

รายงานกรณีศึกษา: เนื้องอกฟีลโลดเต้านมชนิดร้ายแรงที่มีการกลับเป็นซ้ำอย่างรวดเร็วร่วมกับการลุกลามไปยังก้อนในช่องกลางทรวงอก

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Case Report: Aggressive Recurrence of a Malignant Phyllodes Tumor with Rapid Progression and Mediastinal Invasion

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Abstract

Phyllodes tumors are rare breast neoplasms characterized histologically by leaf-like architecture. These tumors can present as benign, borderline, or malignant. This case report describes a 35-year-old female with a rapidly enlarging mass in her right breast over a span of three months. On physical examination, a large mass was observed throughout the entire right breast. The patient underwent surgical treatment with a mastectomy, and the surgical site was closed using abdominal muscle flaps. Postoperative histopathological examination revealed a malignant phyllodes tumor with clear surgical margins. The patient subsequently received adjuvant radiotherapy. During follow-up, metastasis to the lungs was detected nine months post-surgery, along with a mass in the mediastinum, which rapidly invaded the major veins, resulting in bilateral arm swelling and facial edema. This article will discuss the general knowledge and types of phyllodes tumors, the significance of surgical intervention in their treatment, and the recurrence of this tumor type.

Keywords: Phyllode tumor, Metastasis, Breast neoplasm

บทคัดย่อ

เนื้องอกฟีลโลด (Phyllodes tumors) เป็นเนื้องอกหายากที่พบได้ในเต้านม โดยมักจะมีลักษณะทางพยาธิวิทยาเป็นโครงสร้างคล้ายใบไม้ และสามารถมีลักษณะเป็นเนื้องอกที่ไม่ร้ายแรง (benign) กลาง (borderline) หรือร้ายแรง (malignant) ได้ รายงานกรณีนี้เป็นของหญิงอายุ 35 ปีที่มีอาการมีก้อนที่เต้านมขวาขยายตัวอย่างรวดเร็วภายใน 3 เดือนจากการตรวจร่างกายพบก้อนขนาดใหญ่ทั่วทั้งเต้านมด้านขวา ผู้ป่วยได้รับการรักษาผ่าตัดด้วยการตัดเต้านมทั้งเต้า และปิดแผลด้วยกล้ามเนื้อหน้าท้อง ผลชิ้นเนื้อหลังการผ่าตัดพบว่าเป็นเนื้องอกฟีลโลดชนิดร้ายแรง (malignant phyllode) ผลชิ้นเนื้อได้ขอบเขตครบ ผู้ป่วยได้รับการรักษาด้วยการฉายรังสีหลังการผ่าตัด ภายหลังจากการตรวจติดตาม พบว่าผู้ป่วยมีการกระจายไปยังปอดในระยะเวลา 9 เดือนหลังการผ่าตัดและมีก้อนที่บริเวณช่องกลางทรวงอก จนทำให้เกิดการกดเบียด

เส้นเลือดดำใหญ่ในระยะเวลาย้อนรวดเร็ว มีอาการแขนบวมสองข้างและใบหน้าบวม ในบทความนี้จะมีการอภิปรายถึงความรู้ทั่วไปและชนิดของเนื้องอกฟีลโลด ความสำคัญของการผ่าตัดในการรักษาเนื้องอกฟีลโลด และการกลับเป็นซ้ำของเนื้องอกชนิดนี้

คำสำคัญ: เนื้องอกฟีลโลด, การกระจายของเนื้องอก, เนื้องอกเต้านม

Introduction

Phyllodes tumors are uncommon breast neoplasms that arise from both the stromal and epithelial elements.¹ These tumors account for less than 1% of all breast neoplasms and approximately 2.5% of all fibroepithelial breast tumors.² According to the World Health Organization (WHO), phyllodes

tumors are histologically classified into three subtypes: benign, borderline, and malignant based on their histopathological characteristics.³

Globally, the estimated incidence of phyllodes tumors is approximately 2.1 cases per million women per year.² In Thailand, data on phyllodes tumor incidence are limited, but studies suggest a similar prevalence to global trends, with malignant tumors comprising a small subset. A retrospective review at Ramathibodi Hospital reported that phyllodes tumors accounted for 168 patients excised over a 16-year period, with 12.5% being malignant.⁴

Malignant phyllodes tumors are known to be difficult to treat, with a high recurrence rate. The optimal management of malignant phyllodes tumors typically involves surgical resection with clear margins.

In selected malignant cases, adjuvant radiotherapy may reduce the risk of local recurrence, although the role of systemic chemotherapy remains uncertain and is generally reserved for metastatic disease.

Case Presentation

A 35-year-old Thai woman with no significant medical history presented with a rapidly enlarging mass in her right breast, which had grown significantly over the past 3 months. On physical examination, a mass exceeding 12 cm in size was palpated in the right breast. The mass had grown to 15 cm in size in the 2 weeks prior to surgery, and the surface was purple, indicating pressure from the tumor. There was no regional lymphadenopathy. The mass was still mobile on physical examination.

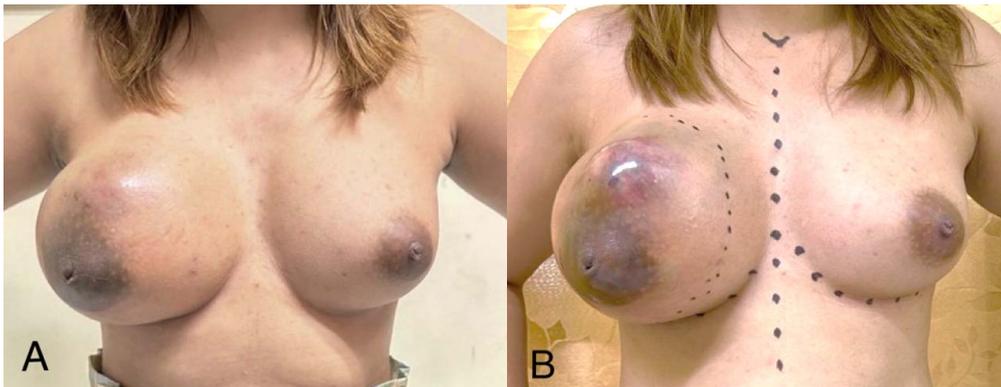


Figure A : The lesion at the initial visit.

Figure B : The lesion after two weeks (one day prior to the operation). It is noticeable that the mass has become more prominent, and the overlying skin appears purplish due to pressure from the rapidly enlarging mass.

Imaging studies, including breast ultrasound and mammography, demonstrated a well-defined, mixed solid and cystic mass measuring 11.0 × 11.2 × 8.8 cm, occupying the entire right breast. The lesion demonstrated internal vascularity without evidence of abnormal axillary lymphadenopathy. Core needle biopsy of the mass revealed atypical cells with multinucleated giant cells and extensive areas of necrosis. During the biopsy procedure, serosanguinous fluid was noted to drain from the mass; therefore,

cytological examination of the fluid was performed, which returned positive for malignancy. A computed tomography (CT) scan of the chest and upper abdomen showed no evidence of distant metastases.

The patient underwent a right mastectomy with sentinel lymph node biopsy (SLNB) and unilateral Transverse Rectus Abdominis Myocutaneous (TRAM) flap reconstruction due to the large tumor size, which precluded primary closure. Because the initial core needle biopsy did not yield a definitive diagnosis regarding the tumor subtype, SLNB was performed as

a precautionary measure. Postoperative histopathological examination confirmed a malignant phyllodes tumor, characterized by a mitotic rate of 10 mitoses per 10 high-power fields (HPFs), moderate to marked stromal atypia, and the presence of heterologous elements including osteosarcoma, chondrosarcoma, and pleomorphic liposarcoma. Surgical margins were clear by more than 1 cm, except for the posterior margin, which measured 1 mm.



Figure C : Postoperative photograph showing the reconstructed breast using a TRAM flap.

Post-surgery, the patient received adjuvant radiotherapy within one month. Although the surgery was successful and the patient resumed normal activities, she developed a persistent cough nine months later. A chest X-ray and CT scan revealed extensive pulmonary metastasis. Subsequently, the patient also presented with swelling in her arms and face, which was attributed to tumor invasion of the superior vena cava and brachiocephalic veins.

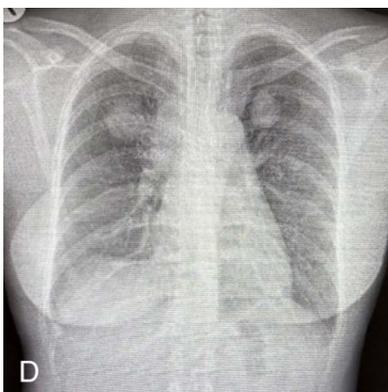


Figure D : The chest X-ray demonstrates multiple pulmonary metastases.



Figure E : Computed tomography (CT) scan reveals multiple metastatic lung and mediastinal masses with direct invasion into the superior vena cava (SVC).

Chemotherapy was recommended as the next step, but after a discussion about the prognosis and potential outcomes, the patient declined further treatment. Her condition rapidly deteriorated, and she was referred for palliative care. This case highlights the aggressive nature of malignant phyllodes tumors and underscores the importance of early detection and vigilant follow-up.

Discussion

Phyllodes tumors are rare fibroepithelial tumors of the breast, with an incidence of approximately 2.1 cases per million women annually. They account for 0.3% to 1% of all breast neoplasms.² These tumors are categorized as benign, borderline, or malignant based on histological characteristics such as tumor margins, stromal overgrowth, necrosis, cellular atypia, and mitotic activity.⁵ The incidence of malignant phyllodes tumors is estimated to be 12-30% of all phyllodes tumors.^{4, 6}

Malignant phyllodes tumors tend to grow rapidly and are typically painless. They are more aggressive than benign phyllodes tumors, with a higher risk of local recurrence and distant metastasis, most commonly to the lungs, where they may appear as solid nodules or thin-walled cavities. Lymph node involvement is rare.⁷ Core needle biopsy has a sensitivity of approximately 13-25% and a

specificity of 90-100%, as it requires differentiation from fibroadenoma and fibroepithelial lesions.⁸

Surgical excision with clear margins, typically greater than 1 cm, is the mainstay of treatment for malignant phyllodes tumors. In cases where negative margins cannot be achieved via breast-conserving surgery, mastectomy is often preferred.⁷ However, obtaining adequate margins can be challenging, particularly when the tumor is large, and complete mastectomy is often required. In this case, the posterior margin was less than 1 cm despite the mastectomy. In selected cases where there is tumor invasion into the ribs or chest wall, chest wall resection may be warranted to ensure adequate oncologic margins.

Adjuvant therapies, including radiation, are frequently used for high-grade malignant phyllodes tumors or those with a risk of recurrence. Some studies have demonstrated that radiation therapy can significantly reduce the risk of recurrence.⁹⁻¹⁰ However, the role of chemotherapy in treating malignant phyllodes tumors remains unclear, as there is insufficient evidence to confirm its benefit.^{7, 11-12} While the patient in this case received radiotherapy, her early pulmonary metastasis highlights the aggressive nature of the tumor and the limited effectiveness of current therapies in preventing metastasis.

The rates of local recurrence and distant metastasis in malignant phyllodes tumors range from 20% to 32%.^{9, 13} Prognosis is poor for those with metastatic disease, particularly when metastasis occurs early, as demonstrated in this case.

Risk factors associated with metastasis include high mitotic activity (>10 per 10 high-power fields), marked stromal atypia, stromal overgrowth, presence of stromal necrosis, large tumor size (>9 cm) with heterologous elements, and positive surgical margins.¹⁴⁻¹⁷ In this patient, nearly all these risk factors were present, except for positive margins.

Due to the possibility of early metastasis, particularly in high-risk patients, long-term follow-up protocols with scheduled imaging (e.g., chest CT) should be developed and individualized based on risk factors.

Conclusion

Malignant phyllodes tumors are rare and aggressive breast neoplasms that require early detection and careful management. While surgical resection with clear margins remains the standard treatment, the benefits of adjuvant therapies, such as radiation and chemotherapy, remain uncertain. The high potential for metastasis and recurrence emphasizes the need for continuous surveillance. This case underscores the importance of a multidisciplinary approach to effectively manage malignant phyllodes tumors and improve patient outcomes.

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