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Case Report

Benign Phyllodes tumor in an 11-year-old premenarchal girl

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Abstract

Phyllodes tumors are rare fibroepithelial tumors that account for less than 0.5% of all breast tumors. Presentation in children is even rarer. In this paper, we describe a case of an 11-year-old adolescent girl with a benign phyllodes tumor. The rare presentation at this age, its distinguishing features, the preoperative diagnostic difficulties, and the management protocols, surgical resection and reconstructive approach of this uncommon tumor may provide useful insight and perspective for patients and clinicians facing similar challenges in the paediatric population.

Keywords: Cystosarcoma phyllodes, Breast lump, Premenarchal girl, Benign tumors

Introduction

Breast lumps are uncommon in children. The most common type of breast mass is fibroadenoma found in the adolescent population. Phyllodes tumors are rare fibroepithelial tumors that account for 0.3 - 0.5% of all breast tumors in females. They are rarely observed in adolescents, with only 20 cases reported[1]. Here, we report a case of a left breast lump in a 11-year premenarchal girl whose histopathology proved a benign phyllodes tumor.

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Case summary

An 11-year-old otherwise healthy premenarchal girl presented to the outpatient department of paediatric surgery regarding a rapidly growing protruding mass from areolar region of left breast one month prior to consultation. The patient denied any redness or thickening, nipple discharge, nipple inversion. The right breast was normal. She had no previous history of breast biopsies or abnormal breast imaging. There was no additional symptoms such as bone or rib pain, headache, shortness of breath. Her family history was insignificant. Local examination revealed a 6 cm x 5 cm x 3 cm, lobulated lump having multiple papillary fronds-like structures, occupying the lower and inner quadrant of the areola of the left breast (Fig.-1). There was no axillary, infraclavicular, or supraclavicular lymphadenopathy appreciated on examination. The patient underwent an excision biopsy using an elliptical incision 2-mm from the margin of the mass. A 6.5 cm x 6 cm x 2 cm lobulated lump having multiple papillary fronds-like structures was excised. The histopathologic analysis revealed that it was a benign phyllodes tumor (low grade) composed of a stromal and glandular component. The stromal component is seen to be over grown than the glandular component, having monomorphic spindle to round nuclei. No mitosis or necrosis is seen. She has been doing well since the time of surgery. Follow-up ultrasonography was normal . Chemotherapy was not currently indicated, and opted for close observation. Workup for metastatic and recurrent disease has been negative to date (Fig.-2).

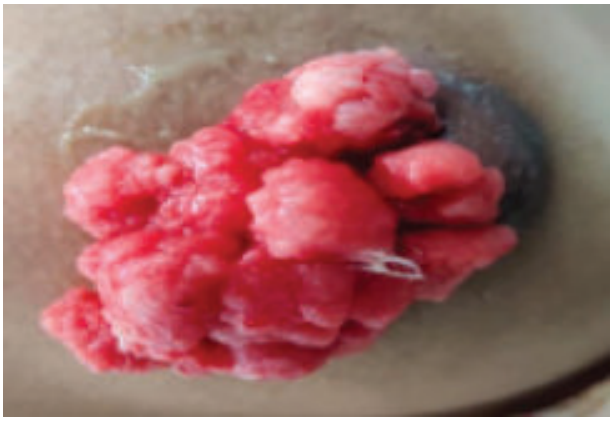

Fig.-1. *Clinical photograph of the patient showing the left breast lump (Source: from Dhaka Medical College Hospital)*

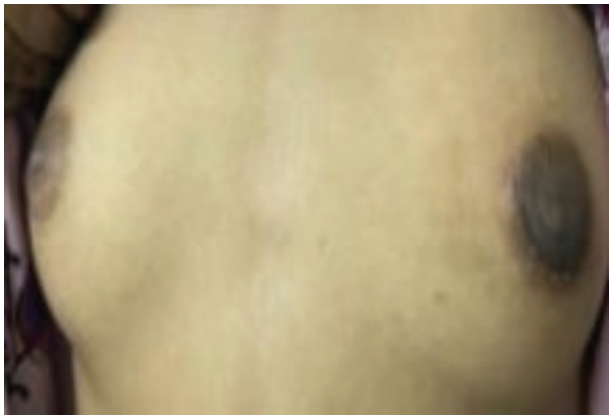

Fig.-2. *Clinical photograph of the patient showing both breast after excision biopsy of the left breast lump (Source: from Dhaka Medical College Hospital)*

Discussion

An adolescent presenting with a breast mass differs substantially from that of an adult due to the differences in breast cancer risk and breast architecture. There is less emphasis on exclusion of malignancy. Paediatric breast masses are typically benign (95% benign fibroadenomas)[2].

Management of paediatric breast masses is primarily conservative. Clinical observation over two to four months is appropriate. Masses that increase by more than 1 cm and those larger than 2 cm warrant ultrasonographic percutaneous biopsies to confirm the benign nature[2]. Phyllodes tumors are rare fibroepithelial tumors that account for 0.3 — 0.5% of female breast tumors, the peak of which occurs in women between the ages of 45 and 49 years[3].This type of tumor is rarely found in adolescents. Only about 20 cases have been reported in children[1].

A large breast lump with history of rapidly increasing size and ultrasound features suggestive of a fibroadenoma (except a size > 2 cm) should arouse high suspicion of a phyllodes tumor[4]. Axillary lymphadenopathy is rare. Another characteristic feature of these tumors is a high rate of local recurrences (5–20%)[5]. Fibroadenomas and phyllodes tumors share many common features. Clinically, both present as rounded, circumscribed, and moveable masses. Histologically, both can be grouped as "fibroepithelial lesions". Preoperative diagnosis poses a diagnostic difficulty, as fine needle aspiration cytology and core needle biopsy may not be able to distinguish a phyllodes tumor from a fibroadenoma[4] .

Microscopically, phyllodes tumors are characterized by a double-layered epithelial component arrayed in clefts and surrounded by a hypercellular stromal mesenchymal component. The stroma often protrudes into the epithelial lining spaces, forming a slit-like space or a leaf-like pattern; hence, the name is phyllodes. The morphologic features that have to be accounted for are the following: 1) the degree of stromal hypercellularity, 2) stromal overgrowth, 3) nuclear atypia, 4) number of mitoses, 5) amount of stroma relative to epithelium, and 6) infiltrative tumor borders. However,the natural history of these tumors is often different, Phyllodes tumors tend to grow more rapidly and may recur if incompletely excised. Moreover, phyllodes tumors may metastasize. In contrast, fibroadenomas usually do not need to be removed, and even when surgery is needed, enucleation is sufficient.

Excisional biopsy is required in the majority of cases of suspected phyllodes tumors. The World Health Organization (WHO) classifies them as either benign (pushing type margin, minimal stromal atypia >5 mitosis/10 HPF, absence of stromal overgrowth), borderline (pushing/infiltrating margin, moderate stromal atypia,<10 mitosis/10 HPF, absence of stromal overgrowth) or malignant (infiltrating margin, severe stromal atypia>/=10 mitosis/10 HPF , presence of stromal overgrowth)based on histopathological features[6] .Recurrence among benign, borderline, and malignant phyllodes tumors is 10–17%, 14–25%, and 23– 30%, respectively[7]. Malignant tumors must have confirmed negative margins [8,9] . The rate of distant metastasis for the

malignant tumors is 15–25% [5].The benign variant is most common, with only 10-25% of cases being malignant. In a review of 36 malignant phyllodes tumors, 11 (30.6%) were given a diagnosis of primary fibroadenomas and experienced recurrence as malignant phyllodes tumors [10] .

Phyllodes tumors are managed by wide local excision. In cases of large lumps, a mastectomy may be necessary. Yom et al concluded that a clear margin of 0.1 mm is equivalent to a margin of 1 cm[11]. Adjuvant therapy has no proven effect. In cases of systemic metastasis, treatment is based on the soft tissue sarcoma protocol[12].

Reconstructive surgery is usually not considered until at least one year after the procedure and after the patient has reached skeletal maturity due to postoperative cosmetic deformity or secondary asymmetry. The breast parenchyma may expand to fill the resulting defect and resolve any deformities. Surgeons should discuss all potential outcomes. Adolescents are said to have a higher rate of recurrence and therefore should be observed closely with routine follow-up.

Conclusion

When a premenarchal girl presents with a large breast lump, the possibility of a phyllodes tumor should be considered, though a rare differential to prevent misdiagnosing and aggressive tumor. A case-based, individualized approach is recommended. Due to the extreme rarity and the frequently benign nature of tumors in this age group, a more conservative approach with regular follow up is advisable. In young adolescent girl, this prevents cosmetic and psychological distress .

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Conflict of Interest: None

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