Malignant Phyllodes Tumor of the Breast: A Case Study

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Malignant phyllodes tumors of the breast are rare, fast-growing tumors that can be difficult to diagnose. A case study is featured about a young adult patient who lacked insurance and received a delayed diagnosis of malignant phyllodes tumor of the breast. This article includes pertinent clinical and age-specific considerations for comprehensive management.

Phyllodes tumors of the breast (PTB) are generally rare tumors and represent less than 0.5 percent of all breast tumors (Reinfuss et al., 1996). Those neoplasms are most commonly diagnosed between 35–55 years of age, with reports of cases in both adolescents and among older adult women (Khosravi-Shahi, 2011; Reinfuss et al., 1996). PTB are classified as benign, borderline, or malignant based on the degree of atypia in the stroma and overall mitotic rate (Kraemer et al., 2007). The five-year survival rate for malignant PTB is reported to range from 66%–82% (Khan & Badve, 2001). These tumors often exhibit an increase in size to 7.8 cm in the four-week period. In addition, the vascularity of the lesion was noted to have increased. A core biopsy demonstrated a low-grade spindle cell neoplasm with a primary differential diagnosis of cellular fibroadenoma versus phyllodes tumor. L.P. was recommended to undergo an excisional biopsy. She was counseled regarding potential need for additional surgery if the mass proved to be malignant.

Pathology from the excision was consistent with a high-grade malignant phyllodes tumor. A computed tomography (CT) scan of the chest was performed to rule out metastatic disease. L.P. was then taken back to the operating room for re-excision. Her second surgery achieved greater than 5 cm tumor-free margins circumferentially. Because of the significant cosmetic defect after her second surgery, L.P. ultimately decided to undergo a completion mastectomy with implant reconstruction.

Diagnostic Evaluation

In general, PTB tend to be large, fast-growing, and difficult to differentiate from benign fibroadenoma in clinical presentation, and radiologic and pathologic findings (Kraemer et al., 2007). PTB are usually palpable and appear as mobile dominant masses in physical examinations (Khan & Badve, 2001). These tumors often exhibit a period of rapid growth even after being stable for months or longer (Telli, Horst, Guardino, Dirbas, & Carlson, 2007). On mammogram, PTB appear well defined and can be oval, round, or lobulated (Telli et al., 2007). Ultrasound findings also include a heterogeneous echo pattern and absent microcalcification (Chao, Lo, Chen, & Chen, 2002).

The diagnosis of malignant PTB is based on histopathologic evaluation. Like their
benign counterparts, malignant phyllodes represent neoplastic proliferations of the breast stromal, or connective, tissue (Kemp, Burns, & Brown, 2008). All phyllodes tumors have a propensity for local recurrence; however, malignant phyllodes also bear metastatic potential. Features that push the diagnosis from benign to malignant include increased mitotic activity (more than 5–10 mitotic figures per high-power microscopic field), severe atypia within the stromal cells, overgrowth and infiltration of the stromal tissue into the surrounding breast tissue, and the presence of necrosis and/or hemorrhage (Kemp et al., 2008). Obtaining the correct diagnosis may be complicated by the fact that malignant phyllodes tumors vary considerably from one area to the next. A core biopsy that happens to sample a less-worrisome appearing region of the tumor may be falsely reassuring (with fine needle aspirations and core needle biopsies yielding false negative results estimated at 63%) (Telli et al., 2007). Both benign and malignant phyllodes tumors may mimic cellular fibroadenomas on limited biopsy specimens. Often, a diagnosis of malignant phyllodes tumor can only be rendered after evaluation of a complete excisional specimen, with thorough assessment of the tumor borders and careful mitotic counts (see Figures 1 and 2).

**Treatment Considerations**

Wide surgical excision (with at least 1 cm clear margins) remains the primary treatment modality for PTB (Barth, 1999; Khan & Badve, 2001). Total mastectomy is considered when adequate margins cannot be achieved through a breast-conserving surgical approach. Surgical axillary staging is usually not performed as PTB rarely metastasize to axillary lymph nodes (with reports of axillary metastases occurring in less than 10% of cases) (Khosravi-Shahi, 2011; Kraemer et al., 2007). The use of adjuvant radiation therapy is controversial, with no firm data to support its use or benefit when negative margins have been obtained (Gnerlich, Williams, Yao, Jaskowiak, & Kulkarni, 2014; Khan & Badve, 2001). In addition, no evidence exists that adjuvant chemotherapy is beneficial in malignant PTB; however, it has been associated with improved survival in other breast sarcomas (Khan & Badve, 2001). Finally, no established role exists for hormonal therapy in the care of malignant PTB (Telli et al., 2007).

Based on data from large case series, an estimated 5%–10% of patients with PTB will develop a distant metastasis, and this incidence increases to 20% among those with malignant PTB (Telli et al., 2007). Metastases are most common in the lungs, but can also occur in the soft tissue, bone, and liver (Reinfuss et al., 1996; Telli et al., 2007). Metastatic PTB is associated with a poor prognosis and average survival is less than two years (Barth, 1999; Telli et al., 2007). Limited case reports document the use of ifosfamide, a combination of cisplatin and doxorubicin, or a combination of cisplatin and etoposide among metastatic PTB, with a range of therapeutic and palliative effects. In addition, radiation therapy has been used in a palliative context with some success (Telli et al., 2007).

**Resolution of Case**

In the interim, L.P. met with the genetic counselor to have a TP53 test to rule out L-Fraumeni syndrome, which yielded a normal (negative) result. The radiation oncologist presented the option of conventional dosing radiation (50 Gy in 25 fractions) to minimize risk of local recurrence; however, L.P. and her family decided against this treatment option. She was also referred to medical oncology and was counseled at length regarding adjuvant treatment recommendations and the lack of evidence supporting chemotherapy. The medical oncologist presented a 25% risk of distal recurrence and a 75% five-year survival to L.P. It was recommended that she have CT imaging at six-month intervals, and she was also counseled regarding smoking cessation.

L.P. discussed her feelings of anxiety, depression, and insomnia since the time of diagnosis. She had to quit her job as a waitress. Although she has an extremely supportive family, her boyfriend (her primary source of social support) is serving overseas in the military. She also has had a difficult time adjusting with the uncertainty of recurrence. L.P. states that she is actively engaged in the young adult cancer online community, and a social worker will follow her case and progress. A referral to psychiatry may be indicated.

**Implications for Nursing**

Given the rarity of malignant PTB, treatment principles to date have relied on small, retrospective series and case reports. Unfortunately, triple assessment involving clinical, radiologic, and histologic examination often results in low diagnostic accuracy for PTB because they have similar characteristics to benign breast disease in all three areas (Telli et al., 2007). Because of this, advanced practice nurses should be aware of PTB as a possible differential and help navigate the patient to surgical excision for definitive diagnosis.
L.P. had financial obstacles, which delayed her presentation to a tertiary academic medical center for eventual diagnosis and treatment. Unfortunately, these insurance issues are pervasive among young adults. Young adults (aged 19–29 years) are the largest demographic of uninsured and underinsured patients in the United States (Nicholson & Collins, 2009). The lack of insurance results in lag times in diagnosis for their cancer (Bleyer, 2005). In addition, because of the age at diagnosis, young adults often require a broader scope of supportive care and psychosocial support (Bleyer, Albritton, Ries, & Barr, 2007). Many young adults report feelings of anxiety and depression, and the cancer diagnosis often requires them to enter a dependent role with their parents again (Bleyer et al., 2007), which was the case for L.P. Finally, because of L.P.’s age and the real possibility of local or distant recurrence, continued support for anxiety regarding this uncertainty is a relevant priority for nursing care delivery.

References