Liposarcoma of the breast - a case report

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Summary

Sarcoma of the breast constitutes less than 1% of all malignant breast tumors and liposarcoma of the breast has an incidence of 0.3% of all the mammary sarcomas. A 90-year-old woman presented with a mass in the upper outer quadrant of the right breast measuring 25 x 15 x 7 cm. Mammography was performed and the mass was diagnosed as a liposarcoma. A wide excision was performed with a 2 cm margin of healthy tissue. The tumor was diagnosed histologically as a fibrous liposarcoma. The patient was discharged and her postoperative recovery was uneventful. We report a case of liposarcoma of the breast and discuss this rare malignant tumor together with the various diagnostic and therapeutic modalities used.

Key words: Soft tissue tumor; Primary sarcoma-liposarcoma-breast.

Introduction

Primary sarcomas of the breast are rare malignant tumors arising from the mesenchymal tissue of the mammary gland [1] with an approximate incidence of 17 new cases per million women. Liposarcoma is highly uncommon in the breast. Mammary liposarcoma can arise as a denovo neoplasm or as a malignant component within cystosarcoma phylloides. The former is extremely rare with a reported incidence of 0.3% of all mammary sarcomas [2]. We report this case because of its rarity and also to throw light on the diagnostic and therapeutic modalities being used.

Case report

A 90-year-female patient presented with a painless lump in the upper outer quadrant of the right breast in September 2004. The patient was multiparous and had no family history of breast carcinoma. She bore a scar mark of two previous excisions done for recurrent swellings three and five years before. The diagnosis of previous swellings had been documented as recurrent lipoma. On examination a smooth, mobile, non-tender, soft to firm lump measuring 25 x 15 x 7 cm was identified in the right breast in the upper outer quadrant. An area of ulceration was identified alongside the lump. The lump was neither fixed to the skin nor muscle. The nipple and areola were normal. Axillary and supraclavicular nodes were not palpable. The left breast did not show any lump or abnormality. Abdominal examination did not reveal anything outside normal limits. A clinical diagnosis of lipoma was suspected. A bilateral mammogram was done which showed a well-defined soft tissue density lesion occupying the upper quadrant region, retroareolar and axillary tail of the right breast. Part of lesion was in the upper outer quadrant region and the retroareolar region was predominantly echogenic with hemorrhagic and necrotic areas. Part of the lesion in the axillary tail was mixed echogenic. No axillary lymphadenopathy was identified. The mass was diagnosed as liposarcoma of right breast.

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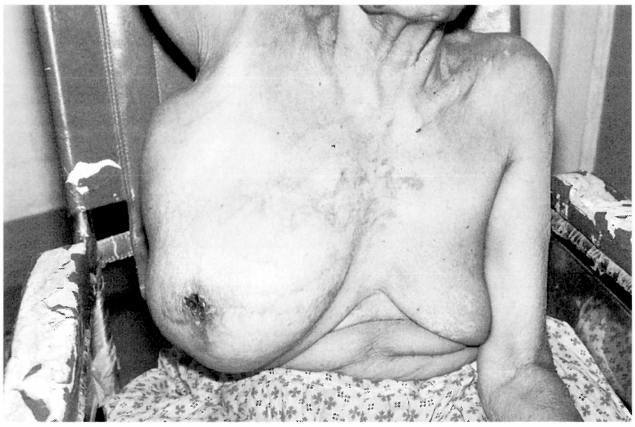
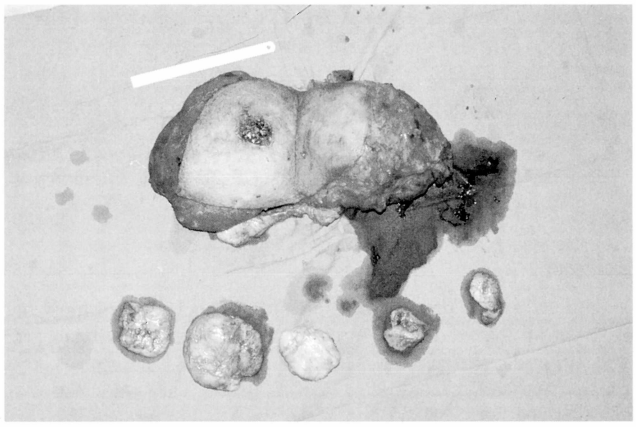
Chest X ray (posterior-anterior view of the right lateral chest wall) showed soft tissue opacity due to the right breast mass. Both lung fields were clear. Heart size, aorta and diaphragm domes did not show any lesion. Tru-cut biopsy was done from the swelling and was diagnosed as a lipoma. A wide local excision of the primary tumor with a 2 cm tumor-free margin was performed without dissection of the axillary lymph nodes. The large mass measured 28 x 17 x 8 cm. On cutting, it had a yellowish surface and showed focal areas of hemorrhage and necrosis.

Microscopically the tumor showed liposarcomatous areas and spindle cell sarcomatous areas. The liposarcomatous areas showed multivacuolated lipoblasts. The overall picture was consistent with fibrous liposarcoma, grade 2. The skin was involved by tumor but all cutaneous margins of resection and base of resection were free of tumor. The postoperative course was uneventful and the patient was discharged on postoperative day 10. At present, five months after surgery, the patient is asymptomatic.

Discussion

Sarcomas are very rare tumors of the breast representing less than 1% of all mammary tumors [3] and are a highly heterogeneous group of tumors. The majority are made up of malignant fibrous histiocytoma, fibrosarcoma, liposarcoma and less commonly angiosarcoma. Liposarcoma of the breast may arise from the periductal-perilobular stroma in the form of a cystosarcoma phyllodes tumor [4, 5] or from the interlobular stroma as a primary liposarcoma. The latter are extremely rare, with a reported incidence of 0.3% of all mammary sarcomas. Because of its rarity only one series has been described in the vast majority of reports [4]. Most reported incidents have occurred in women and there is a wide age spectrum. Most patients are women between 40 and 60 years of age, and only three cases of liposarcoma of the breast have been reported in men. Early reports of mammary liposarcoma describe bulky, multifocal tumors, often involving the entire breast [6]. There has been one reported case with consistent infiltrating ductal carcinoma and one rare bilateral tumor [2].





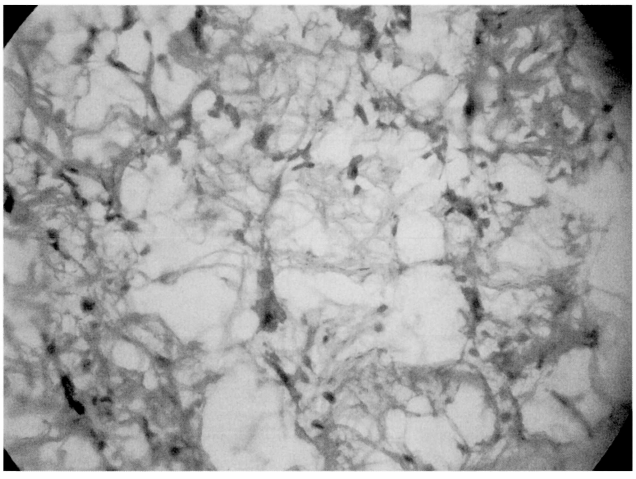


Figure 1. — Photograph of the patient demonstrating a huge lump in the right breast.

Figure 2. — Resected specimen of the main swelling and multiple smaller swellings.

Figure 3. — Microphotograph demonstrating multivacuolated lipoblasts in a background of spindle cell-fibrous liposarcoma (H & E, 400 x magnification).

Radiological investigations do not help much in framing the differential diagnosis of primary breast sarcoma but computed tomography (CT) and magnetic resonance imaging (MRI) mammography do provide an exact preoperative assessment of tumor extension. In addition to the tumor itself, the borders of the affected components (muscle fascia, muscle, cortex of the bone) and the relationship of these structures to the larger vessels and the nerves must be adequately visualized for proper diagnosis. In mammography, the radiographic features of liposarcoma largely depend on the histological type.

Enzinger and Weiss [7] histologically classified liposarcoma into four categories: well differentiated, myxoid, round cell and pleomorphic type. The well-differentiated type, like normal fat [6] or lipoma, shows radiolucency, allowing it to be clearly distinguished from the surrounding soft tissue. In contrast, the myxoid, round cell, and pleomorphic types are radiopaque. During sonography, a liposarcoma reveals a solid mass with irregular internal echoes and irregular margins. Sonography has not been used extensively in the assessment of the histological nature of the tumor. During CT, a liposarcoma displays an extremely variable pattern of enhancement ranging from homogeneous low density to marked areas within the fatty tissue [8].

The well-differentiated type is considered to be a low-grade malignancy, whereas the pleomorphic and round cell types are considered to be high grade, with a high rate of local recurrence and metastasis [7]. The myxoid types are intermediate. Therefore, determination of the histological subtype and degree of differentiation is of utmost importance for selection of proper treatment and prognosis because the clinical behavior of liposarcoma is quite variable depending on the histological type [10]. A tru-cut or an incisional biopsy should be an indication for the definitive diagnosis when a tumor rapidly grows and is not diagnosed radiologically, even if negative histological results are acquired by a fine needle aspiration biopsy.

On gross examination, the tumor is generally very large, yellowish, lobular and rubbery in most of the reported cases [6]. Lesions of the breast requiring cytological differential diagnosis include fat necrosis, lipoma with regressive changes, pleomorphic lipoma and malignant fibrous histiocytoma.

Well-differentiated liposarcoma can be distinguished by four related modifications: lipoma like, inflammatory, sclerosing and dedifferentiated types [7]. Histologically, the well-differentiated type contains irregularly shaped cells with hyperchromatic nuclei that show a slightly greater variation in size and shape than those of normal fat. The myxoid type is composed of the main tissue components: (i) lipoblasts of varying stages of differentiation (ii) a plexiform capillary pattern, and (iii) a myxoid

matrix containing heteroglycans. The round cell type is characterized by a proliferation of small, uniformly shaped round cells. The pleomorphic type has a disorderly growth pattern and an extreme degree of cellular pleomorphism, including bizarre giant cells [7].

The protocol for treatment of liposarcoma of the breast has not been established. Austin and Dupree [4] reported that complete surgical excision of the tumor and tumor free tissue margins (a 2 cm margin of healthy tissue) are required and that total mastectomy or removal of axillary lymph nodes is not necessary unless these procedures are needed for a complete excision. When metastasis of the axillary lymph nodes is found clinically, it is necessary to resect them for total excision of the tumor. In contrast, Gutman *et al.* [11] reported that axillary dissection is not included because the presence of axillary lymph node metastasis is observed as a part of systemic dissemination of the primary tumor.

Adjuvant radiotherapy of 50-60 GY may be advised when microscopic tumor is left behind or a high-grade liposarcoma, such as pleomorphic or round cell type is diagnosed [12, 13]. The prognosis is difficult to predict because of the small numbers of reported cases. The 5-year survival rate of liposarcoma in areas of the body other than the breast is 90% for well-differentiated lesions, 80% for myxoid lesions, and 20% for round cell and pleomorphic lesions. Austin and Dupree [4] reported that metastasis developed in four of 20 patients with liposarcoma of the breast, and all four patients who developed recurrence had pleomorphic liposarcoma. Well-differentiated liposarcoma is a group of tumors that may recur locally but very rarely metastasize. It is less aggressive than pleomorphic or round cell liposarcoma.

Nielsen *et al.* recently identified molecular characterization of soft tissue tumors with cDNA expression microarray and reported a group of genes associated with malignant fibrous histiocytoma, liposarcoma, and a part of leiomyosarcomas. Some of the characterized genes contributed to distinctions among the sarcomas that could be useful markers or targets for diagnosis, prognosis and treatment.

Conclusion

Liposarcoma of the breast is a rare condition. The planning of the surgical procedure is greatly facilitated by careful evalution of mammography, sonography and a definitive diagnosis of the tumor by preoperative histological examination. Based on these examinations a marginal resection should be performed to be sure that all tumor is surgically excised.

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References

[1] Adem C., Reynolds C., Ingle J.N., Nascimento A.G.: "Primary breast sarcoma: Clinicopathologic series from the Mayo Clinic and review of the literature". *Br. J. Cancer,* 2004, *91,* 237.
[2] McGregor A.: "Liposarcoma of the breast". *Can Med. Assoc. J.*, 1960, *82,* 781.
[3] Pollard S.G., Marks P.V. *et al.*: "Breast sarcoma, a clinicopathologic review of 25 cases". *Cancer,* 1991, *66,* 941.
[4] Austin R.M., Dupree W.B.: "Liposarcoma of breast: A clinicopathologic study of 20 cases". *Human Pathology,* 1986, *17,* 906.
[5] Powell C.M., Rosen P.P.: "Adipose differentiation in cystosarcoma phyllodes .A study of 14 cases". *Am. J. Surg. Pathol.,* 1994, *18,* 720.
[6] Mies C.: "Mammary sarcoma and lymphoma". In Bland, K.I. and Copeland, E.M., III (eds.). The Breast: Comprehensive Management of Benign and Malignant Disorders. Philadelphia, Saunders, 1998, 307.
[7] Enzinger F.M., Weiss S.W.: "Soft tissue tumors", 2nd edition, St. Louis, CV Mosby, 1983, 242.
[8] Jelinek J.S., Kransdorf M.J., Shmookler B.M., Aboulafia A.J., Malawer M.M.: "Liposarcoma of the extremeties: MR and CT findings in the histologic subtypes". *Radiology,* 1993, *186,* 455.
[9] Wegener O.H.: "Whole Body Computed Tomography", 2nd edition, Oxford: Blackwell Scientific, 1993, 497.
[10] Jones A.D.: "How would you manage recurrent liposarcoma of the chest wall?". *Eur. J. Surg. Oncol.,* 1995, *21,* 561.
[11] Gutman H., Pollock R.E., Ross M., Benjamin R.S., Johnston D.A., Janjan N.A. *et al.*: "Sarcomas of the breast: implications for extent of therapy". *Surgery,* 1994, *116,* 505.
[12] Arkun R., Memis A., Akalin T., Ustun E.E., Sabah D., Kandiloglu G.: "Liposarcoma of soft tissue: MRI findings with pathologic correlation". *Skeletal. Radiol.,* 1997, *26,* 167.
[13] Johnstone P.A., Pierce L.J., Merino M.J., Yang J.C., Epstein A.H., DeLancy T.F.: "Primary soft tissue sarcomas of the breast. Local-regional control with postoperative radiotherapy". *Int. J. Radiat. Oncol. Biol. Phys.*, 1993, *27,* 671.

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