Borderline phyllodes tumor of breast in a premenarchal girl: A relatively common tumor at an uncommon age

Akhil Kapoor1, Ishwar Charan2, Mukesh Kumar Singhal1, Harvindra Singh Kumar1

1Department of Radiation Oncology, Acharya Tulsi Regional Cancer Treatment & Research Institute, Sardar Patel Medical College, Bikaner, Rajasthan, India
2Department of Surgery, Sardar Patel Medical College, Bikaner, Rajasthan, India

Received December 11, 2015; Revised May 15, 2016; Accepted May 25, 2016; Published Online June 5, 2016

Case Report

Abstract
Phyllodes tumors are relatively rare breast lesions that usually occur in the age group of 35 – 55 years. It is a very rare diagnosis in young girls, particularly at prepubertal age. Because of the uncommon nature of this tumor in children, it may be misdiagnosed leading to inappropriate management. We report a case of a 9-year-old girl who was diagnosed as a case of borderline phyllodes tumor left breast. Simple mastectomy without axillary staging was performed. She has recovered well and is on follow up.

Keywords: Phyllodes tumor; Breast; Prepubertal girl

1. Introduction
Phyllodes tumors, also known as cystosarcoma phyllodes, are rare fibroepithelial lesions accounting for 0.3 to 0.5% of breast tumors in females.1 It is a relatively large tumor with smooth, sharply demarcating margins, the average size being 5 cm. Phyllodes tumor usually occurs in the age group of 35 - 55 years and is very rare in younger age.2

In most of previously published reports of phyllodes tumor in young females, the patients are adolescent and have attained menarche.3 Because of the uncommon presentation of this tumor in children, it may be misdiagnosed leading to inappropriate management. We present a case of a 9- year-old premenarchal girl presenting with a very large lump in her left breast which was subsequently diagnosed as borderline phyllodes tumor.

2. Case Presentation
A 9-year-old girl presented to our department with the complaint of painless, rapidly growing left breast mass from 4 months. On examination, the patient had achieved thelarche. However, menarche was not achieved. The lump measured approximately 12 × 10 cm in size occupying the entire left breast. Dilated cutaneous veins on this huge left breast mass were seen. No axillary lymph nodes were palpable. Ultrasound of the breast showed lobulated, well circumscribed mass with smooth margins, echogenic rim, and low level homogenous internal echoes (Figure 1).

Fluid - filled cleft like cystic spaces with good thorough transmission were noted inside the mass; however no microcalcification were seen. Posterior acoustic enhancement and vascularity within solid component was seen. Mammography was not done as the patient was young. True cut biopsy was performed from the mass and it showed fibroepithelial lesion with hypercellular stroma and moderate pleomorphism (Figure 2). The mitotic rate was 8 per 10 hpf. The overall picture was suggestive of borderline phyllodes tumor. On further study, the tumor stained negatively for estrogen receptor (ER) and showed weak proliferating cell nuclear antigen (PCNA) staining. As the mass occupied entire breast, it was not possible to obtain negative margins with wide local excision alone. Hence, simple mastectomy was performed. Grossing of the specimen showed grey meaty consistency with fibrogelatinous and hemorrhagic areas with leaf like protrusions into the cystic space (Figure 3). On microscopy, the findings were similar to that of core needle biopsy with no new findings. All the margins were free from tumor with closest margin being the deep excision margin (1 cm). The surrounding breast tissue showed normal ducts and stroma. Her post-operative recovery was uneventful.
The term cystosarcoma phyllodes is derived from the Greek words *sarcoma* ("fleshy tumor"), and *phyllon* ("leaf"). Though the tumor grossly displays characteristics of a large, malignant sarcoma, leaf-like appearance is seen when it is sectioned. Al so, it displays epithelial, cyst-like clefts when viewed histologically. Since most of the tumors are benign, the name cystosarcoma is misleading. Hence, nowadays the preferred terminology is phyllodes tumor. It is typically a large, fast growing mass that arises from the periductal stroma of the breast.

Table 1 shows the criteria for classification of phyllodes tumor as proposed by Salvadori *et al.* and Azzopardi *et al.* In our patient, the margins were well circumscribed, pushing type suggestive of benign disease. However, the stromal cellularity and pleomorphism, both were moderate. The mitotic rate was also 8 per 10 high power field; all these features supported the diagnosis of borderline phyllodes tumor in our patient.

Mastectomy is the most reliable procedure with regard to local control, but breast-conservation surgery is preferred, except for very large tumors. In our patient, since the entire breast was replaced by the mass, simple mastectomy was done. Though most of the tumors are benign, there is predilection for local recurrence especially in incompletely resected tumors. The local recurrence rate varies from 10 - 40% with the surgical margin being the only independent predictive factor in multivariate analysis. In most of the cases, the local recurrence is isolated in absence of any distant metastases. In our patient, all the margins were free with the closest margin being 1 cm; hence, the patient was at relatively lower risk of recurrence. Approximately 10% of patients with phyllodes tumors develop distant metastases and these eventually occur in about 25% of patients with malignant histology. Also, the presence of mixed stromal components such as chondro and osteosarcomatous differentiation puts the patient at higher risk of distant metastasis. Such features were fortunately absent in our patient.

### Table 1: Criteria for classification of phyllodes tumor as proposed by Salvadori and Azzopardi.

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Histological Type</th>
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<tbody>
<tr>
<td></td>
<td>Benign</td>
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<tr>
<td>Tumor margins</td>
<td>Pushing</td>
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<tr>
<td>Stromal cellularity</td>
<td>Low</td>
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<tr>
<td>Mitotic rate (per 10 hpf)</td>
<td>&lt;5</td>
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<tr>
<td>Pleomorphism</td>
<td>Mild</td>
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3. Discussion
Though some studies advocate adjuvant radiotherapy in borderline and malignant cases, we followed MD Anderson Cancer Center guidelines according to which radiotherapy is recommended only for cases with positive or near-negative surgical margins and further surgical intervention is not possible.

Another important differential diagnosis of breast lump in a prepubertal girl is fibroadenoma. Goyal et al. presented a case of bilateral giant juvenile fibroadenoma in a 11-year-old-pre-pubertal girl. In a study of cost benefit ratio by Cattin et al, they concluded that diagnostic interventions even if more expensive at the beginning, allows in some cases to avoid further interventions. Thus, tru-cut biopsy, though more expensive and time-consuming than conventional fine needle aspiration cytology was preferred in our patient.

4. Conclusion
The physicians and surgeons should be made aware of breast diseases in adolescents and young children. This report shows that phyllodes tumour can affect children and greater awareness would ensure early presentation to hospital for appropriate treatment.

Conflict of interest
The authors declare that they have no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

Acknowledgement
The authors acknowledge the support of department chairs and also would like to thank the parents of the child to provide the permission to use the data for publication purposes.

References