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Original Research Paper

Pathology

PRIMARY UNDIFFERENTIATED PLEOMORPHIC SARCOMA OF BREAST IN A 24 YEAR FEMALE : A CASE REPORT

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ABSTRACT Undifferentiated pleomorphic sarcoma constitutes less than 5 % of all sarcomas in adults and has been rarely seen in the breast and is defined as a group of pleomorphic, high-grade sarcomas in which any attempt to disclose their line of differentiation has failed . Most undifferentiated pleomorphic sarcomas(UPS) have occurred in their sixth and seventh decades of life and very rarely in adolescents and adults.

We report a case of 24 year old female presented with painless lump in right breast and rapidly increasing in size for last 5 months. Iinitial diagnosis was made as sarcoma breast on the basis of physical and radiological examination. After that total mastectomy was done without axillary lymph node dissection, as lymphatic spread is very rare. But without histopathological examination and most importantly IHC-panel, it is impossible to make a definitive diagnosis of UPS breast.

KEYWORDS : Soft tissue sarcoma, breast sarcoma, undifferentiated pleomorphic sarcoma, young female.

INTRODUCTION:

Primary breast sarcomas are extremely rare, representing less than 1% of all primary breast malignancies [1]. The most common malignant tumor of the breast is adenocarcinoma 93.7%, Sarcomas account for less than 1% of all primary breast malignancies [2]. Undifferentiated pleomorphic sarcoma previously known as malignant fibrous histiocytoma (MFH), is a mesenchymal malignancy originates from the connective tissues of glands and composed of fibroblast and histiocyte like cells, mixed with pleomorphic giant and inflammatory cells [3] that shows no definable line of differentiation [4-6]. Undifferentiated pleomorphic sarcoma constitutes less than 5 % of all sarcomas in adults and has been rarely seen in the breast and is defined as a group of pleomorphic, high-grade sarcomas in which any attempt to disclose their line of differentiation has failed [7,8]. Most undifferentiated pleomorphic sarcomas have occurred in their sixth and seventh decades of life and very rarely in adolescents and adults [9].

Breast sarcomas are commonly seen after radiotherapy. However, primary UPS has been rarely reported to involve the breast [10,11]. Owing to rarity of this entity, the exact treatment guidelines and prognostic variables are yet to be determined [12]. There are nocomprehensive reports on primary UPS of the breast, and individual case reports are rare.

CASE PRESENTATION:

A 24 year old female presented with a lump in the right breast had history of 5 months with complaint of rapidly increasing in size. Detail physical examination were performed. Patient had poorly demarcated, ill defined , non-tender, firm and mobile lump in the right breast measuring 18x12 cm in size underneath the nipple areola complex. There was no evidence of axillary lymphadenopathy. Opposite breast (left) appeared to be normal. The patient had no medical history, no family history of any breast cancer. Ultrasonography showed a low-echoic mass with irregular margins and a heterogeneous internal echo pattern. CT scan revealed a heterogenous lower density areas of haemorrhage and necrosis. All the investigations indicated a strong suspicion of malignancy and all laboratory findings, chest X ray, ultrasonography of abdomen, revealed no abnormalities. Provisional diagnosis was made as sarcoma right breast. The patient underwent total mastectomy in the oncology department without axillary lymph node dissection. Specimen has been sent to the department of Pathology, RIMS Ranchi for final definitive diagnosis.

Macroscopically breast tissue specimen appeared fleshy ill defined tumor border measuring as a whole 30 × 18 × 9.5 cm, partially covered with skin measuring 21 × 14 cm. The cut surface revealed a greyish white, fibrotic nodular tumor measuring 18x12 cm with areas of hemorrhage and necrosis(figure-1). Given specimen were routinely processed as per standered protocol to obtain tissue paraffin blocks, then sections were taken and stained by haematoxylin and eosin stain. After fixation and inclusion in paraffin, microscopic examination of the section from tumor showed oval to spindle cells, hyperchromatic nuclei with marked pleomorphism admixed with bizarre cells, multinucleate giant cells and chronic inflammatory cells with zones of necrosis and myxoid features (Figures-2) with mitotic count >10/10 hpf. The surgical base and overlying skin were free from tumor cells.

Further to confirm the diagnosis immunohistochemistry wad done which was positive for vimentin and CD68. Negative for Cytokeratin (CK) , epithelial membrane antigen (EMA), Smooth muscle actin (SMA), Desmin, CD64, CD45, S-100 protein. By correlating both histological features and immunohistochemical study, diagnosis of undifferentiated pleomorphic sarcoma was made.

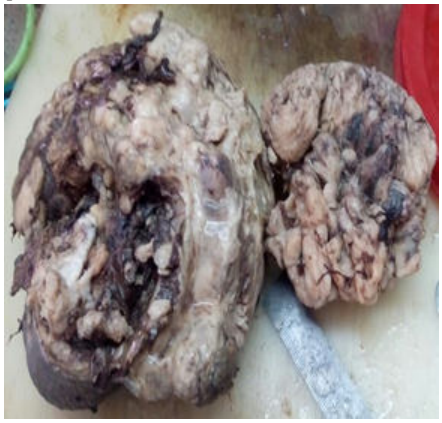

Fig. 1: Figure-1(Macroscopically breast tissue specimen shows fleshy ill defined tumor border measuring as a whole 30 × 18 × 9.5 cm)

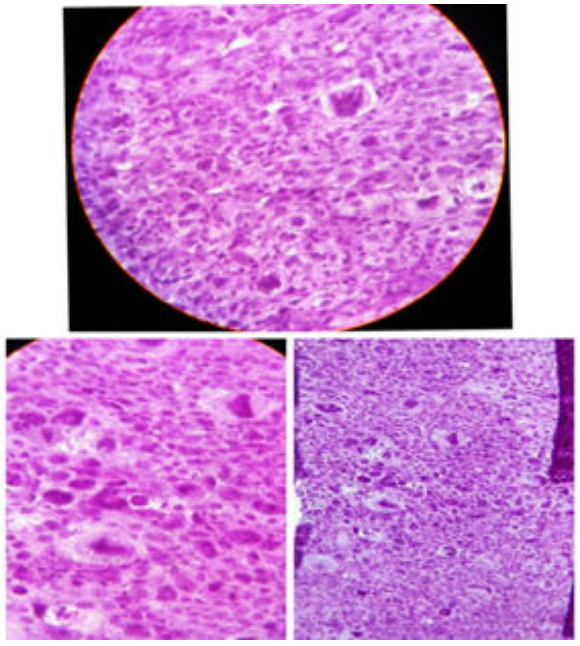

Figure-2(microscopic picture shows oval to spindle cells, hyperchromatic nuclei with marked pleomorphism admixed with bizarre cells, multinucleate giant cells.)

DISCUSSION:

Primary breast sarcomas constitute a rare group of nonepithelial tumors with aggressive behavior, originating from connective breast tissue. They account for less than 1% of all breast malignancies and less than 5% of all soft tissue sarcomas[13]. In 2013, the WHO classification of soft tissue tumors was revised and the concept of undifferentiated pleomorphic sarcoma/ malignant fibrous histiocytoma (UPS / MFH) disappeared, and a major item of undifferentiated / unclassified sarcomas (US) was created on behalf of UPS / MFH. There are 5 subtypes in US. They are undifferentiated round cell sarcoma, undifferenti-ated spindle cell sarcoma, undifferentiated pleomorphic sarcoma,undifferentiated epithelioid sarcoma, undifferentiated sarcoma, and not otherwise specified (NOS). UPS / MFH corresponds to undifferentiated pleomorphic sarcoma. Most undifferentiated pleomorphic sarcomas have occurred in their sixth and seventh decades of life and very rarely in adolescents and adults [9]. Our case is is among the rare age group i.e only 24 year old female.

Undifferentiated pleomorphic sarcomas often grow rapidly and then may be painful.

Imaging methods and macroscopy may be shown well-circumscribed masses with heterogeneous composition. Further, they can be identified as pale fibrous and fleshy areas admixed with zones of (cystic) necrosis, hemorrhage, or myxoid features [14]. Microscopically, lesions exhibit cells showing marked pleomorphism admixed with bizarre giant cells, spindle cells, and variable foamy cells [15]. A storiform growth pattern and variable chronic inflammatory cells are also common [14]. Neither the symptoms nor the physical findings of undifferentiated pleomorphic sarcoma of the breast present any characteristic pattern that would easily suggest the diagnosis. Immunohistochemistry may be useful to distinguish primary breast sarcomas from non-mesenchymal malignant tumours and to delineate the level of differentiation of primary breast sarcomas [16]. Vimentin, CD68, Desmin, Cytokeratin, leukocyte common antigen, CD34, HMB-45, SMA, EMA, and S-100 protein should be performed in sarcoma patients as it mimics many of the malignant epithelial and mesenchymal tumors.

The first documentation of UPS of breast in a young female available in literature was by Banushree et. al.in 2016[17].

In our patient although initial diagnosis was made as sarcoma breast. After that total mastectomy was done without axillary lymph node dissection, as lymphatic spread is very rare. But without histopathological examination and most importantly IHC-panel, it is impossible to make a definitive diagnosis of UPS breast.

CONCLUSION:

Sarcoma of breast is a very rare entity. Undifferentiated pleomorphic sarcoma(UPS) of breast is the diagnosis of exclusion. Usual age of presentation is sixth and seventh decades of life . Very few cases has been reported among young females. Our case report is of 24 year old female which is the youngest till date. Without immunohistochemistry i.e. only on the basis of clinical, radiological and histopathological examination it is very difficult to make a final diagnosis of undifferentiated pleomorphic sarcoma (UPS) of breast.

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