

Malignant Phyllodes Tumor in a Preteen Girl Presenting As a Huge Cystic Mass: A Case Report

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Malignant phyllodes tumors are exceptionally rare in children and adolescent females. Herein, we report a challenging case of malignant phyllodes tumor in a preteen girl who initially presented with a rapidly-growing, huge, palpable mass followed by a discussion of her imaging findings.

Index words: Breast; Breast neoplasm; Malignancy; Phyllodes tumor; Adolescent

Introduction

Phyllodes tumors account for less than 1% of all breast neoplasms and usually occur in women in the 5th or 6th decade of life (1). It is uncommon in children and adolescent females, and the malignant form is exceptionally rare (2). Moreover, there are only a few reports on image findings of malignant phyllodes tumors in children and adolescents. Here, we report a rare case of malignant phyllodes tumor in an adolescent female who presented with a rapidly-growing, huge, palpable breast mass, with imaging features from the mammography,

ultrasound (US) and Magnetic resonance imaging (MRI).

Case Report

A preteen female visited our hospital with a huge, palpable, non-tender mass in her left breast, which had rapidly grown in 6 weeks. Physical examination revealed a hard palpable mass with erythematous skin changes. Mammography showed a large, round, circumscribed, isodense mass involving the entire left breast (Fig. 1a). US (Fig. 1b) depicted a 12 cm-sized, complex, solid and cystic mass with variable echogenicity in the cystic portion. In color Doppler US, vascularity was not evident in the solid portion. MRI (Fig. 1c) showed a circumscribed heterogeneous mass containing multi-loculated fluid, suggesting various stages of hemorrhage within the mass. The solid portion of the mass

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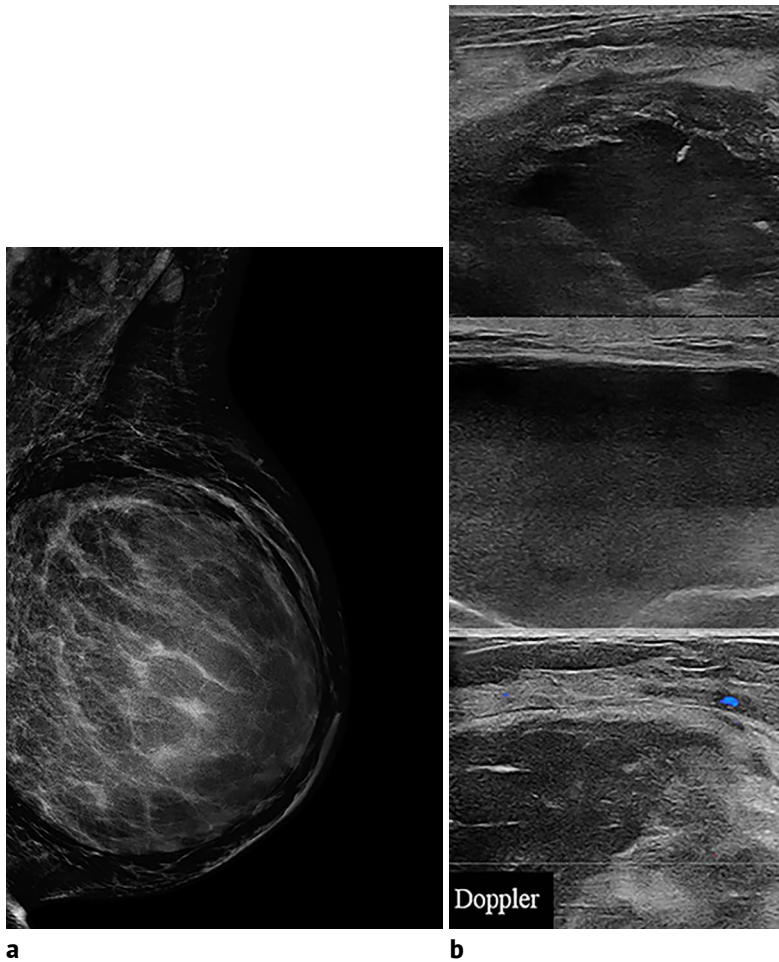
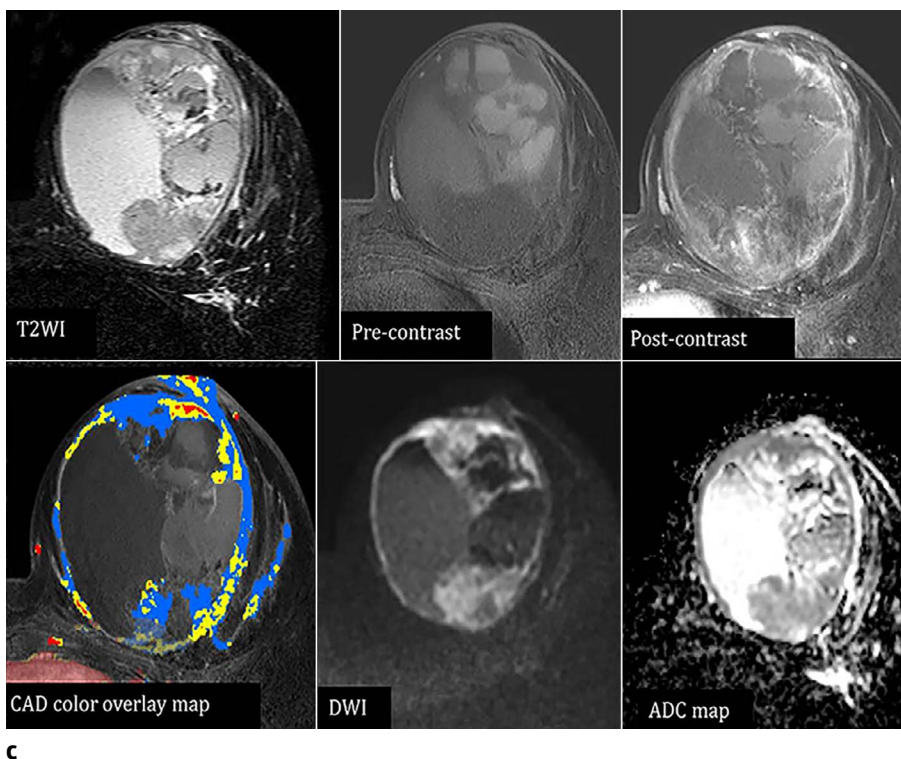


Fig. 1. A preteen girl with malignant phyllodes tumor, presenting as a huge palpable non-tender mass in her left breast.

a. Mammography shows a huge round isodense mass in left breast. **b.** US reveals a circumscribed mass which has large areas of cystic portion with various echogenicity. Blood flow within the mass is nearly absent in the color Doppler US. **c.** Magnetic resonance imaging of the left breast mass: T2WI shows a multiloculated heterogeneously iso-to high signal intensity mass containing hemorrhagic fluid collection of various phases. Image obtained at 1-minute post-gadolinium injection shows thick irregular wall of the mass with heterogeneous enhancement. CAD color map shows various enhancement kinetics in the solid portion. DWI and corresponding ADC map shows diffusion restriction along the peripheral solid portion.



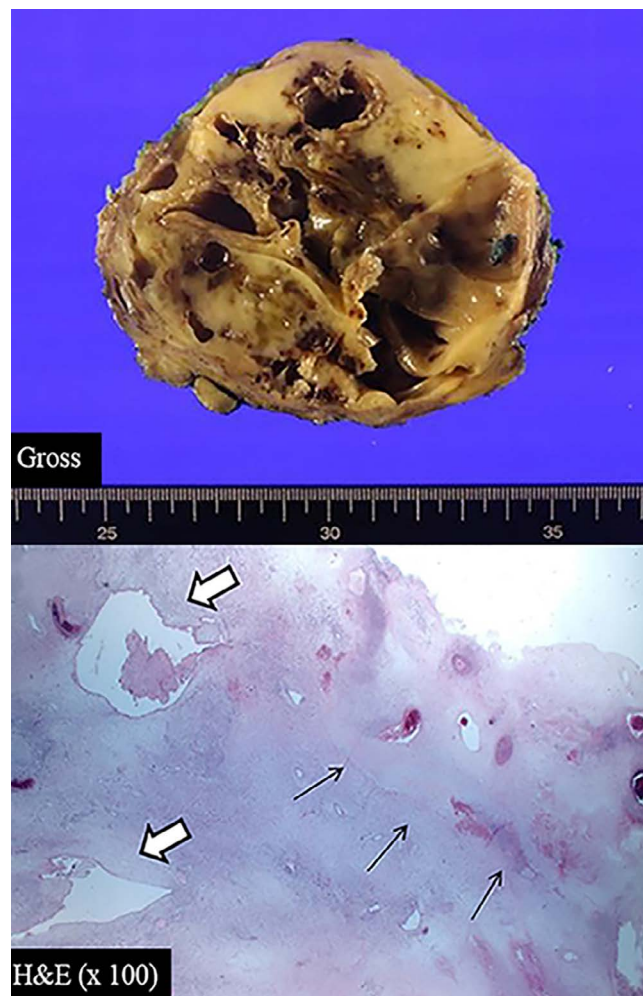


Fig. 1. d. Gross photograph shows a 11 x 10.5 x 10 cm in size and tan colored mass with extensive cystic change. Microscopic examination (x100) shows a tumor mainly composed of stromal component. Epithelial component with cystic change was noted (open arrows) with necrosis (arrows).

showed iso- to high signal intensity (SI) and dark internal septa on T2-weighted image (T2WI) and diffusion restriction on diffusion-weighted image (DWI). On dynamic contrast enhancement, the mass showed heterogeneous enhancement with various enhancement kinetics. The patient underwent mass excision. The gross specimen showed a 11 x 10.5 x 10 cm sized, circumscribed, tan-colored mass with extensive cystic changes. Histopathologic analysis revealed malignant phyllodes tumor (Fig. 1d). Completion mastectomy was subsequently performed.

Discussion

In children and adolescents, the most common malignant tumors of the breast are metastasis, and primary breast malignancy is extremely rare. One of the most frequent histologies of primary breast malignancy in children and adolescents is malignant phyllodes tumor (3).

Oftentimes, phyllodes tumors cannot be clinically distinguished from fibroadenomas, but they should be suspected when a patient presents with a large (>3 cm), rapidly-growing, palpable breast mass (1). Until now, there have been few publications reporting malignant phyllodes in young patients (4), and imaging findings are rarely discussed.

On mammography, phyllodes tumors are usually nonspecific and often show large, dense masses (5). US shows circumscribed, round, heterogeneous, hypoechoic masses with cleft-like cystic spaces (5). However, the findings are often not distinguishable from the findings of fibroadenoma. For the MR findings, the limited information in the literature includes smooth margins, lobular shape, frequent cystic changes or internal septa (6).

Benign and malignant phyllodes tumors show a substantial overlap in tumor size, mammographic and US features (5, 7). MRI is known to be helpful because malignant phyllodes tumors show higher SI than normal breast tissue on T1WI, lower than or equal SI compared to normal breast tissue on T2WI, low ADC on DWI, and cystic changes with irregular walls (7). Our case showed multi-loculated, large, cystic lesions with various SI, irregular thick walls, low SI of solid portion on T2WI, and low ADC, all of which corresponded to the imaging features of malignancy phyllodes.

Although we considered the possibility of angiosarcoma because the mass was rapidly growing in a young patient, the enhancement on MRI was different from that of angiosarcoma, which usually shows multiple, intense, nodular enhancement filling from the peripheral portion of the tumor.

High grade ductal carcinoma with hemorrhagic necrosis was also considered, but the margin of the mass was relatively circumscribed, there was no lymph node enlargement, and the mass had grown too rapidly for ductal carcinoma.

Phyllodes tumors should be completely excised. Axillary lymph node dissection is not necessary since axillary lymph node involvement is rare, even when tumors are malignant (1). The management of malignant phyllodes tumor remains controversial, especially in children or adolescent patients. Adjuvant radiation therapy reduces local recurrence in malignant phyllodes tumors, but after considering the patient's young age and the possibility of secondary malignancy following radiation treatment, we decided against radiation therapy and performed completion mastectomy. Chemotherapy is reserved for highly selected patients with large, high-risk, or recurrent malignant phyllodes tumors, and hormonal therapy is not recommended (2, 8).

In conclusion, although very rare, phyllodes tumor should be considered in the differential diagnosis of a breast mass in circumstances of rapid growth, regardless of patient age. MRI findings may be helpful in suggesting the malignant nature of a mass.

Compliance with ethical standards

Conflict of interest: The authors declare that they have no conflict of interest.

Informed consent: This study was approved by our institutional review board with waiver of informed consent (IRB 2019-11-218-001).

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십대 초반 여아에서 발생한 악성 엽상종: 증례보고

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악성엽상종은 십대 초반이나 더 어린 나이에는 매우 드물게 발생한다. 최근 십대 초반의 여아에서 매우 빨리 자라는 거대한 낭성 종괴로 발현했던 악성 엽상종의 영상의학적 소견에 대해 보고하고자 한다.

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