In instructive case

An adolescent with a phyllodes tumor: A case report and review

Deepa Makhija, Hemanshi Shah*, Jyoti Bothra, Shalika Jayaswal

Dept. of Paediatric Surgery, TNMC & BYL Nair Hospital, Mumbai Central, Mumbai, Maharashtra, Pin: 400008, India

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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 adolescent with a phyllodes tumor. The rare presentation at this age, its distinguishing features, the preoperative diagnostic difficulties, and the management protocols of this uncommon tumor are highlighted.

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1. Introduction

Breast lumps are uncommon in children. The most common type of breast mass found in the adolescent population is a fibroadenoma. 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 an adolescent with a lump in her left breast which the histopathology revealed to be a benign phyllodes tumor.

2. Case summary

An 11-year-old pre-menarcheal girl presented with a lump in her left breast that was gradually increasing in size. Local examination revealed a 15 × 15 cm, well-circumscribed, firm, and freely mobile lump occupying the entire left breast (Fig. 1). The right breast was normal, and there was no axillary lymphadenopathy. Ultrasound revealed a 9 × 5 × 6 cm homogenous, hypoechoic, and encapsulated lesion with minimal vascularity and no calcification (Fig. 2). Cytology was suggestive of a fibroadenoma. The patient underwent an excision biopsy using an inframammary incision. A 9 × 9 cm encapsulated lump weighing 250 gm with smooth surfaces was excised. The histopathologic analysis revealed that it was a benign phyllodes tumor with margins reaching the inked surface (Fig. 3).
3. Discussion

The evaluation of an adolescent presenting with a breast mass differs substantially from that of an adult because of marked differences in breast cancer risk and breast architecture. There is less emphasis on exclusion of malignancy, as pediatric breast masses are typically benign (95% benign fibroadenomas) [2].

Management of pediatric 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].

According to Stanford University, the following criteria for juvenile fibroadenoma are used: 1] circumscribed and rarely multiple; 2] biphasic stromal and epithelial process lacking a leaf-like growth pattern in a uniformly hypercellular stroma; 3] a lack of atypical features and a stroma-like periductal increase in cellularity, stromal overgrowth, and cytologic atypia, as well as a mitotic rate <3/hpf; 4] frequent epithelial and myoepithelial hyperplasia; and 5] an age of between 10 and 20 years [3].

Giant fibroadenomas are defined as tumors >500 g. 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 [4]. 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 [5]. Axillary node involvement is rare. Another characteristic feature of these tumors is a high rate of local recurrences (5—20%) [6].

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 [5].

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 phyllodes, which means “leaf-like” in Greek. 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, and the differences are tabulated in Table 1. 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, borderline, or malignant based on histopathological features [7] (Table 2).

The benign variant is most common, with only 10–25% of cases being malignant. The rate of distant metastasis for the malignant tumors is 15–25% [6].

Phyllodes tumors are managed by wide local excision. In cases of large lumps, a mastectomy may be necessary. A recent study by Yom et al concluded that a clear margin of 0.1 mm is equivalent to a margin of 1 cm [8]. Due to the rarity of the condition in younger age groups, an individualized, case-based approach, and regular follow up are advisable. As per the National Comprehensive Cancer Network (NCCN) guidelines, in cases with local recurrence, resection with wide, tumor-free surgical margins should be performed. Adjuvant therapy has no proven effect. In cases of systemic metastasis, treatment is based on the soft tissue sarcoma protocol [9].

Patients may experience postoperative cosmetic deformity or secondary asymmetry. However, reconstructive surgery is usually not considered until at least one year after the procedure and after the patient has reached skeletal maturity. Additionally, the breast parenchyma may expand to fill the resulting defect and resolve any deformities over the course of development. Surgeons should discuss all potential outcomes, expectations of aesthetic results, and the need for reconstructive surgery with patients prior to surgery and at subsequent follow-up appointments [10].

4. Conclusion

When an adolescent presents with a large breast lump, the possibility of a phyllodes tumor, though a rare differential, should be considered. A case-based, individualized approach is recommended, as there are no set protocols. Due to the extreme rarity of this tumor and because of the frequently benign nature of tumors in this age group, a more conservative approach with regular follow up is advisable. This prevents cosmetic and psychological distress in young adolescent girls.

Conflict of interest

The authors have no conflict of interest to report.

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Department of Histopathology, Tata Memorial Hospital, Mumbai, Maharashtra, in India.

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