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Research Paper

Malignant Phyllodes tumor with necrosis – a rare case report

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ABSTRACT:- Phyllodes tumor of the breast is a biphasic fibro epithelial neoplasm. Phyllodes tumor (PT) accounts for 0.3-1% of all primary tumors and for 2.5% of all fibro epithelial tumors of the breast. Phyllodes tumor was originally described under the name cystosarcoma phyllodes, but phyllodes tumor is the current preferred term. Phyllodes tumors are further classified as benign, borderline, or malignant based on a variety of pathologic variables outlined in the current World Health Organization (WHO) classification. Overall average distribution of benign, borderline and malignant phyllodes are 60%, 20%, 20% respectively. The average in published data suggests a 21% rate of local recurrence, overall, with a 17%, 25% and 27% rate in benign, borderline and malignant PTs, respectively, and a 10% rate of metastases overall, with a 0%, 4% and 22% rate in benign, borderline and malignant PTs, respectively. Local recurrence after surgery is strongly dependent on the width of the excision margins. Malignant PTs develop on average 2-5 years later than benign PT. We report a case of malignant phyllodes tumor in a 22 year female who was present with complaints of pain, ulcer and palpable mass with fungating growth in left Breast. The histopathological appearance of this tumor was Malignant PT with necrosis.

Keywords:- Breast, Malignant, Phyllodes

I. INTRODUCTION

Although PTs may have been described as early as 1774, the lesion was first fully characterized in 1838 by Johannes MUller. The term cystosarcoma phyllodes (phyllo =Greek for leaf) was used to emphasize the leaf-like pattern, fleshy and cystic gross appearance of the lesion. PTs have been reported in patients ranging in age from 6 to 86 years. ²

The median age at diagnosis is about 45 years, approximately 20 years older than the median age of patients with fibro adenomas (FAs). A population-based study conducted in Los Angeles County, California, revealed an annual age-adjusted incidence of malignant PT of 2.1 per 1 million women. ⁴ The average size of PTs is 4 to 5 cm, ranging from 1 cm to larger than 20 cm. ^{2,3} Although malignant PTs tend to be larger than benign variants, there are many exceptions, with high-grade malignant PTs being smaller than 2 cm and some of the largest lesions being histologically benign. PTs with microscopically invasive borders usually appear well circumscribed grossly.

A fully malignant or high-grade PT typically features hypercellular stroma. Stromal overgrowth is also common. In most cases, this is combined with greater than 5 mitoses per 10 HPF and an invasive tumor border. The neoplastic stromal cells typically have high-grade and pleomorphic nuclei.

Rarely, the stroma contains specific sarcomatous elements such as Angiosarcoma, liposarcoma, chondrosarcoma, myosarcoma, or steosarcoma $^{7,\,8,9,10,11}$

Vascular invasion was identified in 3 of 31 high-grade malignant PTs studied by Tan et al. ³

The epithelial component may show usual ductal hyperplasia or metaplastic changes, such as squamous or apocrine. Stromal cellularity and mitotic activity are often increased in the periglandular areas.

II. CASE STUDY

We present a case of 22 year old female who visited to Surgery OPD with a one year history of the progressive left breast lump which later became associated with pain and ulcer and fungating mass. It was located on the left breast and involve nipple and areola. On the skin a large fungating mass measuring 10x9x7 cm was present. The lump was irregular, painful, firm, mobile and not attached to underlying muscle.

Her routine investigations like hemogram, routine urine microscopy and biochemistry were normal. Her chest X-ray is clear and no opacity was noted. Her general physical condition was normal. She was operated for left

mastectomy and axillary clearance under general anesthesia. The left breast with an axillary tail was removed and send to histopathological examination.

Pathologic finding- **Gross** – Left mastectomy specimen was measured 17x8x5 cm in size. A large fungating mass measuring 10x9x7 cm was present on external surface of the specimen (fig-1). The nipple and areola were obscured by mass. The tumor mass was multinodular, firm. The cut surface of the Mass was variegated, fleshy, and a few clefts were visible and showed necrosis, degeneration and hemorrhage at places (fig-2). The whole of the breast was involved with growth and normal breast tissue was not identified. Growth reached up to the skin. Total twelve lymph nodes were identified. Largest one measuring 3.5x3x2 cm in size. The cut surface of the lymph node was gray white.

Multiple sections from growth and all the lymph nodes were taken, stained with hematoxyline and eosin and examined under microscope.

Microscopic finding-

Section shows highly cellular tumor comprised of predominantly spindle cells in nested, leaf like and dispersed pattern with pleomorphic, hyperchromatic nuclei and plenty of bizarre mitoses (fig-3). Cells are round polygonal to spindle in shape, having moderate to abundant eosinophilic cytoplasm. Cell boundaries are distinct and cell membrane are irregular. Nuclei are highly pleomorphic and bizarre, Vesicular to hyperchromatic chromatin, Nucleoli are prominent in most of the cells. Numerous bizarre mitotic figures are present (8-10/10hpf) (fig-4). In between the tumor cells necrosis and hemorrhage are present (fig-5). At places blood vessels present. Multiple uninucleated, binucleated, & multinucleated giant cells are also seen (fig-4). The peripheral nodular areas showed compressed leaf-like benign ducts enclosed by proliferating benign stromal fragments.

All the twelve lymph node shows metastatic deposit (fig-6).

On the basis of clinical, gross and histopathological finding a diagnosis of Malignant phyllodes tumor with necrosis was offered.

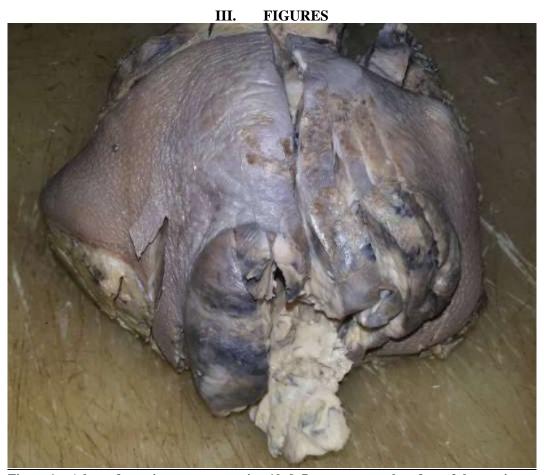


Figure 1- A large fungating mass measuring 10x9x7 cm on external surface of the specimen.

The nipple and areola are obscured by mass.



Figure 2- cut surface of tumor showing grey white variegated, fleshy appearance with necrosis and haemorrhage at places and few clefts seen.

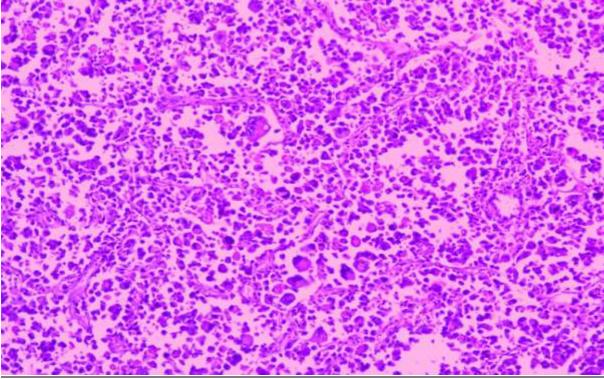


Figure 3- Section shows highly cellular tumor with pleomorphic, hyperchromatic nuclei and plenty of bizarre mitoses with numerous giant cells.

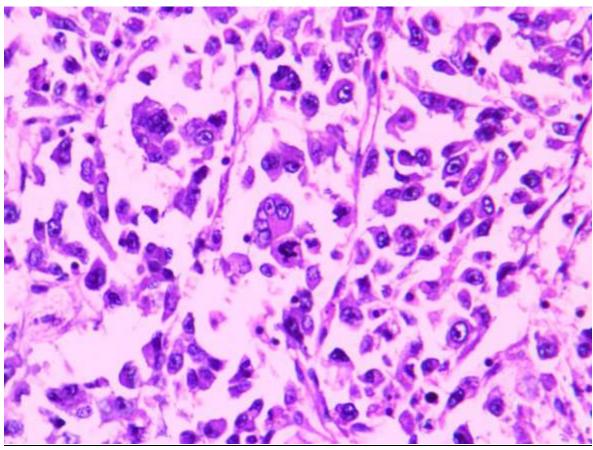


Figure 4- section shows bizarre mitotic figure with bi and multinucleated giant cells

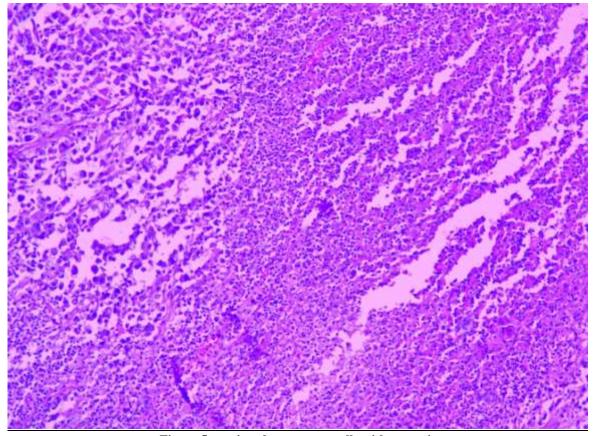


Figure 5- section shows tumor cells with necrosis

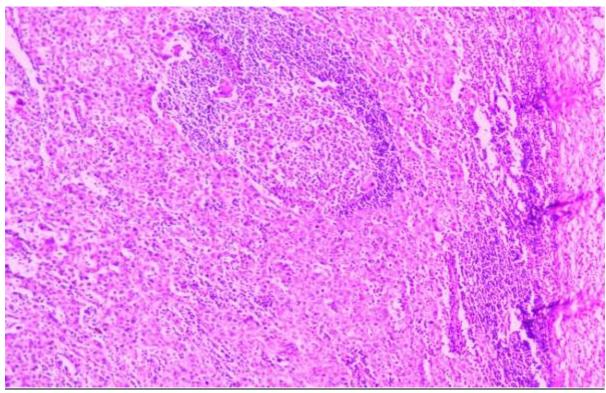


Figure 6 section from lymph node showing metastatic deposit

IV. DISCUSSION

Phyllodes tumors of the breast are fibroepithelial neoplasms that have the potential for recurrence and metastases. Phyllodes tumors can be divided into two major categories: low grade, and high-grade.

- Low-grade: A tumor with a "pushing" margin, mild cytologic atypia, and less than three mitotic figures per 10 hpf. Low-grade PT has a potential for local recurrence, but it is very unlikely to metastasize. A low-grade PT is a tumor with low malignant potential.
- High-grade (synonyms include malignant Phyllodes tumor and cystosarcoma Phyllodes): A tumor with either an infiltrating or pushing margin, moderate to severe nuclear atypia, and three or more mitotic figures per 10 hpf. Furthermore, the presence of stromal overgrowth is highly indicative of a high grade tumor. A high-grade PT is a tumor with high malignant potential.

It should be noted that the current WHO classification (2003) of tumors of the breast and female genital organs separates Phyllodes (phyllodes) tumors into benign, borderline, and malignant categories, mainly based on the mitotic activity, type of margin, stromal overgrowth, and cellular pleomorphism. ¹² For practical purposes, however, benign and borderline tumors can be viewed as low-grade PTs. Malignant tumors represent high grade PTs.

Although histological features have been helpful to some extent in predicting biologic behavior, specific parameters that can define the likelihood for recurrence are not universally accepted. Various investigators have found cellular pleomorphism, stromal overgrowth, tumor necrosis and heterologous stromal elements, or a combination of histological features to be prognostically useful. On the other hand, several authors have concluded that the adequacy of surgical margins is of paramount importance and that histological factors have an inconsistent influence on biologic behavior. ³

Sarcomatous stromal elements, including Angiosarcoma, chondrosarcoma, leiomyosarcoma, osteosarcoma, and rhabdomyosarcoma, are rarely encountered in malignant phyllodes tumors. Liposarcomas may also develop as stromal components of phyllodes tumors ⁷⁻¹⁰. Liposarcomatous differentiation in phyllodes tumors may consist of well differentiated, myxoid, round cell, and pleomorphic liposarcomatous elements. The finding of a malignant heterologous element places the tumor into a malignant category.

Phyllodes tumors are highly variable in their gross appearance, but a majority display a solid, fleshy mass with cystic areas. The tumors may be small or very large, ranging in size from 1 to 45 cm. The characteristic whorled appearance with curved clefts resembling leaf buds is seen in large tumors.

Biphasic neoplasm composed of a benign epithelial component (two cell layers of epithelial and myoepithelial cells) and a cellular, spindle cell stroma. The hallmark of the tumor is formation of leaf like processes protruding into cystic (dilated) spaces.

Well-developed fronds consist of epithelial-lined stromal projections that protrude into dilated glands or cystic spaces. The glands are usually widely spaced, dilated, and irregular with prominent side branches. The epithelium is often hyperplastic, and atypical columnar cell hyperplasia is a common finding. Rare tumors show lobular and ductal carcinoma in-situ. Apocrine and squamous metaplasia are occasionally seen

V. CONCLUSION

The high-grade malignant phyllodes tumor is a very rare but aggressive breast malignancy. Phyllodes tumor is always include in differential diagnosis when a rapidly growing mass present in breast in adult female.

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