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AN UNUSUAL BREAST TUMOR: LEIOMYOSARCOMA REVIEW OF THE LITERATURE

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Bu çalışma Ulusal Cerrahi Kongresi 2004'de sunulmuştur.

BACKGROUND: Primary leiomyosarcoma of the breast is a very rare type of breast tumors. Only 37 cases have been reported in the world literature.

Case Presentation: Herein we report a 40-year old lady with breast sarcoma and analyze the previous cases aiming to determine the diagnostic and therapeutic options. She admitted to the clinic with a complaint of slowly growing mass. No specific imaging findings were present. The diagnosis was reached after the examination of pathological specimen using immunohistochemistry. Simple mastectomy was done since it is the best therapeutic option to prevent local recurrence and there is no need to do axillary lymphadenectomy since lymphatic metastasis is an exception in breast leiomyosarcomas.

CONCLUSION: A long disease-free survival interval is no guarantee of a cure since local or systemic recurrences may appear 15-20 years after the primary surgery.

NADİR BİR MEME TÜMÖRÜ: LEYOMİYOSARKOM LİTERATÜR TARAMASI

ÖZET

Memenin primer leiomiyosarkomu çok nadir bir meme tümörüdür. İngilizce literaturde şu ana kadar 37 vaka gönderilmiştir. Bu yazıda meme sarkomlu 40 yaşında bir kadın hastayı sunmayı ve literatürü tarayarak tanı ve tedavide seçeneklerini ortaya koymak amaçladık. Hasta kliniğimize yavaş büyüyen bir kitle şikayeti ile başvurdu. Özellikli bir görüntüleme bulgusu saptamadık. Kesin tanı çıkarılan kitlenin patolojik ve immünohistokimyasal incelenmesi ile ortaya kondu. Hastaya simple mastektomi uygulandı çünkü lokal rekürrensi önlemek için en iyi tedavi seçeneği buydu. memenin leiomiyosarkomunda lenfatik metastaz görülmediği için aksiller diseksiyona gerek duyulmadı. Simple mastektomi meme sarkomlarında seçilecek tedavi yöntemidir. Ancak primer cerrahiden sonraki 15-20 yıl içerisinde dahi lokal veya sistemik rekürrens görülebileceğinden uzun süreli hastalıksız sağkalım kür olarak kabul edilmememlidir.

Sarcomas of the breast are rare tumors accounting for about only 1% of all breast malignancies. Leiomyosarcoma is one of the very rare type of primary breast sarcomas which constitutes only 7% of the all breast sarcomas (1). Preoperative diagnosis of leiomyosarcoma of the breast is difficult. Since these are extremely rare tumors, treatment approaches, extension of surgery and prognosis are differing in the literature. This report describes a case of primary leiomyosarcoma of the breast in a 40-year-old woman with the review of the literature.

Case report

A 40-year-old lady was admitted to the hospital with a slowly growing mass lesion on her right breast. The mass was first appeared nine months ago. The incisional biopsy was performed in another hospital two months ago and revealed a mesenchymal tumor and then referred to our clinic. The mass was located at the upper outer quadrant and retroareolar region. It was hard on palpation, lobulated and 8 x 8cm in size. A 4-cm circumareolar incision scar was present. On physical examination there was no lymphadenopathy neither on axilla nor on supraclavicular region. Mammography (MG) revealed no calcification but a well-demarcated, lobulated, phyllodes tumor with a diameter of 8 cm (Fig. 1) which was hypoechoic on ultrasonography (US). Simple mastectomy was performed in order to achieve negative surgical margins.

On pathological examination, the tumor was well definated from the surrounding structures (Fig. 2); many bundles of spindle-shaped cells were found under low magnification using hematoxylin and eosin (H&E) and many pleomorphic cells with significant mitotic activity (more than 10 mitotic figures per 10 high-power fields) were detected. Immunohistochemical study showed strong positive staining with antibodies to smooth muscle actin and vimentin which confirmed that the tumor originated from smooth muscle cells. The tumor did not stain for S-100 protein and MNF116 (Fig. 3). The final diagnosis was primary leiomyosarcoma of the breast. Staging investigations were performed and no metastatic spread was detected.

The patient was discharged uneventfully and followed with six-month interval. No local recurrence or systemic metastases has been detected for four years.

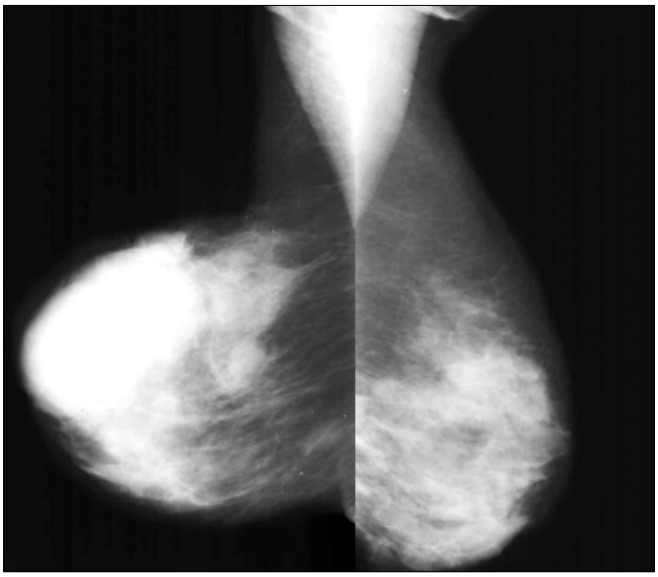

Figure 1. Mammography revealed no calcification but a well-demarcated, lobulated, phyllodes tumor with a diameter of 8cm

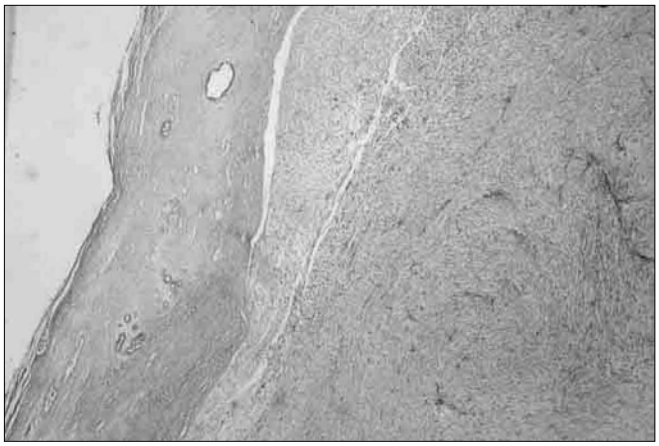

Figure 2. On hematoxylin and eosin staining of the specimen, the tumor was well delinated from the surrounding structures.

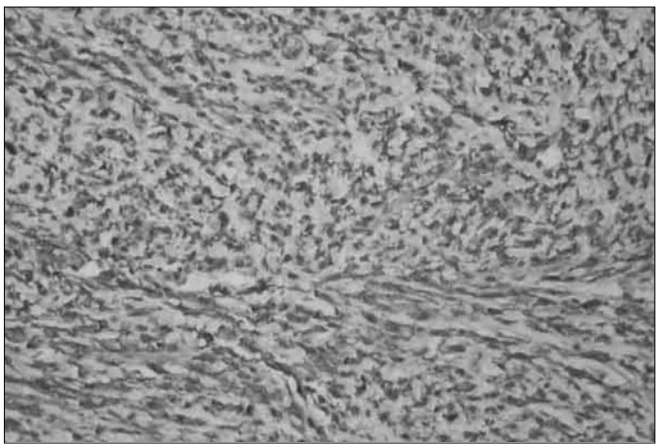

Figure 3. The tumor did not stain for S -100 protein and MNF116 on immunohistochemistry.

Discussion

Leiomyosarcoma of the breast is an extremely rare tumor and to the best of our knowledge, previously only 37 cases of primary leiomyosarcoma of the breast have been reported in the English literature excluding cases for no clear histopathologic diagnosis is available (Table 1) (2-32). The tumor originates in the smooth muscles around the nipple or in vessel and in the mesothelium of the lactiferous duct.

The most of the cases were female (only two male patients) and the median age of presentation was 56 years (range 24-86) (20). The patients usually present with a longstanding breast lump as the tumor is slowly growing. The tumor may reach large size without nipple discharge and without involvement of the skin or surrounding structures.

Definitive preoperative diagnosis of leiomyosarcoma is difficult. MG and US usually shows a dense, well-circumscribed mass which may reveal pictures similar to phyllodes tumor or fibroadenoma (28, 32) and are not associated with microcalcification. Magnetic resonance imaging was performed only in one case but did not reveal any specific findings for leiomyosarcomas (2).

Fine needle aspiration (FNA) cytology is mostly inconclusive since the aspirate is poorly cellular and a distinction from other spindle cell tumors in the breast may not be possible on cytologic specimens but FNA with immunohistochemistry may give a correct preoperative diagnosis.

Complete excision by simple mastectomy seems to be the best therapeutic option to prevent the local recurrence, since the tumor have a strong tendency towards local recurrence. Only local excisions (enucleations) are followed by a high rate of local recurrences. Although almost half of the authors did radical or modified radical mastectomy in the literature, it was unnecessary since no axillary lymphatic involvement was detected at the time of diagnosis or during follow up in the vast majority of patients. Adjuvant therapy with chemo, radio, and/or hormonotherapy (28, 32) was not effective. Almost 25% of published cases had distant metastases and metastatic disease was usually due to hematogenous route. Long term follow-up is recommended as local recurrence or metastatic disease may develop 15-20 years after the primary treatment.

Pathological diagnosis of leiomyosarcomas requires identification of features suggesting malignancy and differentiation from other breast sarcomas especially malignant phyllodes tumors and carcinosarcomas. The diagnosis of malignancy for smooth muscle cells primarily requires identification of high mitotic activity (>3 per 10 high power fields). Immunohistochemistry was positive for dem-

Table. Review of cases of leiomyosarcoma

Reference	*Sex*	*Age*	*Size (cm)*	*Treatment*	*Follow up (years)*	*Outcome*
2	F	59	12	RM	-	LR, LuM
3	M	51	5	RM	-	-
4	F	77	8	SM	14	NT
5	F	49	7	RM	6mo	NT
6	F	55	3	SM	5	Dead
7	M	53	4	RM	1.2	NT
8	F	59	5.6	SM	15	LiM
9	F	50	9	RM	2	NT
10	F	56	1.5	RM	4.7	NT
11	F	24	1.5	Exc	14	MM, dead
12	F	62	2	RM	5	NT
13	F	62	2.5	SM	2	NT
14	F	50	4.5	Exc	13	LR
15	F	58	4	Exc	1	NT
16	F	52	3	SM	4mo	LuM, BM
17	F	36	-	RM	10mo	BM, BoM
18	F	56	4	RM	6	NT
18	F	45	2.2	Exc	3.4	NT
19	F	35	3	SM	6mo	NT
20	F	83	6	RM	1	NT
20	F	86	6.5	RM	2	LR
21	F	47	2	SM	18	NT
22	F	62	3	SM	17	NT
23	F	80	6.5	RM	2	NT
24	F	59	12	RM	-	LR, LuM
25	F	55	1	Exc	3.5	LR, LuM
26	F	42	14	RM	3	NT
26	F	65	2	Exc	1.5	NT
27	F	52	1.5	Exc	3mo	NT
28	F	58	4	RM	1	NT
29	F	40	4.5	SM	-	LR
30	F	44	4.5	SM	1.5	NT
30	F	52	3.0	Exc	1.5	NT
31	F	67	2	Exc	7mo	LR,LiM,BM
31	F	55	4	SM	6	MM
32	F	48	2	RM	1.5	NT
present case	F	40	8	SM	3	NT

M: male, F: female, RM: radical mastectomy, SM: simple mastectomy, Exc: Excision, NT: No evidince of tumor, LR: local recurrence, LiM: Liver metastasis, LuM: Lung metastasis, BM: Brain metastasis, BoM: Bone metastasis, MM: multiple metastasis

sin, vimentin and muscle-specific actin and negativity for cytoceratins, myoglobin and S-100 protein.

Currently, no established prognostic criteria exist on the basis of the few cases published but in general leiomyosarcomas are tumors with a better prognosis than other breast sarcomas (16, 28).

In conclusion, leiomyosarcomas should be kept in mind in the differential diagnosis of breast tumors especially tumors having pictures similar to phyllodes tumors. The treatment of choice is simple mastectomy. There is no need for axillary lymphadenectomy. There is no certain prognostic factor, but long-term follow-up is recommended.

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