



Primary Embryonal Rhabdomyosarcoma of an Adolescent Girl Breast: A Case Report

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Abstract

Rhabdomyosarcoma (RMS) of the breast is extremely rare condition. A 15-year-old girl presented to our clinic complaining of left breast mass that is 5 cm x 5 cm, immunohistochemistry approached it initially as giant fibro-adenoma but later diagnosed as high grade embryonal RMS. Diagnosis confirmed which showed markers suggestive of RMS. Patient initially underwent simple lumpectomy, but after histopathology results she underwent surgical margin revision and chemotherapy. Unfortunately, after chemotherapy she developed local recurrence. Wider local excision or mastectomy is to be considered in such cases.

Keywords: Breast; Primary sarcoma; Rhabdomyosarcoma

Introduction

Primary Rhabdomyosarcoma of the breast is a rare neoplasm. Sarcomas in general represent less than 1% of all malignant breast neoplasm [1]. This rare malignant disorder arises from connective tissues of the breast. Primary connective tissue of the breast is highly heterogeneous tumors, further subdivided into: Malignant Fibrous Histiocytoma (MFH), liposarcoma, leiomyosarcoma, fibrosarcoma, angiosarcoma, rhabdomyosarcoma, clear cell sarcoma, neurogenic sarcoma and soft tissue sarcoma [2].

Breast sarcomas can develop primary (*de novo*) or post radiation therapy and arm lymphedema. The clinical features of the breast sarcomas mimic those of carcinomas [3]. However, the clinical behavior and management is completely different. The authors' purpose in this article is to describe briefly behavior and to discuss briefly its histological features.

Case Presentation

A 15 – year-old female presented to Breast Clinic in King Salman Armed Forces Hospital in August 2015, with complain of left breast mass, which rather progressively increased in size over last 3 months. It was located at 9 O'clock position. No regional lymph nodes were palpable. On physical examination of other breast and routine examination no abnormality detected.

Ultrasonography study was done Figure 1 showed 4.5 cm x 4 cm mass with evidence of central necrosis, heterogeneous echogenicity, posterior enhancement and internal vascularity. No skin invasion, dilated ducts or signs of inflammation noted in the study. The other breast and the axilla were unremarkable.

Fine Needle Aspiration showed picture of juvenile fibroadenoma; so, it's initially approached as a case of large fibroadenoma. Surgical resection was done on August 17th, 2015 for excisional biopsy.

Grossly the tumor size was 5 cm x 7 cm, Anaplasia is present grade III out of III without lymphovascular invasion. Margins were anterior: 3 mm, superior: 4 mm, medial: 5 mm, lateral: 5 mm, posterior: 6 mm and inferior: 6 mm.

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Figure 1: Ultrasonography of the diseased side.

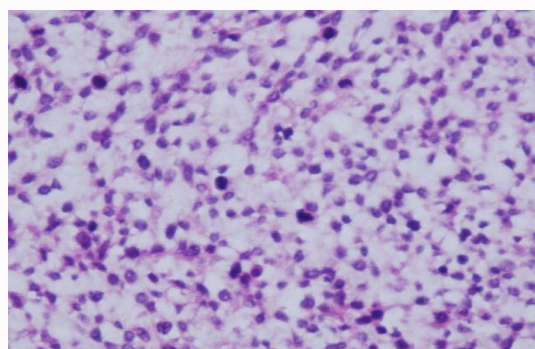


Figure 4: Round to spindle shaped cells in myxoid stroma.

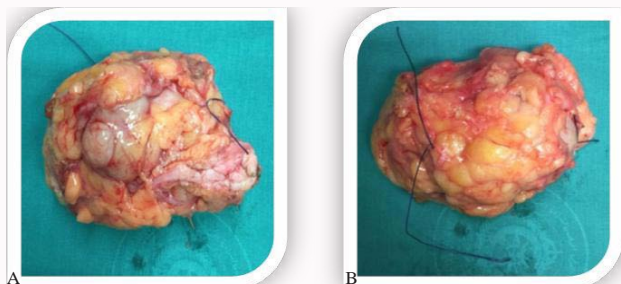


Figure 2A, 2B: Well-circumscribed excised nodular mass covered by adipose tissue.

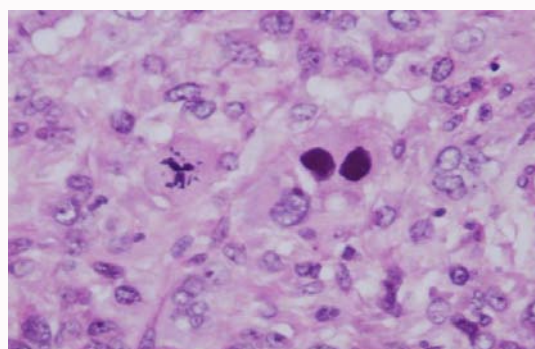


Figure 5: Marked nuclear pleomorphism and a tripolar mitosis.

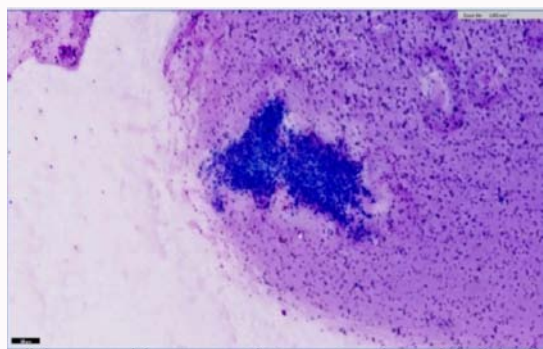


Figure 3: Hypercellular stromal fragments with impression of juvenile cellular fibroadenoma.

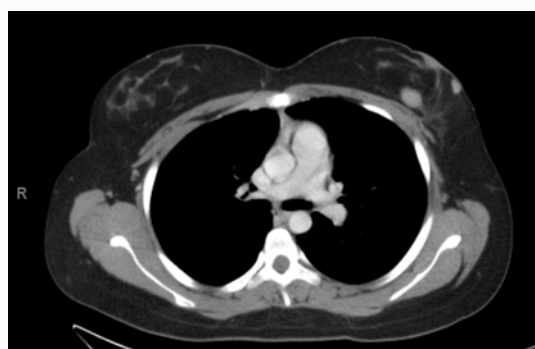


Figure 6: Postoperative staging computed tomography.

Histopathology results showed malignant tumor forming irregular fascicles and sheets Figure 2. It showed large areas of necrosis and peritheliomatous arrangement of malignant cells. The cells were round to oval to spindles shaped showing marked pleomorphism, it was embedded in myxoid stroma with high mitotic index (10/10 per HPF). Abnormal mitosis and multinucleate giant malignant cells were also frequent (Figures 3-5). Special staining (immunohistochemistry) was positive for: Desmin, Vimentin, CD99, MyoD1, Myoglobin, Muscle Specific Actin (focal).

Postoperative Staging Computed Tomography (Figures 6-10) for chest, abdomen and pelvis showed left breast mass with multiple bilateral sub-centimetric lymph nodes. No metastasis noted.

For conformation the specimen was sent to a regional reference Pathology Lab (KFSH&RC) and they agree with the diagnosis, also it was sent to Germany Bioscentia labs, and their diagnosis was undifferentiated sarcoma. She was staged T2a N0 M0 (stage 3).

Patient underwent Revision of surgical margins to achieve acceptable negative margins. She received 12 cycles of adjuvant chemotherapy (Vincristine, Actinomycin Band furosemide). Patient is followed up in Sarcoma Unit in a National Center for Oncology. Unfortunately, she developed recurrence post chemotherapy.

Follow up was done by post chemotherapy CT study which showed recurrence in operative site. No metastasis identified in the study, but incidental ovarian enlargement noted which is benign looking. She is planned for mastectomy.

Discussion

The majority of breast sarcomas have no underlying cause. The established etiology for breast sarcomas is prior radiotherapy. Predisposing factors include: genetic syndromes such as Li-Fraumeni & Gardner's syndromes & neurofibromatosis type 1. Environmental factors include use chemotherapeutic agents, arsenic compounds, vinyl chloride & immunosuppressive agents, human

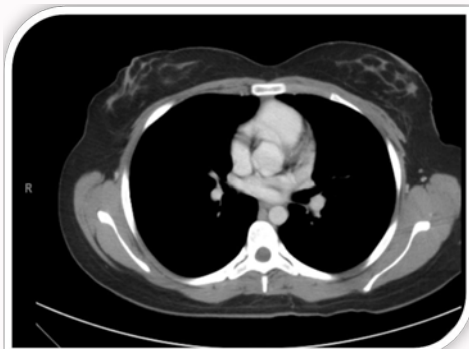


Figure 7: Left breast mass with multiple bilateral sub-centimetric lymph nodes. Urology consultation was done to exclude renal source.

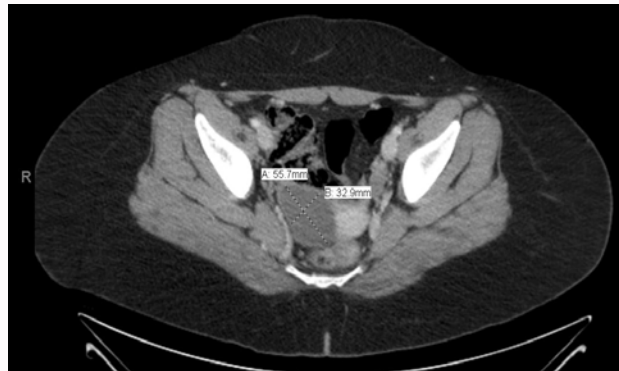


Figure 9: Incidental ovarian enlargement noted which is benign looking.



Figure 8A, 8B: New left breast two lesions in keeping with recurrent sarcoma of the breast. No evidence of metastatic disease neither to the axillary lymph nodes nor to the chest, abdomen or pelvis.



Figure 10: 5 years follow up CT showed no recurrent or metastatic disease.

immunodeficiency virus & human herpes virus type [4]. Pure non-epithelial primary malignant neoplasms of the female breast are rare, accounting for less than 1% of all breast malignancies [4]. Primary rhabdomyosarcoma of the breast in adults is extremely rare. Rhabdomyosarcomas in adults account for less than 3% of all adult primary soft-tissue sarcomas [6].

The clinical features of non epithelial breast malignancies are similar to those of breast neoplasms in many ways, but the prognosis and management of these types of tumors differ dramatically, thus it is important to distinguish between the two [4].

The origin of primary rhabdomyosarcoma of the breast is obscure and speculations include. Misplaced mesenchymal rests rhabdomyoblastic differentiation of tumors of mesenchymal origin and pluripotential cells from periductal & acinar mesenchymal cells undergoing rhabdomyoblastic differentiation have been proposed. Primary rhabdomyosarcoma of the breast accounting for less than

1% of all breast malignancies. The annual incidence of breast sarcoma is 44.8 new cases per 10 million women. The majority of breast sarcomas have no known cause. The established etiology for breast sarcomas is prior radiotherapy [4].

Primary breast sarcomas usually present as large painless breast lumps with no associated skin or nipple changes or axillary lymphadenopathy. Typically, the affected patient is a woman in her 50 s (age range 17 to 89 years), but it is also seen in men [4].

The treatment of primary breast sarcomas requires a multi-disciplinary approach. Surgery remains the mainstay of therapy. Wide local excision with negative margins is adequate. If there is local recurrence, then salvage mastectomy should be done. Axillary dissection is not required, as primary breast sarcomas do not present usually with axillary lymphadenopathy; it is required in cases of palpable lymphadenopathy, carcinosarcoma or liposarcoma. Radiotherapy is controversial and does not improve disease-free survival, but it may be useful in the treatment of high-grade lesions, larger tumors, and questionable or positive margins. Chemotherapy has no clearly defined role in primary breast or soft-tissue sarcomas [4].

In a study of 25 cases of primary breast sarcoma, it was found that tumor size very important prognostic factor. Five years overall survival and cause specific survival were both 91% for tumor <5 cm and 50% for tumors >5 cm. Mean tumor size with recurrence or metastasis was 7.7 cm, compared with 4.9 and 4.3 cm respectively for patients without recurrence or metastasis [5]. The predisposing factors for breast sarcomas are prior radiotherapy, genetic syndrome like Li-Fraumeni and Gardner's syndrome, environmental associations such as arsenic, vinyl chloride, chemotherapeutic agents, certain viruses

such as herpes and human immunodeficiency virus.

The standard therapy for breast sarcoma is mastectomy, which, according to Pollard, should include the excision of both underlying pectoral muscles, in order to reduce the recurrence rate [2]. Other approaches include a breast preserving Wide Local Excision (WLE) with adequate negative margins, particularly for small sarcomas (<5 cm). The staging or therapeutic role of routine axillary lymph node dissection has not been defined. As lymph node involvement is low, many believed that lymph node dissection is unjustified [6]. Routine lymphadenectomy does not confer a survival benefit. Rather than performing axillary sampling for these patients, a consideration may be the routine use of sentinel lymph node biopsy [7].

Conclusion

Primary RMS of the breast should be kept as a rare possibility in the differential diagnosis of female adolescent with breast mass.

This concept should be considered in absence of strong family history of invasive breast cancer with positive BRCA 1 or 2. The diagnosis & treatment is challenging & it necessitates complete clinical & histopathological correlation.

Surgery is the only potentially curable modality for primary breast sarcomas the extent of excision depends on the size of the tumour, breast size as well as histology. Regional lymph nodes spread is rare 5 or less.

As with soft tissue sarcomas arising in other areas of the body, a multidisciplinary approach involving surgical, medical & radiation oncology is preferred.

Finally, the prognosis in general depends on the stage, histologic grade & tumor size. The increased use of multimodality therapy will have a positive impact, significant & better outcome.

Recommendations

- Use of core needle biopsy for large fibroadenomas or suspicious cases.
- Surgical excision with clear margin is the gold standard way of treatment either wide local excision or mastectomy depending on the mass site, size and its proportion to the breast.

References

1. Mardi K, Gupta N. Primary pleomorphic liposarcoma of breast: A rare case report. *Indian J Pathol Microbiol.* 2011;54:124-6.
2. Pollard SG, Marks PV, Temple LN, Thompson HH. Breast sarcoma. A clinicopathologic review of 25 cases. *Cancer.* 1990;66:941-4.
3. Wiklund T, Huuhtanen R, Blomqvist C, Tukiainen E, Virolainen M, Virkkunen P, et al. The importance of a multidisciplinary group in the treatment of soft tissue sarcomas. *Eur J Cancer.* 1996;32A(2):269.
4. Chugh R, Baker L. Nonepithelial malignancies of the breast. *Oncology.* 2004;18(5):665-73.
5. Adem C, Reynolds C, Ingle JN, Nascimento AG. Primary breast sarcoma: Clinicopathologic series from the mayo clinic and review of the literature. *Br J Cancer.* 2004;91:237-41.
6. De Cesare A, Fiori E, Burza A, Ciardi A, Bononi M, Izzo L, et al. Malignant fibrous histiocytoma of the breast. Report of two cases and review of the literature. *Anticancer Res.* 2005;25:505-8.
7. Gullett NP, Rizzo M, Johnstone PA. National surgical patterns of care for primary surgery and axillary staging of phyllodes tumors. *Breast J.* 2009;15:41-4.