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Recurrent giant juvenile fibroadenoma

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ABSTRACT

Breast masses in children, though rare, present a difficult clinical challenge as they can represent a wide variety of entities from benign fibroadenomas to phyllodes tumors. Rapidly growing or recurrent masses can be particularly concerning to patients, families and physicians alike. Clinical examination and conventional imaging modalities are not efficacious in distinguishing between different tumor types and surgical excision is often recommended for both final diagnosis and for treatment of large or rapidly growing masses. While surgical excision can result in significant long-term deformity of the breast there are some surgical techniques that can be used to limit deformity and/or aid in future reconstruction. Here we present a case of recurrent giant juvenile fibroadenoma with a review of the clinical presentation, diagnostic tools and treatment options.

1. Introduction

Breast masses in children and adolescents are rare entities with a cited prevalence of about 3% [1]. The majority of breast masses are benign with the fibroadenomas being the most common breast tumor in children and adolescents [2,3]. The differential diagnosis of a breast mass in adolescents includes fibroadenoma, giant fibroadenoma, juvenile hypertrophy, phyllodes tumor, benign fibrocystic changes, and malignancy. Breast malignancy in children and adolescents is even more rare, with a reported prevalence of 0.9% [2]. Malignancies of the adolescent breast include metastatic lesions, primary breast carcinoma (lobular or ductal) and lymphoma [2].

Juvenile fibroadenomas are considered giant when they weigh over 500 g, are greater than 5–10 cm, and/or comprise greater than 80% of the breast. These tumors represent approximately 1% of breast masses in adolescents [2] and are characterized by rapid growth and large size. They are histologically characterized by proliferation of both stromal and glandular elements. While treatment is similar, distinguishing these lesions from phyllodes tumors on histology is important, as phyllodes tumors may be malignant resulting in need for a more extensive resection as compared to fibroadenoma. Treatment of giant juvenile fibroadenomas includes surgical excision with preservation of any uninvolved breast tissue [4].

Here we present a case of recurrent giant juvenile fibroadenoma with reactive features on histology treated with resection of the mass and rearrangement of the remaining breast tissue.

2. Case report

An 11-year-old female presented with complaints of a rapidly growing recurrent left breast mass. The patient had first noted swelling of her left breast approximately 1 year prior. She was evaluated at an outside hospital and underwent incisional biopsy of a large breast mass approximately 4 months after initially noting the left breast swelling. Final pathology from those biopsies demonstrated “fibroepithelial lesion with pseudoangiomatous stromal hyperplasia”. She subsequently underwent a “left simple mastectomy” with reported removal of a 20 cm wide lobulated mass. Pathology indicated a mass measuring 19 × 14 × 9.2 cm, weighing 1347 g, with histologic features consistent with giant fibroadenoma with pseudoangiomatous stromal hyperplasia. Margins were noted to be close on final pathology. The patient reportedly noticed a lump in her left breast approximately one month following surgery. An ultrasound obtained approximately 4 months after surgery demonstrated a 3.7 × 5.4 × 3.4 cm mass of the left breast. Ultrasound guided biopsies were subsequently obtained, pathology of which demonstrated gynecomastoid changes with pseudoangiomatous stromal hyperplasia. She was subsequently referred to our clinic.

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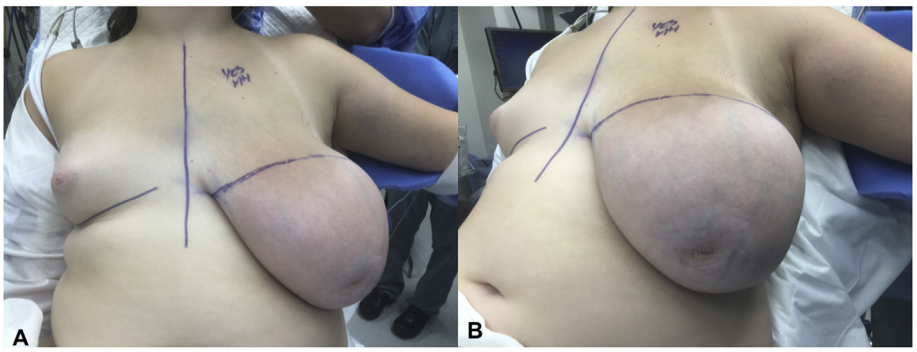

Fig. 1. Preoperative photos demonstrating a large left breast mass with associated dilated superficial veins and nipple retraction. (A) Anterior view (B) Oblique view.

On clinical examination she was noted to have a significantly enlarged left breast as compared to the right with what appeared to be two masses, one superiorly measuring approximately 15 × 10 cm and one inferiorly measuring approximately 8 × 4cm (Fig. 1). She was noted to have nipple retraction and dilated superficial veins. Given the size of the mass and pathology indicating recurrent pseudoangiomatous stromal hyperplasia, surgical excision was recommended. An MRI was obtained for pre-operative planning which was significant for a 15 × 15 × 16 cm heterogeneous mass with pseudocapsule of the left breast (Fig. 2). She was taken to the operating room where she underwent excision of a 23 × 17.5 × 7cm left breast mass (Fig. 3) followed by excision of left breast skin and local tissue rearrangement reconstruction of the left tissue defect (Figs. 4 and 5). Pathology again demonstrated fibroadenoma with myofibroblastic and pseudoangiomatous stroma. She did well post-operatively without complication and no evidence of recurrence was noted at her 1-month follow up visit.

3. Discussion

Fibroadenomas are the most common breast mass in the pediatric/adolescent population. Giant fibroadenomas, however, account for only 1% of breast masses in this population [2]. Giant fibroadenomas, while rare in the pediatric population, present a challenge to the medical community, as there are a lack of clear guidelines regarding the diagnosis and treatment of these lesions.

Giant fibroadenomas clinically present as rapidly enlarging breast mass, either unilateral or bilateral, occasionally associated with overlying skin changes. In the case of giant fibroadenomas, the mass reaches over 5 cm in size on clinical exam. Other causes of significant breast enlargement include juvenile hypertrophy, pseudoangiomatous hyperplasia, and phyllodes tumors. Juvenile hypertrophy is an abnormal response of the breast tissue to estrogen resulting in enlargement without a defined mass. Phyllodes tumors and pseudoangiomatous hyperplasia are rare tumors of the breast and often present as a rapidly growing mass. These tumors are indistinguishable from giant fibroadenomas on physical exam requiring histologic analysis for diagnosis.

Imaging studies are often obtained on these patients with the majority of patient undergoing ultrasonography [4]. Ultrasound characteristics of fibroadenoma are well-circumscribed round or oval masses with uniform hypoechogenicity [5,6]. They also appear as well-defined oval or round masses on mammography and can be associated with calcifications [5,7]. Appearance of fibroadenoma on MRI is variable in regards to T2 hyperintensity though fibroadenomas were noted to have low signal intensity on T1 images and smooth margins [8,9]. Unfortunately, MRI does not precisely differentiate between fibroadenoma and phyllodes tumor [9]. While imaging studies are not diagnostic they are often obtained in an effort to further define the mass and assist with operative planning.

Giant juvenile fibroadenomas are typically treated with surgical excision as their large size can lead to complications of skin ulceration and psychosocial distress. Mastectomy is typically not necessary as these lesions often have a pseudocapsule that separates them from normal breast tissue. Current recommendations are for removal of the mass with preservation of developing breast tissue and the nipple areolar complex (NAC) if possible. Giant fibroadenomas present an additional challenge as enucleation of the mass alone results in significant asymmetry. Reconstruction is often performed at the time of mass excision with the knowledge that additional procedures may be required in the future as the breast develops. Many different reconstructive techniques have been described following excision of large breast masses in the

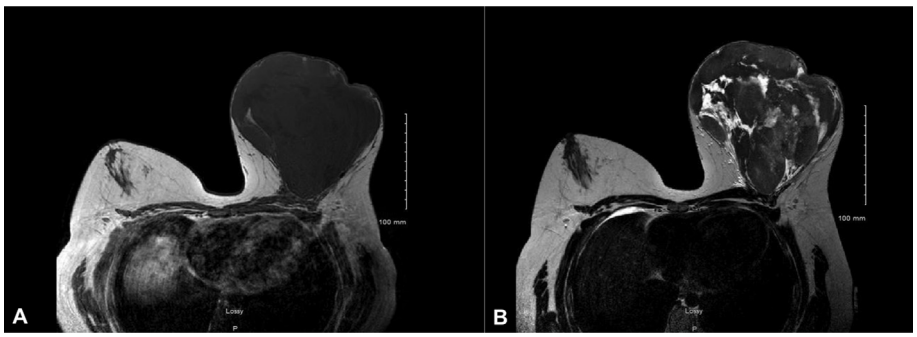

Fig. 2. A preoperative MRI was obtained for operative planning. This demonstrated a 15 × 15 × 16 cm heterogeneous mass of the left breast with a pseudocapsule. (A) T1 weighted image. (B) T2 weighted image.