunohistochemistry.

Conflict of Interest

The authors declare that there are no conflict of interests.

Ethical Considerations

The written informed consent was obtained from the patient for publication of this case report. The copy of the consent is available with the authors.

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Table 1 Comparison of clinicopathological variables of all the cases of primary leiomyosarcoma of breast reported in the English literature.

Author	Year	Age/Sex	size (cm)	Mitosis (/10hpf)	Treatment	Ct/Rt	Final follow up
Haagensen	1971	77/F	8	Very frequent	SM	-	Alive, 14 yrs
PardoMindan et al.	1974	49/F	7	16	SM	-	Alive, 6 months
Barnes and Pietruszka	1977	55/F	3	10	SM	CT	Died 4 yrs later
Hernandez	1978	53/M	4	15	MRM	-	Alive 1 yr
Chen et al.	1981	59/F	5.6	3	SM	CT	Alive 15yrs
Callery et al.	1984	56/F	2	-	SM	-	Alive 39 months
Callery et al.	1984	54/F	3	-	SM	-	Alive 53 months
Yatsuka et al.	1984	56/F	1.5	21	RM	-	Alive 4 yrs
Gobardhan	1984	50/F	9	5	MRM	-	Alive 2 yrs
Nielsen	1984	24/F	1.5	2	WLE	-	Died 20yrs later
Yamashina	1987	62/F	2.5	11	SM	-	Alive 2yrs
Arista-Nasr et	1989	50/F	4.5/2.3	4	WLE	-	Alive 6yrs
Parham et al.	1992	52/F	3	29	SM	RT	Alive 6 months
Lonsdale and Widdison	1992	60/F	2	10	SM	-	Alive 3 months
Waterworth et al	1992	58/F	4	10	WLE+AC	-	Alive 1yr
Wei et al.	1993	36/F	4	-	MRM	-	Died 14 months
Boscaino et al.	1994	56/F	2.5/4	2	WLE	-	Alive 9 yrs
Boscaino et al.	1994	45/F	1.9	2	WLE	-	Alive 40 months
Levy et al.	1995	35/F	4	2	SM	-	Alive 6 months
Falconieri et al.	1997	83/F	6	20	RM	RT	Alive 10 months
Falconieri et al.	1997	86/F	8	11	SM	-	Alive 8 months
Ugras et al.	1997	47/F	2	3	SM	-	Alive 1.5yrs
González-Palacios	1998	62/F	3	10	SM	P CT	Alive 17yrs
Gupta et al.	2000	80/F	6.5	5-8	SM+AC	-	Alive 2yrs
Székely et al.	2001	73/F	4.8	20-22	SM	-	Alive 1yr
Kusama et al.	2002	55/F	0.5	few	WLE		Alive 4yrs
Shinto et al.	2002	59/F	12	19	SM		Alive 8 months
Wei et al.	2003	52/F	4	22	WLE		Alive 3 months
Markaki et al.	2003	42/F	14	50	MRM		Alive 3yrs
Markaki et al.	2003	65/F	5.2	10	E		Alive 18 months
Liang et al.	2003	25/F	4	5	E		Alive 32 months
Adem et al.	2004	67/F	2	-	E		Died 7 months l
Adem et al.	2004	55/F	4	-	SM		Died 77 months
Jayaram et al.	2004	55/F	12	-	MRM		Local recurrence
Lee et al.	2004	44/F	3	6-12	SM		Alive 13months
Lee et al.	2004	52/F	4.5	6-12	SM		Alive 17 months
Stafyla et al.	2004	53/F	23	-	MRM		Alive 2yrs
Munitiz et al.	2004	58/F	4	14	MRM		Alive 1yr
Gupta	2006	37/F	8	15	WLE		Alive 36 months
Vu et al.	2006	-/F	23	-	SM		Alive 10 months
De la Pena and Wapnir	2008	50/F	3.2	-	SM		Alive 11 months
Wong et al.	2008	52/F	1.5	7	SM		Alive 4 days
Cobanoglu et al.	2009	64/F	3.5	12	MRM		Alive 22 months
Fujita et al.	2010	18/F	7.2	10	SM		Alive 5 yrs
Swapnil et al	2011	19/F	7	20-25	WLE		Alive 3yrs
Zulfikar et al	2012	48/F	10	-	MRM		Alive 1 month
Amaadour et al	2013	44/F	9.2/7.6		-		Died 1 month
Sokolovskayaet al	2014	58/F	15/15cm	-	MRM		Alive after 2 years
Present Case	2015	40/F	25/25cm		MRM		Alive after 1 year