

Rare case of malignant phyllodes in a young female

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ABSTRACT

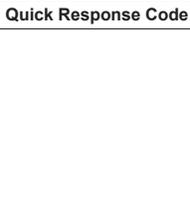
Introduction: Phyllodes tumors (PTs) of the breast are rare fibroepithelial tumors that constitute 0.3–0.5% of primary breast tumors. Incidence is maximum in women aged 45–49 years. Majority of PTs have been described as benign (35–64%). We are presenting a case of a 21 year female with bilateral breast lumps one of which turned out to be malignant PT. **Case Report:** We report a rare case of young female who presented with slowly progressive, minimally painful lump in the left breast along with multiple small painless lumps in the right breast. On examination, there was a left sided 4 × 3 cm tender, globular firm lump under the nipple areolar complex with nipple retraction with no axillary lymphadenopathy. Right breast showed multiple, non-tender lumps. Ultrasonography suggested a complex fibroadenoma on left breast with multiple fibroadenomas on right. Fine-needle aspiration cytology from the left breast suggested a highly atypical cytology and patient underwent excision of both breast lumps. Histopathological examination suggested a left sided Malignant PT with high grade sarcoma with resected margins positive for malignant cells and right-sided fibroadenomas. IHC report was positive for progesterone receptors and Vimentin receptors. Whole body positron emission tomography scan showed no FDG avid lesions in either breast or anywhere else. Patient is now currently under follow-up and a wide local excision is being planned. **Discussion:** PTs occur most commonly during the late fifth decade of life in females. Our case adds to the data pool where malignant PTs are found in a younger age group. Surgical management is the mainstay but type of surgery has always been a source of debate. The use of radiation therapy and adjuvant chemotherapy is controversial. **Conclusion:** Accurate preoperative diagnosis and vigilance for malignant PTs irrespective of the age group of the patient are vital for the appropriate treatment and a disease-free survival.

Key words: Phyllodes tumors, Fibroadenoma, Lump

INTRODUCTION

Phyllodes tumors (PTs) of the breast are rare fibroepithelial tumors that constitute 0.3–0.5% primary breast tumors.^[1] Incidence is maximum in women aged 45 to 49 years. The tumor is rarely found in adolescents

and the elderly.^[2] Majority of PTs have been described as benign (35–64%), with the remainder divided between the borderline and malignant subtypes based on the degree of stromal cellular atypia, mitotic activity, infiltrative versus circumscribed tumor margins and presence or absence of stromal overgrowth.^[2] Phyllodes are characterized by hypercellular stroma growth into epithelial lined cyst spaces and the epithelial element is responsible for the distinction from stromal sarcomas.^[3] Accurate preoperative pathological diagnosis allows correct surgical planning and avoidance of reoperation, either to achieve wider excision or for subsequent tumor recurrence. Malignant PTs, if inadequately treated, have a propensity for rapid growth and metastatic spread.^[1] National Comprehensive Cancer Network

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Figure 1: Phyllodes tumors (PTs)



Figure 2: Left breast lumps - cystic masses with solid components

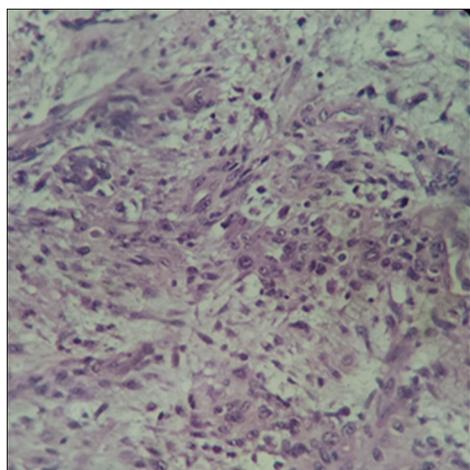


Figure 3: H&E (40 ×) hyper and hypocellular areas

guidelines for the management of PTs recommend wide excision with margins ≥ 1 cm and recommend against axillary staging.^[3] Malignant PTs are a rare entity and encountering such pathology in a younger age group is

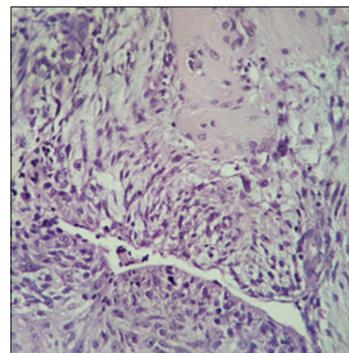


Figure 4: H&E (40 ×) Marked stroma atypia with ITC showing high pleomorphism

what makes our case fascinating. We are presenting a case of a 21 year female with bilateral breast lumps one of which turned out to be malignant PT.

CASE REPORT

We report a rare case of young female who presented with slowly progressive, minimally painful lump in the left breast along with multiple small painless lumps in right breast. There was no ulceration over skin or nipple discharge. She had no significant past or family history. On examination, there was a left sided 4 × 3 cm tender, globular firm lump under the nipple areolar complex with nipple retraction with no axillary lymphadenopathy. Right breast showed multiple, non-tender lumps. Ultrasonography suggested a complex fibroadenoma on the left breast with multiple fibroadenomas on the right. Fine needle aspiration cytology from the left breast suggested a highly atypical cytology and patient underwent excision of both breast lumps [Figure 1]. Histopathological examination suggested a left-sided malignant PT with high grade sarcoma with resected margins positive for malignant cells and right sided fibroadenomas [Figures 2-4]. IHC report was positive for Progesterone receptors and Vimentin receptors. Whole body positron emission tomography scan showed no FDG avid lesions in either breast or anywhere else. Patient is now currently under follow-up and a wide local excision is being planned.

DISCUSSION

Primary breast neoplasms are extremely rare in adolescent females and most are benign in nature. PTs occur most commonly during the late fifth decade of life in females and are even more rarely do they occur in men.^[4] Our

case adds to the data pool where malignant Phyllode tumors are found in a younger age group. Surgical management is the mainstay but the type of surgery has been a source of debate over the years. Studies have shown no differences between breast conserving surgery versus mastectomy in terms of metastasis-free survival or overall survival, despite the higher incidence of local recurrence that comes with breast conserving surgery. Re-excision of borderline and malignant PTs identified after local excision should be considered. As malignant PTs undergo mainly hematogenous spread and lymph node metastases are rare so routine axillary clearance is not recommended. Local recurrence rates for PTs are 15–20% and are correlated with positive excision margins. Stromal cellularity, stromal overgrowth, stromal atypia, mitotic activity, tumor margin, and heterologous stromal elements were also significantly correlated with metastasis.^[5]

The use of radiation therapy and adjuvant chemotherapy is somewhat controversial and depends on extent of disease. Radiotherapy has been used with good results for local control of the disease and it may be considered for high-risk PTs, including those >5 cm, with stromal overgrowth, with more than 10 mitoses per high power field, or with positive margins.^[6]

Given the unique and uncommon nature of malignant phyllodes in younger females, appropriate assessment and management must be achieved to prevent misdiagnosing these rare, aggressive tumors.

CONCLUSION

Accurate pre-operative diagnosis and vigilance for this rare entity irrespective of the age group of the

patient are empirical for the appropriate treatment and increased survival. Surgery with negative margins is the mainstay of treatment and regular follow-up is necessary to keep a check on recurrence and metastasis.

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