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JUVENILE GIANT FIBROADENOMA: A CASE REPORT

Clinical Research

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ABSTRACT

Fibroadenomas are common benign breast tumors. Fibroadenomas that exceed 5 cm in diameter, weigh more than 500 g, or replace more than four-fifths of the breast are characterized as giant. A fibroadenoma diagnosed in patients during childhood or adolescence is characterized as juvenile. Patients with giant juvenile fibroadenoma presented at a mean age of 13.92 years and usually after menarche. Juvenile fibroadenomas are usually unilateral, occurring either in the right or the left breast; the majority of them are noticed and diagnosed late when their size is already more than 10 cm and are most frequently treated with total lump excision. Differential diagnosis includes phyllodes tumors and pseudo-angiomatous stromal hyperplasia. Surgical excision is recommended to patients with suspicious imaging features or when the mass grows rapidly or for cosmetic reasons.

KEYWORDS

Juvenile Fibroadenoma; Giant Fibroadenoma; Adolescence; Breast Disease

INTRODUCTION

Fibroadenomas are common breast tumours in children and adolescents, although it is rare among paediatric and adolescent patients, and its presentation is different when compared to adults. Fibroadenomas represent 30–50% of palpable breast masses during childhood and adolescence and 44–94% of operated cases are in the same age group [1,2]. A fibroadenoma can be characterized as gigantic or giant if its diameter exceeds 5 cm, weighs more than 500 g, or replaces more than four-fifths of the breast [1,2]. Majority of fibroadenomas reported among minors are giant, even though they represent a rare clinical entity. When a fibroadenoma is diagnosed in patients between 10 and 18 years old, it is characterized as juvenile fibroadenoma [3]. A broader definition of juvenile fibroadenoma includes all cases in childhood and adolescence, up to 19 years on the definition of adolescence by the World Health Organization (WHO) [4]. Fibroadenomas are benign masses consisting of oestrogen-sensitive epithelial and stromal tissue and are not a pre malignant lesion. Malignancy in a pre-existing juvenile fibroadenoma is very rare [5]. However, in women with a significant family history of breast cancer and in cases of proliferative changes in the histopathologic examination of the mass, an increase in breast cancer risk has been reported [6]. Overall, breast cancers in adolescents are extremely rare accounting for 0.1% of all breast cancers in women regardless of age and less than 1% of all paediatric cancers [1].

Case Report

A case of an 13-year-old girl, who presented in our centre with a palpable mass in her both her breasts (Rt >> Lt). The mass was identified during self-palpation 6 months before presentation, as it increased rapidly in size. However, no other complaints such as pain, fever, dyspnea, cough, or loss of appetite were reported. No significant family history was noticed. Upon clinical examination, an asymmetry of the breasts was evident, with the right breast having greater size than the left one. However, no skin discoloration or discharge from the nipples was observed. During palpation on the right side, a firm solitary mass with discrete margins involving whole of the breast was detected. On the left two small masses of approximate 1-2 cm size was detected. No axillary lymph nodes were palpated. Residual systemic examination was unremarkable.

Ultrasound examination of the breast showed a hypoechoic mass involving entire right breast. Its maximum diameter measured 6 cm, with no significant internal vascularity.

Fine-needle aspiration cytology (FNAC) of the mass shows highly cellular smear with epithelial hyperplasia and increased stromal cellularity. Malignancy cannot be ruled out.

Subsequently, the patient was admitted and underwent a total surgical excision of the mass (Figure 1-4). Reconstruction was not required as no visible asymmetry was

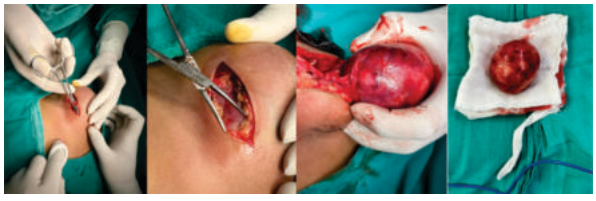

Figure 1-4. Total surgical excision of the fibroadenoma without reconstructive surgery.

The histopathologic evaluation (HPE) of the surgical specimen confirmed the diagnosis of giant juvenile fibroadenoma. The macroscopic examination revealed a single large globular soft to firm tissue mass measuring 8 x 6 x 5.5 cm. External surface is grey brown to grey white. Cut surface is smooth, solid and grey white and shows multiple slit like spaces. Microscopically, sections examined show peri - canalicular growth pattern with epithelial hyperplasia and increased stromal cellularity. No evidence of atypia / malignancy noted in the sections examined.

During the follow-up consultation on 10th post operative day, stitches were removed and no wound gaping was noticed. However, patient lost to follow up and didn't reported for next visit.

DISCUSSION

The most common breast tumors in children and adolescents are fibroadenomas, which are benign tumors with glandular and stromal components. Typical clinical findings include a painless, slowly enlarging mass that may cause breast enlargement or asymmetry. Clinical examination reveals a usually smooth, firm, non-tender mass that is mobile during palpation. Nonetheless, a fibroadenoma might occasionally be painless and present as a rapidly growing mass. With regard to age at diagnosis of giant juvenile fibroadenomas, Michael Sosin et al., in a systematic literature review based on 153 patients, reported a mean age of 16.7 years; among these cases, the mean lesion diameter was 11.2 cm [7].

The precise etiology of juvenile fibroadenomas remains unknown. Reproductive hormones could play a role, since estrogen and progesterone receptors are expressed in fibroadenomas, and these lesions occur more frequently in puberty, pregnancy, and in individuals who take oral contraceptives [13]. Genetic predisposition also plays a role, since fibroadenomas appear more commonly in African-American females, and occasionally patients report positive family history of breast fibroadenomas[8]. With regard to imaging, fibroadenomas appear as hypoechoic during ultrasonography and show diffuse enhancement on magnetic resonance imaging (MRI). Typical diagnostic workup includes patient history, physical examination, and radiographic evaluation with ultrasonography being

the reference standard technique [7]. A study by Smith et al. showed that in 78.8% of fibroadenoma cases, histopathology confirmed the diagnosis made through ultrasound, whereas color Doppler revealed non-vascularization or minimal internal vascularity in most cases [14]. The differential diagnoses in cases of giant juvenile fibroadenomas include phyllodes tumors and pseudoangiomatous stromal hyperplasia. Of note, the majority of fibroadenomas naturally grow slowly and eventually regress before they reach 5 cm in diameter. Fibroadenomas in adolescents are not considered as a premalignant state, even though some reports have suggested the possibility of rare malignant transformation, and metachronous lesions are frequently reported [10]. In particular, the risk of malignant degeneration of a juvenile fibroadenoma is reported to be less than 0.3%, and the most common fibroadenomarelated malignancy is lobular malignant tumor [11,15]. Histopathologically, proliferative changes, such as sclerosing adenosis, duct epithelial hyperplasia, epithelial classification, and papillary apocrine changes are thought to be indicative of increased breast cancer risk [16]. Many authors suggest that pediatric and adolescent patients with fibroadenomas, presenting typical findings, both clinical and ultrasonographic, can be managed conservatively rather than surgically. The surgical excision of a fibroadenoma can cause scarring at the incision site, dimpling of the breast, duct system damage, and ultimately mammographic post-surgical changes that lower the diagnostic accuracy of mammography later in life. In addition, a recurrence is expected in 10–25% of the patients [17]. In cases managed conservatively, a safe management option is to regularly follow up the mass with ultrasound in order to confirm the stability of the lesion over time. With regard to surgical management of juvenile fibroadenomas, the indications include suspicious imaging characteristics or rapid growth. In fact, juvenile fibroadenomas, especially giant, are commonly treated with excision because of their rapid growth in contrast to typical fibroadenomas that appear in adulthood [7]. In the majority of giant juvenile fibroadenoma cases, lump excision is surgically feasible. Mastectomy is performed in very rare cases of recurrent giant fibroadenomas, and surgeons will have to take into consideration the potential morbidity, both physical and psychological [7]. Mammoplasty should be performed after the mastectomy to minimize the psychological trauma. Prosthesis or autologous tissue reconstruction may be used [12]. Another possible treatment method that has been described in the literature is cryoablation, which can be performed after core needle biopsy: however, the available data refer only to adult fibroadenomas [18-20]. Juvenile fibroadenomas are a separate and distinguished clinical entity in comparison to adult fibroadenomas, and their management may differ. As mentioned above, reports of malignant degeneration of a juvenile fibroadenoma are extremely rare [5]. Adult fibroadenomas, however, have been considered as a long-term risk factor for the development of breast cancer. A study published in the New England Journal of Medicine showed that the risk of invasive breast cancer was 2.17 times higher in women with fibroadenomas in comparison to the controls (CI 95%M 1.5 to 3.2), and this risk was found to be higher in patients with complex fibroadenomas, proliferative disease or family history of the disease [6,21]. This possible association is also supported by a clinicopathologic study of women with carcinomas that developed within fibroadenomas, where the mean age of such patients was higher in comparison to patients with benign fibroadenomas [21]. For this reason, a mammography is also recommended to older women to exclude cancerous lesions. In mammography, fibroadenomas appear as well-circumscribed, homogeneous nodules, which may include inner calcifications [21]. Because of the increased incidence of carcinoma in older women, most clinicians recommend the excision of persistent breast masses, such as fibroadenomas, as patients approach adulthood [22]. In younger patients, conservative management with frequent clinical evaluations can be more safely performed; in fact, several investigators suggest that the cut-off age should be 25 [21]. Even though the breast cancer risk has been well described in the literature to differ between fibroadenomas in adolescence and adulthood, the underlying cause has not been explored. One could argue that the aforementioned findings might reflect the well-known increasing risk of breast cancer with age. On the other hand, it is possible that distinct genetic profiles of juvenile versus adult fibroadenomas contribute to the aforementioned difference in breast cancer risk. The management of giant fibroadenomas also differs from that of simple fibroadenomas. Giant fibroadenomas appear to be more cellular and have less lobular components in comparison to simple fibroadenomas [21]. Since they are larger in size, they can compress normal breast tissue, causing greater symptoms of discomfort and pain, and therefore, they are more commonly excised. Another reason for this is that giant fibroadenomas cannot be easily distinguished from phyllodes tumors through physical examination or ultrasound [23]. On the other hand, management of simple fibroadenomas entails careful follow-up, since most simple fibroadenomas in adolescents decrease in size and may even disappear with time [24].

CONCLUSIONS

Enlarging breast masses in children and adolescents can be of great concern to young patients, and fibroadenomas should be included in the differential diagnosis. Generally, the management of breast disease in children and adolescents is different from that in adults. Special surgical consideration needs to be placed on maintaining lactation ability, preserving the developing breast parenchyma through minimal injury of the breast, and achieving good cosmetic results because of the patients' young age. A further consultation with a plastic surgery specialist may also be required, especially in those cases when breast symmetry is not easily achieved after a standard lump excision. Patients need to receive appropriate counseling and to be closely followed up for future breast masses or abnormalities that may form as they progress through adolescence and adulthood. In summary and based on our review, the authors conclude that ultrasonography is the most common method of evaluation and total lump excision is the most common treatment option. Therefore, ultrasonography is recommended as an initial diagnostic tool. However, the currently available data do not suffice for the purpose of proposing standard recommendations for adolescent patients presenting with such breast lesions.

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