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Intramuscular Atypical Lipomatous Tumor/ Well-Differentiated Liposarcoma of the Pectoralis Major Masquerading as a Breast Tumor: Management and Review of the Literature

Nikolaos S. Salemis

Breast Cancer Surgery Unit, Army General Hospital, Athens, Greece

Atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDL) of the pectoralis major muscle is an exceedingly rare clinical entity. We describe here a case of intramuscular ALT/WDL of the pectoralis major muscle in a female patient who presented with clinical manifestations of a rapidly growing breast tumor. Diagnostic evaluation and management of the patient are discussed along with a review of the relevant literature. We conclude that although the clinical examination may be inconclusive, the mammogram and especially the magnetic resonance imaging scan can precisely delineate the anatomic location and extent of the ALT/WDL of the pectoralis major muscle, thus allowing a correct preoperative diagnosis and adequate preoperative surgical planning. Complete resection is the treatment of choice for ALT/ WDL. Long-term follow-up, however, remains mandatory because of the risk of local recurrence or delayed dedifferentiation.

Key words: Atypical lipomatous tumor – Well-differentiated liposarcoma – Pectoral muscle – Breast tumor

Atypical lipomatous tumor (ALT)/well-differentiated liposarcoma (WDL) is the most common histologic subtype of liposarcoma, accounting for 40% to 45% of all aggressive adipocytic tumors.[1,2] Histologically, ALT/WDL is further divided into adipocytic, sclerosing, inflammatory, and spin-

Corresponding author: Nikolaos S. Salemis, MD, PhD, Consultant Surgical Oncologist, Director of Breast Cancer Surgery Unit, 19 Taxiarhon Street, 19014 Kapandriti, Athens, Greece.
Tel.: 302295023559; E-mail: nikos_salemis@hotmail.com

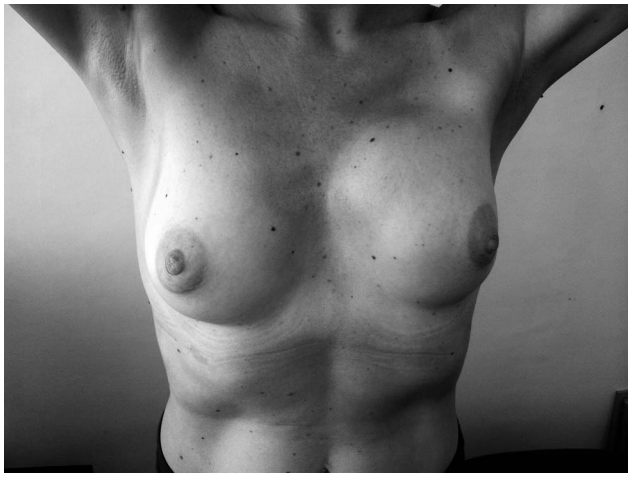

Fig. 1 Preoperative photo showing a large mass in the upper-inner quadrant of the left breast.

dle-cell subtypes.[1] The tumor predominantly affects adults in their fifth to seventh decades of life and most commonly occurs in the deep soft tissue of the extremities.[3–6] The most frequent sites include the thigh, followed by the retroperitoneum, the paratesticular area, and the mediastinum.[1] Rare locations include the subcutaneous tissues and the skin.[1] Adipocytic tumors arising from the pectoralis major muscle are uncommon and in most cases represent benign lipomas.[7–9] ALT/WDL of the pectoralis major muscle is an exceedingly rare clinical entity. We describe here a case of an intramuscular ALT/WDL of the pectoralis major muscle in a female patient who presented with clinical manifestations of a rapidly growing breast tumor. Diagnostic evaluation and management of the patient are discussed along with a review of the relevant literature.

Case Presentation

A 44-year-old premenopausal woman presented with a 2-month history of a rapidly growing painless mass in the upper inner quadrant of her left breast (Fig. 1). Her past medical history was unremarkable, and she had no family history of breast or ovarian cancer.

On clinical examination, a nontender, well-circumscribed, movable, large mass measuring approximately 12 × 10 cm was palpated in the upper-inner quadrant of the left breast. There were no skin alterations or nipple discharge, and there were no palpable axillary lymph nodes.

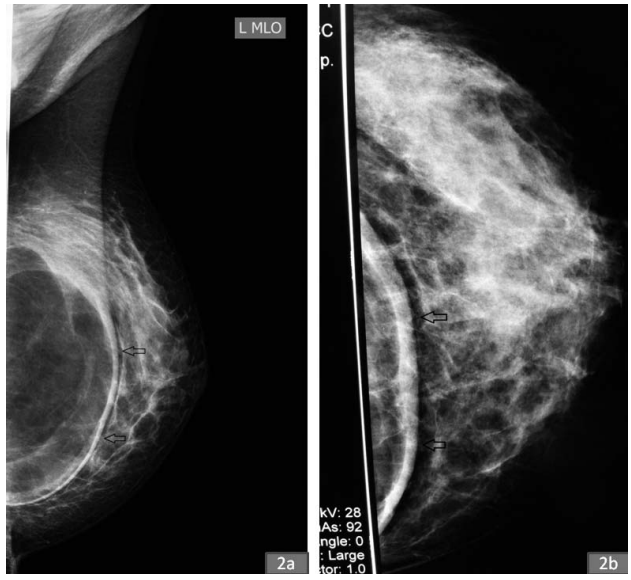

Fig. 2 Left mediolateral oblique (2a) and craniocaudal (2b) mammograms showing a radiolucent mass causing anterior displacement of the pectoralis major muscle margin (arrows).

The mammogram revealed a radiolucent mass of fat density probably originating from the pectoralis major muscle displacing its margin anteriorly (Fig. 2). On ultrasonography the mass was hypoechoic with smooth margins without internal or peripheral vascularity. Based on mammography findings, a tumor originating from the pectoralis major muscle was suspected, and a magnetic resonance imaging (MRI) scan was obtained for further evaluation and preoperative treatment planning. MRI revealed a large, well-defined, lobulated mass arising from the pectoralis major with signal intensity identical to that of the subcutaneous adipose tissue (Figs. 3 and 4). The differential diagnosis based on MRI findings was between atypical lipomatous tumor and lipoma. Fine needle aspiration cytology was suggestive of lipoma. Complete excision of a multi-lobulated yellowish mass measuring 13 × 8.5 × 4 cm was performed through an inframammary incision. Histologic findings were suggestive of an ALT/WDL (Figs. 5 and 6) with focally close posterior (thoracic wall) margins. There was no evidence of dedifferentiation. The patient received adjuvant radiotherapy. She is doing well, without any evidence of recurrence 18 months after surgery.

Discussion

Primary liposarcoma of the pectoralis major muscle is an exceedingly rare clinical entity with only a few

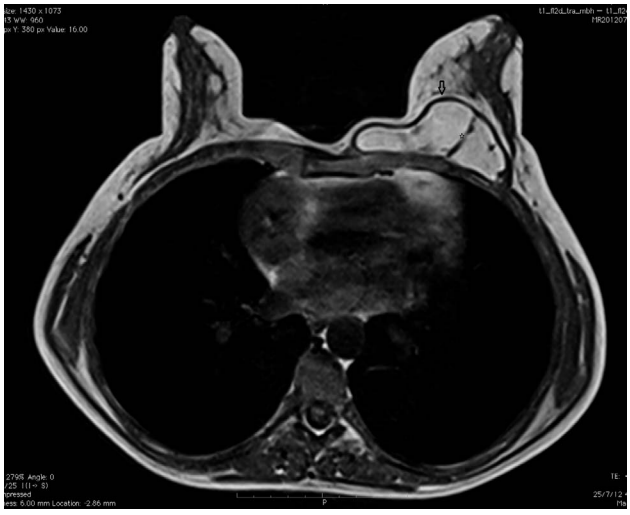

Fig. 3 Axial T1 nonenhanced magnetic resonance imaging (MRI) scan showing a multi-lobulated large mass within the pectoralis major muscle with signal intensity identical to that of the subcutaneous adipose tissue (arrow). Note the presence of thick septa (star).

cases reported in the literature so far.[10,11] However, we were unable to find any cases of ALT/WDL of the pectoralis major muscle presenting as a rapidly growing breast tumor.

ALT and WDL are synonyms describing tumors that are identical both morphologically and karyotypically and in terms of biologic behavior.[1] The use however of the term ALT or WDL is primarily

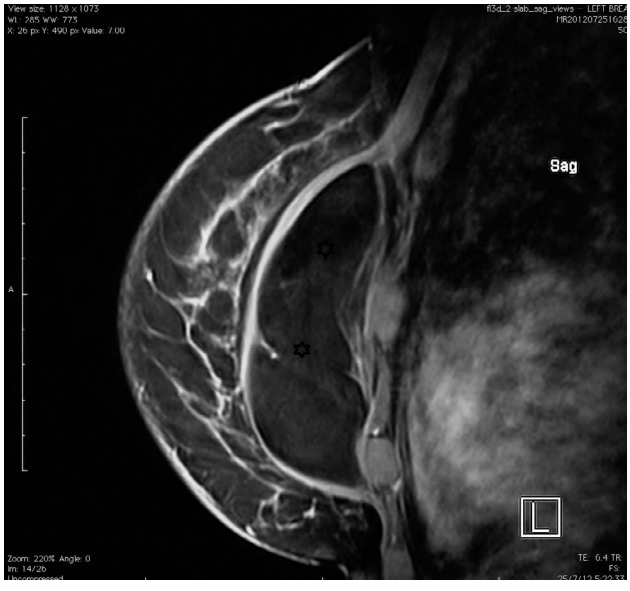

Fig. 4 Sagittal T1 contrast-enhanced MRI showing the large mass arising from the pectoralis major muscle (stars).

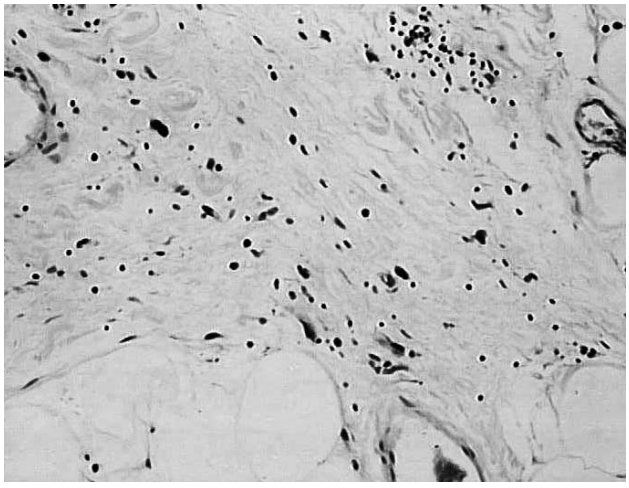

Fig. 5 Fibrous tissue septa containing variably sized adipocytes. Cellularity and mitotic figures are uncommon. (H&E ×100).

determined by tumor location and resectability.[1] The term ATL is used for tumors arising at surgically amenable locations such as the limbs and the trunk, where a radical resection is feasible and curative. On the contrary, the term WDL is reserved for tumors arising at anatomic sites such as the retroperitoneum and mediastinum, where a complete surgical resection with negative margins cannot always be achieved, thus leading to repeated local recurrences and possible fatal outcomes.[1,12] The main cause of death is the uncontrolled local effects on the surrounding structures and the dedifferentiation

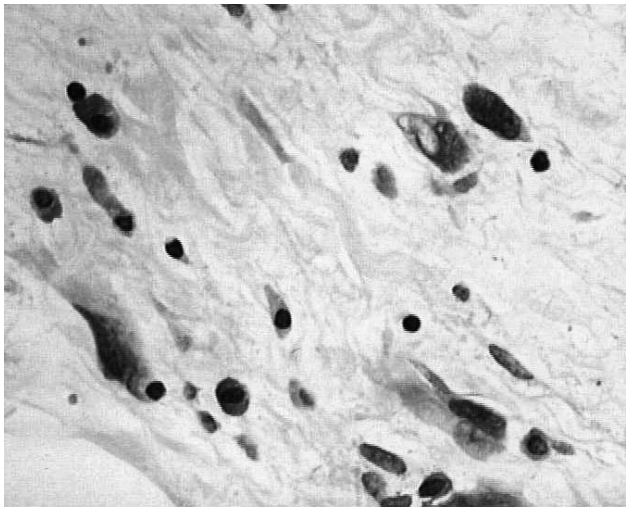

Fig. 6 Spindle cells with large, deep-staining nuclei and marked nuclear enlargement. Some nuclei have sharply outlined vacuoles. (H&E ×400).

and subsequent development of metastases.[1] The choice of terminology should be based on the principle of avoiding either inadequate or excessive treatment.[1,2]

Patients with ALT/WDL usually present with a painless slow-growing mass,[3,4,13] which can attain a size up to 20 cm in diameter, especially when originating from the retroperitoneal space.[1,14] In our patient, however, a rapid tumor enlargement was noted within a 2-month period, making this case exceedingly rare.

On gross examination, ALT/WDL is a well-circumscribed mass with a yellow to white cut surface, whereas gray to white areas indicate the presence of fibrous tissue.[15] Areas of fat necrosis have been reported in larger lesions.[1] Histologically, ALT/WDL is an intermediate malignancy characterized by mature adipocytes of varying size and scattered atypical stromal cells with hyperchromatic nuclei.[15,16] The main differentials of ATL/WDL include the benign lipoma and nonadipocytic lesions such as inflammatory myofibroblastic tumor and Castleman's disease.[2,15]

MRI has been used in differentiating ALT/WDL from lipoma with sensitivity rates ranging from 90.9% to 100%.[13,17] Brisson *et al*,[17] in a retrospective review of 87 patients with histologically proven lipomatous tumors found that factors such as patient age over 60 years, tumor dimension over 10 cm, and the presence of nonfatty areas, increased the likelihood of ALT/WDL by a factor of 2.61 to 6.25 times. The most reliable imaging discriminators of ALT/WDL were the size of the lesion and the fat content.[17] In another retrospective study, Kransdorf *et al*[14] reviewed the computed tomography (CT) and MR images of 60 patients with histologically verified fatty tumors and found that the most statistically significant radiologic predictors of malignancy were male sex, presence of thick septa, and associated nonadipose masses, which increased the likelihood of malignancy by 13-, 9-, and 32-fold respectively.[14]

However, since ALT/WDL and lipoma have overlapping MRI features, the definitive discrimination should be based on molecular pathology rather than imaging.[17] Preoperative differentiation of ALT/WDL from benign lipoma is important because of the differences in initial treatment, follow-up, and prognosis.[13]

On karyotypic analysis, ALT/WDL is characterized by the presence of supernumerary ring or giant marker chromosomes lacking α-satellite centromeric sequences.

Both ring and giant markers contain amplified genomic sequences derived from the 12-q13-15 chromosomal region including the *MDM2* gene.[1,2]

Complete surgical resection with clear margins is the standard treatment for ALT/WDL.[4,15] The most important prognostic factor is the anatomic location of the tumor.[1,6,16,18] Smith *et al*[5], studied 1266 patients with WDL with a median follow-up of 45 months and found that retroperitoneal location and increasing age were associated with worse overall and disease-specific survival. Retroperitoneal location was associated with significantly worse outcomes independent of tumor size, but nonretroperitoneal locations had statistically similar overall survival.[5] Positive resection margins and sclerosing histology have been associated with reduced local recurrence-free survival.[4,19]

Although ALT/WDLs do not metastasize, they have been associated with an increased risk of local recurrence and a low risk of late dedifferentiation.[2,3,16,18–20] Dedifferentiation of ALT/WDL has been more frequently reported in retroperitoneal tumors and usually suggests a more aggressive tumor.[6]

The risk of dedifferentiation is over 20% for retroperitoneal tumors and lower than 2% for tumors arising in the limbs.[1] Weiss and Rao,[6] studied 92 patients with ALT/WDL with no evidence of dedifferentiation at diagnosis and suggested that dedifferentiation is not a site-specific phenomenon but is more likely a time-dependent phenomenon seen in situations with a high likelihood of clinical persistence of the disease for a long period. The progression of the disease after dedifferentiation depends on a number of factors including the amount of dedifferentiation and the type of therapy.[6]

The overall mortality related to ALT/WDL ranges from 0% for tumors arising in surgically amenable soft tissues to more than 80% for retroperitoneal tumors.[15] Long-term follow-up is essential in patients with ALT/WDL because of the risk of local recurrence and delayed dedifferentiation.[3,4,13]

In conclusion, ALT/WDL of the pectoralis major muscle is an exceedingly rare clinical entity. Patients may present with clinical manifestations of a rapidly growing breast tumor. Although the clinical examination may be inconclusive, the mammogram and especially the MRI can precisely delineate the anatomic location and extent of the tumor, thus allowing a correct preoperative diagnosis and adequate preoperative surgical planning. Complete

resection is the treatment of choice, and long-term follow-up is essential.

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