Case Report

Metastatic Recurrent Malignant Phyllodes in 17-Year-Old Female

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Abstract

Malignant phyllodes tumor of the breast is extremely rare but what is rarer is its occurrence in adolescent girls younger than 18 years. It has a propensity for rapid growth and metastases. We present an uncommon presentation of recurrent and metastatic malignant phyllodes tumor in a 17-year-old female.

Keywords: Malignant phyllodes, metastatic, recurrent

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Introduction

Phyllodes tumors, also known as cystosarcoma phyllodes, of the breasts are rare fibroepithelial lesions that represent <1% of all mammary neoplasms.[1] The World Health Organization classifies phyllodes tumors as either benign, borderline and/or malignant according to certain histologic features which include nuclear atypia, mitotic activity, stromal cellularity, stromal overgrowth, and tumor margins.[2,3] Malignant forms of phyllodes tumor represent an extremely rare entity and it constitutes only about 10%–30% of all phyllodes tumors.[4] What is rarer is the occurrence of malignant form of phyllodes tumors in adolescent girls below the age of 18 as the median age at presentation is 45 years.[1] Phyllodes tumors should be regarded as a spectrum of fibroepithelial neoplasms rather than a single disease entity because they display a broad range of clinical and pathological behaviors. At one extreme, malignant phyllodes tumors, if inadequately treated, have a propensity for rapid growth and metastatic spread.[3] With the nonoperative management of fibroadenomas widely adopted, the importance of phyllodes tumors today lies in the need to differentiate them from other benign breast lesions.[1]

Case Report

A 17-year-old female patient presented to the general outpatient department with a 3-month history of the left breast swelling and 1-month history of left breast pain. The swelling was initially small, but it progressively increased in size within 3 months making the left breast twice the size of the right. There were no other visible or palpable swellings in the right breast or other parts of her body and no nipple discharge, bleeding, or wound in both breasts. There was no history of intervention before presentation. There was neither significant constitutional symptoms nor past medical history. She attained menarche at 12 years of age with a regular flow of 4–7 days within a 30-day cycle. She had a positive family history of breast disease in a paternal aunt but none for ovarian disease. She resides with her parents; her mother is a teacher and her father a retired civil servant.

Clinical examination of the left breast showed a centrally located mass that measured 16 cm × 14 cm × 10 cm, was freely mobile, nontender, and with visible distended vessels. There were no palpable masses in the right breast or anywhere else. A clinical impression of giant fibroadenoma was made, and a fine-needle aspiration cytology was done at the pathology department. A diagnosis of atypical most likely benign (C3)' was made. An excisional biopsy was sent to the pathology

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laboratory for analysis.

Grossly, it was a grayish-white mass measuring 20 cm × 10 cm × 5 cm weighing 35 g [Figure 1]. Cut surface showed grayish white mucoid to cystic areas. Microscopic examination revealed infiltrating sheets of spindle- and round-to-oval-shaped cells having hyperchromatic nuclei, prominent nucleoli, and abundant cytoplasm. Focal necrosis, rhabdoid differentiation, and 1–3 mitoses/1 high-power field were seen [Figure 2]. A diagnosis of malignant phyllodes tumor was made with the involvement of all resection margins.

Four weeks later, the patient felt a lump growing back at the excision site. Clinical examination revealed a mass measuring 17 cm × 16 cm with associated peau d'orange. A clinical impression of recurrent malignant phyllodes was made. A simple mastectomy was done and sent to the pathology laboratory for the analysis.

Grossly, we received a mastectomy specimen that measured 22 cm × 16 cm × 18 cm and weighed 1.2 kg. It had a scar that measured 13 cm in length and was 3 cm away from the nipple [Figure 3]. Cut section showed a nodular grayish-white tumor with a diameter of 7 cm. It was surrounded by yellowish fatty tissue streaked by grayish-white tissue.

Microscopic examination revealed a keratinizing pigmented squamous epithelium overlying a tumor composed of sheets of atypical epithelial and mesenchymal stromal cells. The cells showed marked pleomorphism with spindle and round-to-oval nuclei, prominent nucleoli, and moderate cytoplasm. Zonal necrosis, rhabdoid differentiation, and 1–6 mitosis/1 high-power field were noted. The capsule was breached by tumor cells. The tumor margins were devoid of tumor [Figure 4]. A diagnosis of recurrent malignant phyllodes tumor was made.

The patient presented 7 months later with massive right-sided pleural effusion which was completely drained. It reaccumulated 2 weeks after drainage. A chest computed tomography scan was done which revealed massive right-sided pleural effusion.

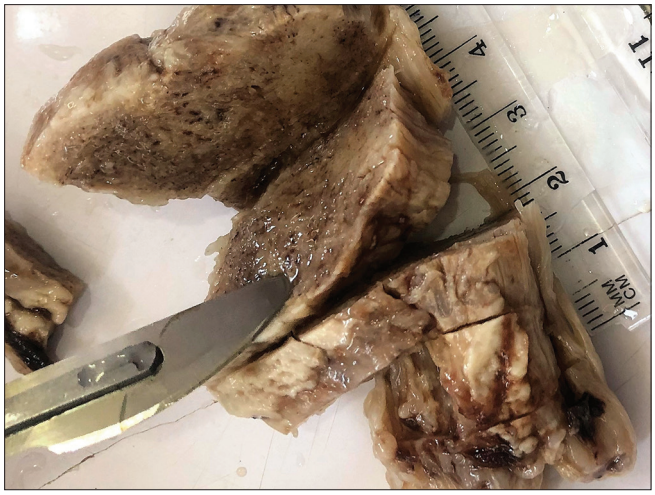

Figure 1: Grayish-white biopsy from the left breast

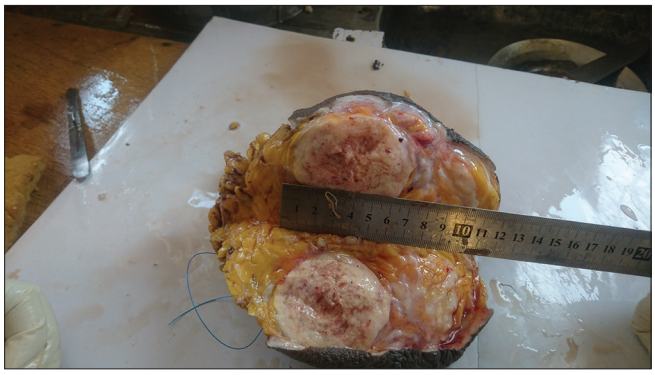

Figure 3: Cut surface of left mastectomy shows a nodular grayish-white tumor with a diameter of 7 cm surrounded by yellowish fatty tissue streaked by grayish-white tissue

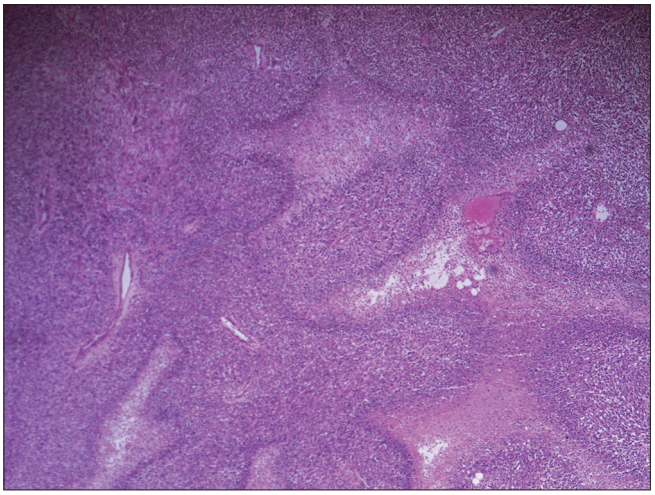

Figure 2: Histological section shows sheets of atypical epithelial and mesenchymal stromal cells displaying a leaf-like pattern, ×4

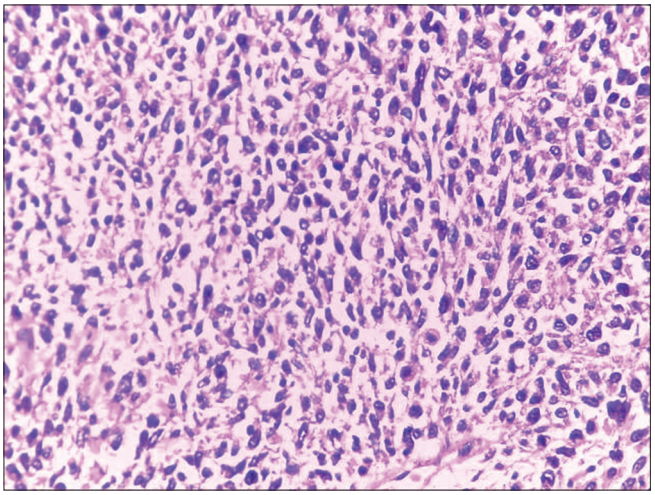

Figure 4: Histological section shows sheets of atypical epithelial and mesenchymal stromal cells showing marked pleomorphism with spindle and round-to-oval nuclei, prominent nucleoli, and moderate cytoplasm, ×100

Right-sided posterolateral thoracotomy was done and a gray-white fleshy mass excised and sent to the pathology laboratory.

Grossly, it was a grayish-white to tan tissue measuring 7 cm × 3 cm × 2 cm a weighing 40 g. Transection showed grayish-white to tan fleshy surfaces [Figure 5].

Microscopic examination revealed infiltrating sheets of pleomorphic round-to-oval shaped to spindle-shaped cells having hyperchromatic vesicular nuclei and moderate cytoplasm. Heterologous stromal differentiation, necroinflammation, and 1–4 mitosis/1 high power field [Figure 6]. A diagnosis of metastatic malignant phyllodes tumor was made with the involvement of all resection margins. Immunohistochemical analysis – CD34 and CD11 – were carried out and are all negative [Figures 7 and 8].

Patient developed generalized edema, severe pallor, chest pain, difficulty in breathing and reaccumulation of right-sided pleural fluid 4 days after surgery. Chest tube was inserted which drained serosanguinous effluent. Packed cell volume was done with a value of 27%. She was transfused with 2 pints of blood but later developed desaturation after oxygen exhaustion. Patient eventually died 2 weeks after surgery.

Discussion

Phyllodes tumors of the breast are generally rare, and malignant cystosarcoma phyllodes is the rarest in the spectrum. It constitutes 0.3%–0.9% of all breast tumors.[5] In the mid-80s, only about 10 cases in individuals under the age of 20 years were described.[6] These tumors are very aggressive and usually have greater tendencies for recurrence.[6,7] They are believed to arise from fibroadenomas, which are the most common breast tumors in children and adolescents.[8] In a series of 821 women diagnosed with malignant phyllodes tumor, the median age at diagnosis was 50 years (range, 12–92 years).[9] However, cases in a 6 and an 11-year-old girls have been reported.[10,11] Malignant phyllodes tumors are the most common primary pediatric breast malignancies closely followed by infiltrating ductal carcinoma, with carcinomas being overall generally better represented than sarcomas.[12] Several studies have reported that phyllodes tumors may

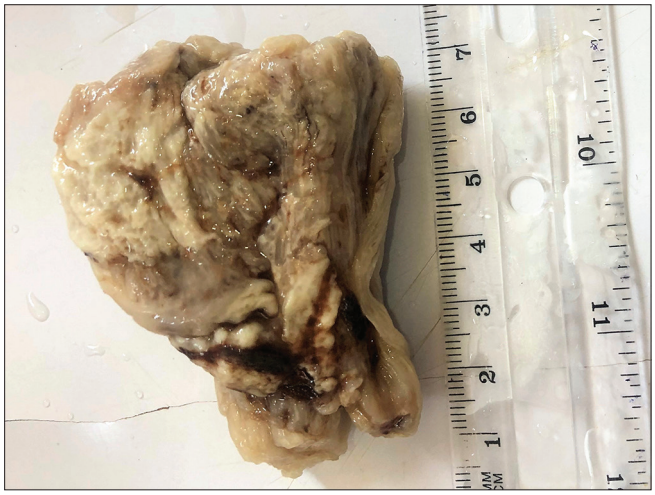

Figure 5: Grayish-white to tan tissue from intrathoracic area

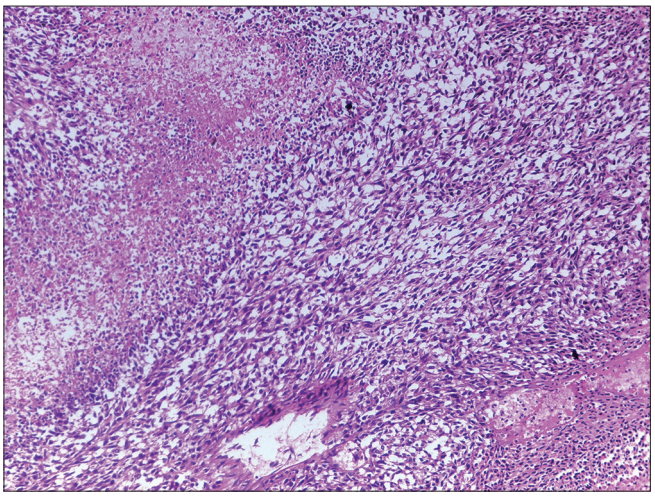

Figure 6: Histological section of intrathoracic lesion shows infiltrating sheets of atypical round-to-oval- to spindle-shaped cells with hyperchromatic vesicular nuclei and moderate cytoplasm, ×100

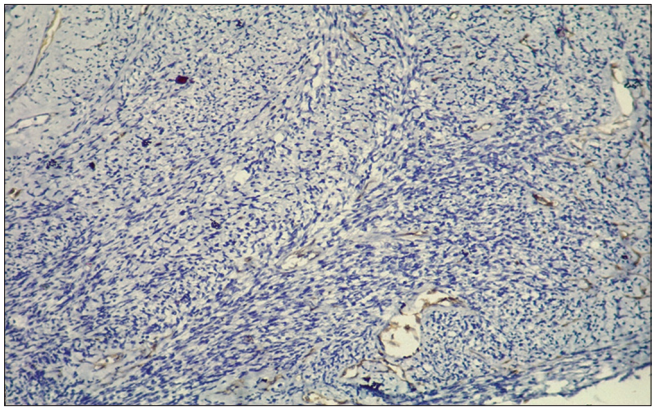

Figure 7: Photomicrograph showing no membranous staining in almost 100% of malignant cells (CD 34) ×10

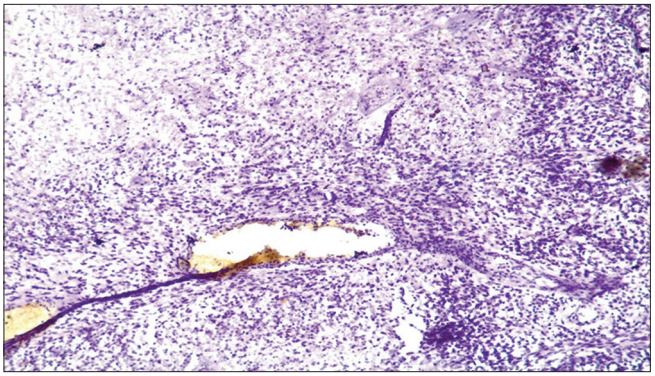

Figure 8: Photomicrograph showing malignant phyllodes tumor demonstrating no cytoplasmic CD117 immunostaining × 10

vary between 0.5 and 27 cm with a mean between 5 and 7.2 cm. It presents as a rounded nodule or multiple nodules, usually mobile, painless with a rapid growth pattern.[9,12,13] Our patient is a 17-year-old that presented with a rapidly growing left breast lump of 3 months duration that measures 20 cm × 10 cm × 5 cm.

Imaging studies such as mammography, ultrasonography, and magnetic resonance mammography (breast magnetic resonance imaging) are often used in the diagnosis of breast neoplasms. However, these techniques are not specific in the diagnosis of malignant phyllodes tumor and other breast tumors.[14,15]

Histologically, these tumors have a biphasic growth pattern comprising of epithelial and mesenchymal components. Stromal cellularity and mitotic figures help classify these tumors from benign to borderline to malignant. Cystic changes, necrosis, and hemorrhage may be seen in large tumors.[7,9,12,15]

Risk factors for local recurrence include tumor diameter, stromal overgrowth/mitosis, and resection margins. Asoglu *et al*. found no significant difference in terms of local recurrence when wide local excision or mastectomy was performed for benign and malignant forms but larger tumors and those with stromal overgrowth tend to recur.[16] Previous studies have reported a 5-year overall survival rate of 72%–86%.[7,12,16,17]

Surgical removal remains the treatment of choice for phyllodes tumors. In cases of malignant phyllodes tumor or local recurrence, mastectomy becomes very necessary.[5,9,12,15] The role of radiotherapy in the treatment of malignant phyllodes tumors is vague because of few reported cases. The function of adjuvant chemotherapy in phyllodes tumors is not fully established.[7,16,18] The index patient had surgical excision of the tumor including all the involved margins, but it later recurred which necessitated for mastectomy. She had no demonstrable nodal involvement.

Extension into the chest from local recurrence or from metastasis to the lungs and bones is the most common cause of death among patients.[7,16,19] Our patient had pleural effusion from the deposition of tumor cells into the chest. She died of metastatic disease 8 months later.

Conclusion

Malignant phyllodes tumor is rare and seldom constitutes a diagnostic dilemma, especially among adolescents. Therefore, it should be considered in the differential diagnosis of breast lumps in females regardless of age or ethnicity, specifically for those tumors associated with rapid growth.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Parker SJ, Harries SA. Phyllodes tumours. Postgrad Med J 2001;77:428-35.
2. Lakhani SR, Ellis IO, Schnitee SJ, Tan PH, van de Vijver MJ. World Health Organization Classification of Tumours of the Breast. Lyon: IARC; 2012.
3. Mishra SP, Tiwary SK, Mishra M, Khanna AK. Phyllodes tumor of breast: A review article. ISRN Surg 2013;2013:361469.
4. Belkacémi Y, Bousquet G, Marsiglia H, Ray-Coquard I, Magné N, Malard Y, *et al*. Phyllodes tumor of the breast. Int J Radiat Oncol Biol Phys 2008;70:492-500.
5. Rowell MD, Perry RR, Hsiu JG, Barranco SC. Phyllodes tumors. Am J Surg 1993;165:376-9.
6. Cornelius IC, Anas ME, Leopoldo L. Fetal metastatic cystosarcoma phyllodes in an adolescent female: Case report and review of treatment approaches. J Surg Oncol 1986;33:176-81.
7. Ben Hassouna J, Damak T, Gamoudi A, Chargui R, Khomsi F, Mahjoub S, *et al*. Phyllodes tumors of the breast: A case series of 106 patients. Am J Surg 2006;192:141-7.
8. Noguchi S, Yokouchi H, Aihara T, Motomura K, Inaji H, Imaoka S, *et al*. Progression of fibroadenoma to phyllodes tumor demonstrated by clonal analysis. Cancer 1995;76:1779-85.
9. Macdonald OK, Lee CM, Tward JD, Chappel CD, Gaffney DK. Malignant phyllodes tumor of the female breast: Association of primary therapy with cause-specific survival from the Surveillance, Epidemiology, and End Results (SEER) program. Cancer 2006;107:2127-33.
10. Sasa M, Morimoto T, Ii K, Tsuzuki H, Kamamura Y, Komaki K, *et al*. A Malignant phyllodes tumor of the breast in a 6-year old girl. Breast Cancer 1995;2:71-5.
11. Sorelli PG, Thomas D, Moore A, Khan M, Hoque H. Malignant phyllodes tumor in an 11-year-old premenarchal girl. J Pediatr Surg 2010;45:e17-20.
12. Gutierrez JC, Housri N, Koniaris LG, Fischer AC, Sola JE. Malignant breast cancer in children: A review of 75 patients. J Surg Res 2008;147:182-8.
13. Juliana AS, Elvira FM, Camila G, Deíse SG, Thiago Q, Luciana G, *et al*. Malignant phyllodes tumor of the breast: Case report. Rev Assoc Med Bras 2011;57:495-97.
14. Yilmaz E, Sal S, Lebe B. Differentiation of phyllodes tumors versus fibroadenomas. Acta Radiol 2002;43:34-9.
15. Tian W, Xu D. Diagnosis and management of multiple carcinosarcoma of the breast in a young Chinese patient. Breast Care (Basel) 2012;7:147-9.
16. Asoglu O, Ugurlu MM, Blanchard K, Grant CS, Reynolds C, Cha SS, *et al*. Risk factors for recurrence and death after primary surgical treatment of malignant phyllodes tumors. Ann Surg Oncol 2004;11:1011-7.
17. Jang JH, Choi MY, Lee SK, Kim S, Kim J, Lee J, *et al*. Clinicopathologic risk factors for the local recurrence of phyllodes tumors of the breast. Ann Surg Oncol 2012;19:2612-7.
18. Guillot E, Couturaud B, Reyal F, Curnier A, Ravinet J, Laé M, *et al*. Management of phyllodes breast tumors. Breast J 2011;17:129-37.
19. Guerrero MA, Ballard BR, Grau AM. Malignant phyllodes tumor of the breast: Review of the literature and case report of stromal overgrowth. Surg Oncol 2003;12:27-37.