Original Research Paper

Volume - 11 | Issue - 07 | July - 2021 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

General Surgery

REITERATIVE MALIGNANT SWELLINGS OF THE BREAST : A CASE REPORT

Dr Mohammed Raza — Professor And Unit Chief , Department Of General Surgery , JSS Medical College & Hospital , Mysuru (KA)

Dr Abhishek Gawri* — Postgraduate In General Surgery , JSS Medical College & Hospital , Mysuru (KA).*Corresponding Author

ABSTRACT Phyllodes tumor of the breast is a rare condition affecting most commonly, middle-aged women between 35-55 years of age. Histopathologically they can be classified as benign, borderline and malignant. These tumors have been known to grow to enormous proportions, often mimicking malignancy. The approach to Phyllodes tumor is similar to other breast lumps, viz. Triple assessment with clinical examination, Ultrasonography, where features of Fibroadenoma and Phyllodes tumor tend to overlap leading to misdiagnosis, and tissue diagnosis, wherein core biopsy is more sensitive than FNAC in diagnosing the condition. The treatment consists of wide local excision with negative margins. Positive margins almost invariably result in recurrence, which may occur over a short period of time. In this case report, we discuss a rare case of recurrent, metastatic, malignant Phyllodes tumor of the breast.

KEYWORDS : Phyllodes tumor, Cystosarcoma Phylloides, Recurrent phyllodes tumor, pulmonary metastasis, tumor progression, malignant phyllodes tumor

INTRODUCTION

Phyllodes tumor of the breast, also known by the names of "Cystosarcoma Phyllodes", "serocystic disease of Brodie", is a rare condition, contributing to 1 % of all breast tumors, of which malignant phyllodes is rarer (16 to 18 %). Phyllodes tumor is a fibroepithelial tumor of the breast with a diverse range of biological behavior. Pathologically they are divided into benign, borderline and malignant forms on the basis of stromal atypia, mitotic activity, stromal overgrowth, and the tumor margins.

These tumors may appear as benign fibroadenoma-like lesions, locally recurrent aggressive tumors, or widely metastatic malignant forms. Recurrent and malignant phyllodes have high metastatic potential, grow rapidly to horrendous proportions and ulcerate either due to tumor infiltration of pressure necrosis and palliative surgery is to be contemplated to improve the quality of life and alleviate symptoms of associated pain, infection, malodor, massive discharge, and bleeding for the short remaining life span of the patient.

They occur in females aged 35–55 years, typically 15–20 years, older than females with fibroadenomas and with a higher incidence in Asian females. (5) Imaging findings and clinical findings of fibroadenoma and phyllodes tumor overlap and they are commonly misdiagnosed. Triple assessment consisting of an initial physical examination, followed by radiological imaging (ultrasound and/or mammography) and histological sampling either by fine-needle aspiration cytology (FNAC) or core biopsy is the standard diagnostic pathway for palpable breast lesions. In case of phyllodes tumor, a core biopsy is more sensitive to rule out possibility of benign fibroadenoma.

Due to the fact that most Phyllodes tumors are not fully diagnosed pre-operatively, initial surgery does not always provide adequate margins necessitating frequent post-operative re-excision of the margins (6)

Here we discuss a case of recurrent phyllodes tumor which presented five times over a period of five years causing significant morbidity and poor quality of life for the patient along with financial burden and prolonged periods spent in different hospitals which finally was diagnosed to be a metastatic malignant phyllodes .

CASE REPORT

Case description

A 55 year old female, homemaker from rural Karnataka presented to us with a lump in the left breast region since 3 months associated with pain and an erosion over the skin. She was a known case of recurrent breast lumps.

In the past she had developed a similar swelling in the same region four times, the first appearing four years ago with a similar rapidly progressive history and had attained a size of 12 X 12 cm. She had been treated at a local hospital and excision of the lump was done. The following year recurrence of the swelling appeared at a different quadrant in the breast and again the lump was excised. The histopathology reports of the first two excision biopsies were lost.

The subsequent year she developed a lump occupying more than 2 quadrants of the breast and excision biopsy with a significant margin of breast tissue and the nipple areola complex was excised. Histopathology report revealed features suggestive of borderline phyllodes tumour with moderate stroma cellularity and atypia and no feature of positive margins, with the presence of occassional foci of microscopic invasions around tumor margins.

The fourth time appeared a year later and attained a huge size of 20 X 15 cm and simple mastectomy with muscle fibres from the Pectoralis major was removed. Histopathology report was suggestive of borderline phyllodes tumor with all margins showing tumor positivity. Grossly tumor mass was interspersing between the muscle fibres. Microscopically mild nuclear atypia, stromal cell predominance, and prominent nucleoli were present. IHC markers CD 34 was positive in spindle cells and CD 10 was focally positive. . Following this, no radiotherapy or chemotherapy was taken.

Currently , 9 months after the last surgery, she presented with a lump , initially of size 3 X 3 cm which gradually progressed over 2 months to a size of 4X 4 cm and then rapidly progressed to a size of 12 x 10 cm. She gave history of recent onset of pain , a pricking continuous type. She also had an erosion at the summit of the lump of size 2 x 3 cm which developed over a week and had no significant discharge. There was no history suggestive of malignant spread or swellings elsewhere in the body. Comorbidities included bronchial asthma for which she was on nebulization. She had attained menarche at the age of 12 and menopause at the age of 50, one year prior to appearance of the first lump. Her obstetric score was Para 1 Living 1, 21 years at the age of delivery and breast feeding was done for 9 months from both breasts. She gave no family history of breast, colon or ovarian carinomas. On examination, there was a mastectomy scar, healed by primary intention, across the left chest region with hypertrophy of scar over the sternum and absent nipple areola complex. A Spherical swelling of 12 X 10 X 10 (vertical, horizontal and Antero-posterior dimension) was present extending vertically from 3 rd to 6 th intercostal space and horizontally from left parasternal to anterior axilary line. Swelling was non tender with no local rise of temperature. The swelling had a bosselated surface with irregular borders , variable in consistency with soft cystic fluctuant regions and firm regions. A 2 X 3 cm oval erosion at the inferior aspect of the swelling was present with surround erythematous and indurated skin. Skin was fixed to the swelling only at this indurated region, rest was easily pinchable. There was decreased mobility of the swelling across the chest wall along the pectolaris major muscle fibre direction. There was no tetheting or dimpling. Engorged veins were present over the entire chest wall. There were no palpable axillary, supraclavicular lymphnodes.

Systemic examination revealed ronchi bilaterally , per abdomen and spine, long bones were normal.

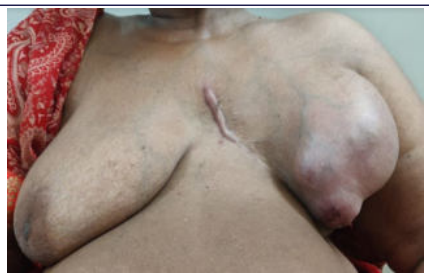

Fig1 Showing Large Left sided breast lump with post mastectomy scar

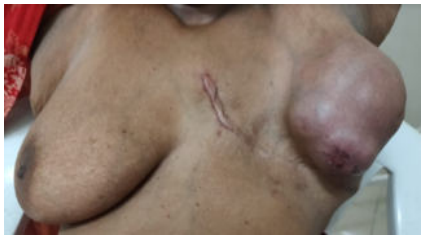

Fig 2 shows Left sided lump superior to the mastectomy scar with skin ulceration

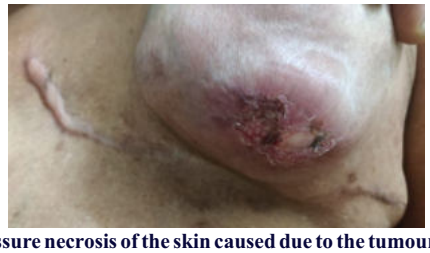

fig 3 Pressure necrosis of the skin caused due to the tumour

Treatment strategy and surgical technique:

Detailed metastatic work up was done which revealed pulmonary metastasis in CT thorax: Few soft density nodular lesions in the apico posterior segment of the right upper lobe, lateral segment of the right middle lobe, lingular segment and posterior basal segment of the right lower lobe, largest abutting the fissural and diaphragmatic margin measuring 3 X 2 cm.

USG Abdomen, Liver function tests, mammography of the opposite breast were normal.

She was taken up for excision biopsy of the tumor. An elliptical incision was taken encompassing previous surgical scar and the skin erosion. Intraoperatively superior, inferior and lateral margins of the tumor was excised with more than 1 cm of remaining breast / fatty tissure. Superficially the tumor was found abutting the skin with no interspersing adipose or breast tissue. Deep, the tumor was found to penetrate through the remaining pectoralis major muscle into chest wall and entire pectoralis major muscle and parts of pectoralis minor muscle was shaved off with the specimen. Axillary vein was seen pulled down to the surgical field due to fibrosis and contracture of surrouding adipose. No significantly enlarged lymph nodes were encountered. Adequate closeure of skin flaps were achieved. Drains were placed into the axillary and main tumor bed region.

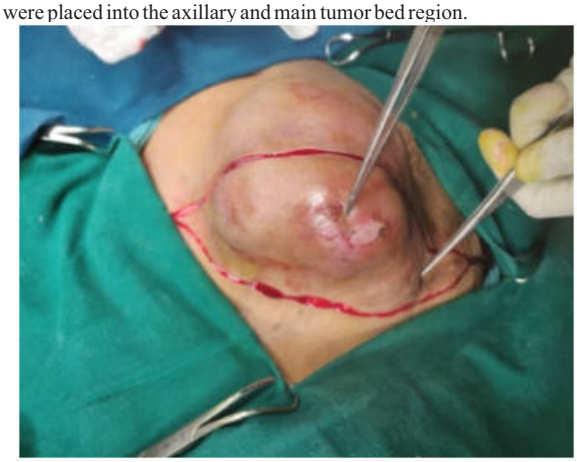

Fig 4 Intra op showing the incision

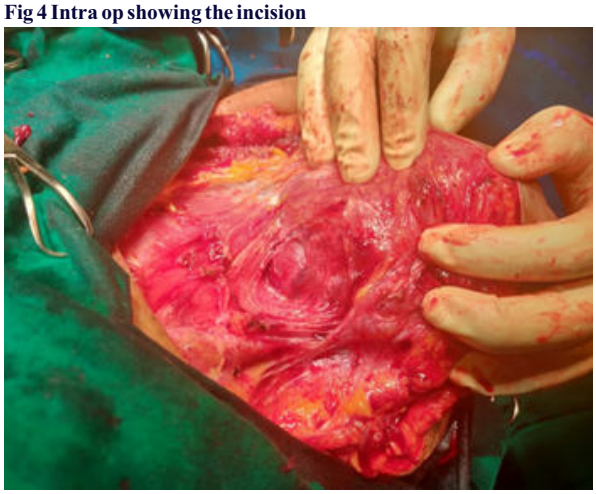

Fig 5 shows tumour infiltrating the pectoralis muscle

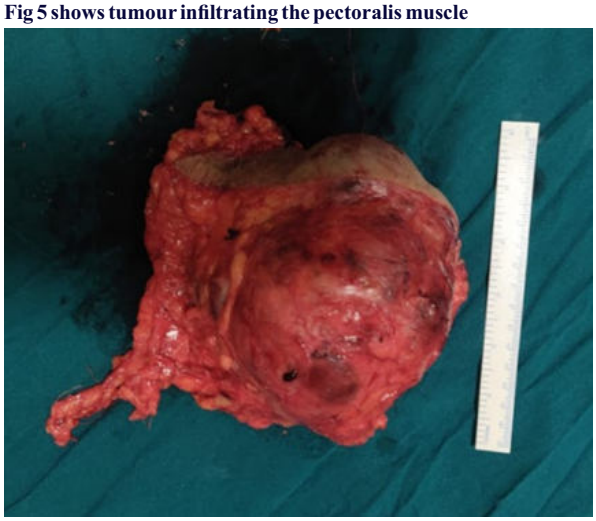

Fig 6 showing tumour excised in toto

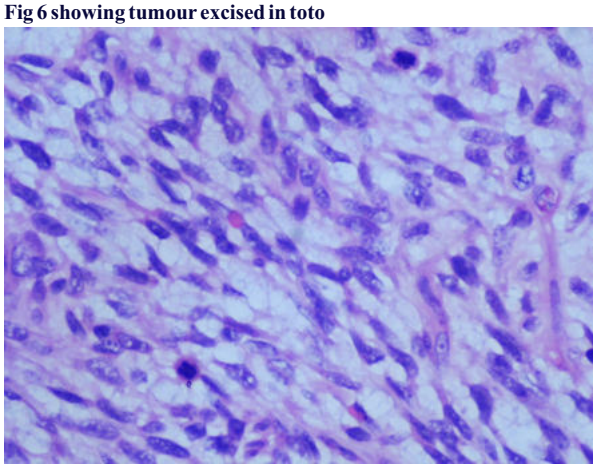

Figure 7 showing Histopathological picture of the tumour showing multiple mitoses

OUTCOME

DISCUSSION

The characteristic leaf-like architecture of the tumor on microscopic examination attributes to the given name of cystosarcoma phyllodes which contains varying degrees of epithelial and mesenchymal elements. (1). Malignant phyllodes tumors can recur as well as metastasize and have a uniformly poor prognosis. The usual survival for metastatic phyllodes is less than 2 years. (2)

Histologically WHO classifies phyllodes tumor as benign, borderline and malignant. Benign tumors exhibit low cellularity, circumscribed borders, less than five mitoses per ten high-power fields , monomorphic stromal cell nuclei and lack stromal overgrowth.

Malignant tumors show increased cellularity, infiltrative borders, more than ten mitoses per ten HPFs, marked stromal nuclear pleomorphism and stromal overgrowth ± stromal malignant heterologous elements (*e.g.* malignant bone/cartilage/fat). Borderline tumors fall between benign and malignant, possessing some but not all of the characteristics of malignant tumors.

Local recurrences are known to occur in all PTs with rates of 5–17%, 14–25% and 23–47% for benign, borderline and malignant PTs, respectively (7) The recurrences may reflect the original tumour or may show histological upgrade due to tumor progression in 25–75% cases. Metastases are very rare (<2%) and are usually encountered with malignant Phyllodes tumors (7).

Clinically they are staged similar to sarcomas according AJCC 8. The etiopathophysiology of phyllodes is also similar to sarcomas with very rare instances of lymph node involvement. Metastasis are also most common to lungs and long bones.

Surgery is the primary option for the treatment of malignant phyllodes tumor. However, the extent of surgery remains controversial. Excision of the tumor with negative margins of 1 to 2 cm is recommended. (4) Re-excision is recommended in cases with a positive surgical margin and stromal overgrowth and malignancy [8]. The giant phyllodes that is more than 10 cm in any dimension presents with several unique management problems. A simple mastectomy is performed for giant phyllodes tumours , those that are multifocal, in cases of recurrence or in phyllodes of 'borderline' histology but no recurrence cannot be guaranteed. Breast-conserving surgery is traditionally avoided in large multifocal phyllodes because of the risk of inadequate local excision and associated high local recurrence. In our case the 3rd and 4rd histopathology reports had evidence of tumor margins being infiltrated with tumor, so recurrence had to be expected.

Patients with large fungating malignant phyllodes with metastasis experience poor quality of life owing to foul-smelling discharge, pain, bleeding, disfigurement, and limitation of movement and psychosocial problems In the case in discussion it was the 5 th time the phyllodes tumor had presented and the patient had consulted carious doctors across the region with no conclusive treatment plan. Inspite of the poor prognosis owing to the factor of pulmonary metastasis the palliative surgery significantly improved the quality of life.

The initial two surgery reports and histopathology reports were missing but there is a possibility that initially the lumps could have been fibroadenomas which progressed to benign and then malignant phyllodes due to tumor progression. Recurrent fibroadenomas in the same breast, especially those of large size with rapid growth rate suggest a high transformation possibility from fibroadenoma to phyllodes tumor. (3)

Surgeons must be cautious in explaining the noncurative nature of the palliative surgical procedures so as to provide realistic treatment goals to these patients and their caregivers. The patient was counselled regarding enrolment in clinical trials for chemotherapy in the neighboring government facility as a possible treatment plan to extend the life span post surgery.

The five year survival for benign, borderline or malignant tumours is 96%, 74% and 66% respectively [9]

Radiotherapy can be planned for a patient for local control if distant metastasis are not present although its impact on disease-free or overall survival remains controversial (4) The role of adjuvant chemotherapy also needs a further study. There is no consensus on the role of chemotherapy in malignant phyllodes tumor.

In recent advances , malignant phyllodes seem to show over-expression of genes involved in angiogenesis including *VEGFA, Angiopoietin-2, VCAM1, PDGFRA*, and *PTTG.* The most common pathogenic mutations included *TP53* and *PIK3CA*. Overexpression of molecular biomarkers of increased angiogenesis, EGFR and immune checkpoints provides novel targeted therapy options in malignant phyllodes tumors of the breast. (10)

Studies that have evaluated the expression of hormone receptors ER expression and PR expression in phyllodes tumours with one report documenting HER2/c-erbB2 reactivity as well. Currently there is limited date to imply any significance to hormonal therapy in phyllodes tumor. C D 1 0 immunohistochemical staining can help to predict poor behavior of phylloides tumor in terms of recurrence and metastasis but this expression should be interpreted in conjunction with tumor category and other histological features especially mitotic activity and necrosis. CD34 reactivity, which is well described in the stromal cells of phyllodes tumours, has been reported to be inversely related to adverse histological features. (4) In our case, both CD 10 and CD 34 had been tested positive with the histopathology specimen of the 4 th surgery.

In conclusion, a high index of suspicion must be kept in mind for benign tumors presenting with rapid increase in size for the possibility of phyllodes tumor. In all cases of phyllodes tumor, the first surgery should achieve adequate clearance with minimum of 1 cm tumor free margins. If margins are found positive in histopathological examination , reexcision to attain local clearance must be done. Recurrence occurs when this is not achieved and can lead to detrimental morbidity and progression to malignant metastatic disease.

REFERENCES

1. . Singer A, Tresley J, Velazquez-Vega J, Yepes M. Unusual aggressive breast cancer: Metastatic malignant phyllodes tumor. J Radiol Case Rep. 2013 Feb 1;7(2):24–37. doi: 10.3941/jrcr.v7i2.1430.
2. Mituś JW, Blecharz P, Walasek T, Reinfuss M, Jakubowicz J, Kulpa J. Treatment of patients with distant metastases from phyllodes tumor of the breast. World J Surg. 2016 Feb;40(2):323–8. doi: 10.1007/s00268-015-3262-7.
3. Sanders LM, Daigle ME, Tortora M, et al. Transformation of benign fibroadenoma to malignant phyllodes tumor. Acta Radiol Open 2015;4:1–3.
4. Tan BY, Acs G, Apple SK, et al. Phyllodes tumours of the breast: a consensus review. Histopathology2016;68:5–21
5. Tse GM, , Niu Y, , Shi HJ. Phyllodes tumor of the breast: an update. *Breast Cancer* 2010; 17: 29–34. doi: 10.1007/s12282-009-0114-z
6. Guillot E, , Couturaud B, , Reyal F, , Curnier A, , Ravinet J, , Lae M, et al. . Management of phyllodes breast tumors. *Breast J* 2011; 17: 129–37. doi: 10.1111/j.1524-4741.2010.01045.x
7. Lakhani SR, , Ellis IO, , Schnitt SJ, , Tan PH, , van de Vijver MJ; International Agency for Research on Cancer, World Health Organization. *WHO classification of tumours of the breast*. 4th edn. Lyon, France: International Agency for Research on Cancer; 2012. pp. 240.
8. Taira N, Takabatake D, Aogi K, Ohsumi S, Takashima S, Nishimura R, Teramoto N. Phylloides tumour of the breast stromal overgrowth and histological classification are useful prognosis predictive factor for local recurrence in patients with a positive surgical margin. Jpn J Clin Incol. 2007;37(10):730–6.
9. Stebbing JF, Nash AG. Diagnosis and management of phyllodes tumour of the breast: experience of 33 cases at a specialized centre. Ann R Coll Surg Eng. 1995;77:181–184.
10. Gatalica Z, Vranic S, Ghazalpour A, et al. Multiplatform molecular profiling identifies potentially targetable biomarkers in malignant phyllodes tumors of the breast. *Oncotarget*. 2016;7(2):1707–1716. doi:10.18632/oncotarget.6421