Case Report

Bilateral Giant Juvenile Fibroadenoma of the Breast Developing Metachronously in a 10-Year-Old Girl

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Abstract

Giant juvenile fibroadenoma (GJF) is very rare and represents about 0.5% of all fibroadenomas. We describe a case of a 10-year-old premenarche girl that presented with huge bilateral GJFs. The right breast mass appeared first and was managed accordingly. The left mass developed rapidly within three months necessitating a second subtotal mastectomy.

Keywords: Breast, giant fibroadenoma, subtotal mastectomy

Introduction

Breast masses are uncommon in children and when they occur, are most often of benign nature such as inflammatory lesions, physiologic hypertrophy, and benign proliferative lesions, most notably fibroadenomas.[1,2] Breast fibroadenomas occurring in childhood and adolescent age groups of up to 19 years are named juvenile fibroadenomas (JFs),[1,3] JFs are dubbed "Giant" if they grow to a size >5 cm in dimension or weight of more than 500 g or replace at least 80% of the breast.[4] About 4% of all fibroadenomas are juvenile while 0.5% of all fibroadenomas are giant JF (GJFs).[5] Thus, both conditions are rare clinical presentations more so in prepubertal girls.

Fibroadenomas are biologically benign with very little or no risk for malignant transformation.[3] Conservative approach in management is sometimes adopted, as 10%–40% of cases are known to resolve spontaneously.[4] However, JFs are known for rapid growth and enlargement to big masses within a short interval of time. Surgical excision is most often needed in the treatment of GJFs as large, pendulous breast masses in an adolescent girl are often a source of physical discomfort and emotional burden.[2,3,6]

In terms of laterality, most JFs tend to be unilateral in location. Eleftheriades *et al*. in a recent extensive review of PubMed literature found only 87 published cases of JFs out of which 82.2% (72/87) were unilateral.[3] In their review, only 15 out of 87 (17.2%) published cases of JFs occurred bilaterally. Almost all bilateral JFs in literature are presented synchronously. Metachronous development of bilateral JF is rarely reported.

We present a rare case of metachronous bilateral GJFs in a 10-year-old prepubertal girl requiring bilateral subtotal mastectomy. This to the best of our knowledge is the first case of JF to be reported from Sierra Leone.

Case Report

A 10-year-old premenarche girl presented with a rapidly enlarging right breast mass of six-month duration associated with dull, nonradiating ache, which was gradual in onset and exacerbated by physical activity. The swelling was associated with low-grade, intermittent fever, and easy fatigability. History of trauma, nipple discharge, anorexia, or weight loss was absent. There was no family history of such an occurrence. On examination, there was a slightly tender, huge, well-circumscribed mass in the right breast, which was firm in consistency. The overlying skin of the breasts was tense and shiny with prominent superficial veins. The right

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breast mass measured 10 cm × 8 cm and was not fixed to underlying structures [Figure 1a]. There was no associated regional lymphadenopathy. The patient had fine-needle aspiration cytology (FNAC) which revealed a benign (C2) lesion consistent with fibroadenoma. Subsequently, the patient and mother were counseled and she had a subtotal mastectomy with excised tissue sent for histopathological assessment.

Two weeks after the surgical resection of the right breast, enlargement of the left breast became noticeable and grew rapidly to surpass the previously resected right breast mass in three months. The mass measured about 13 cm × 12 cm and showed similar clinical features as the resected right mass [Figure 1b]. The left breast mass was also excised by subtotal mastectomy following the clinical workup.

Histopathological assessment of resected right and left breast specimens showed circumscribed masses weighing 2.5 kg and 3 kg, respectively, with solid and grayish-white cut surfaces. Histology of both masses showed a biphasic proliferation of stromal and epithelial elements with ductal structures disposed in an expansive sheet of moderately cellular stroma [Figure 2]. The glands are compressed into slit-like spaces in areas. There was no evidence of malignancy. Diagnoses of GJF were made. Postoperative follow-up of the patient for over 18 months has been generally uneventful. The patient consent was sought and obtained for this report.

Discussion

JF of the breast is defined as circumscribed, often large, breast mass occurring in adolescent females, with stromal and epithelial hypercellularity but lacking the leaf-like growth pattern of phyllodes tumours.[7] The diagnostic criteria for JF are circumscribed breast mass which is rarely multiple; biphasic stromal and epithelial process, in which pericanalicular pattern is common in the uniformly hypercellular stroma. Other features infrequently seen include cytologic atypia and mitotic rate >3/hpf, with a mean age of 15 years.[7]

Giant fibroadenoma on the other hand is defined as a tumour >500 g or disproportionately large compared to the rest of the breast.[7] In our case, the right breast tumour weighed 2.5 kg while the left mass weighed 3 kg. GJF is more common in young and African patients.[7] In agreement with this profile and with reviewed literature, our patient is 10 years old and black. Bilateral occurrence of GJF is not a common presentation. It is seen in only about 17% of cases.[3] Bilateral GJF occurring metachronously as in this case is much rarer. The etiology of GJF is still unknown, however, plausible theories in explanation of its development have included hormonal factors, most notably estrogenic influences.[6,8]

A number of breast conditions present in a similar fashion as JFs and are thus important differentials, which need to be distinguished from JF because treatment approaches may differ depending on the condition. These conditions include benign phyllodes tumours, virginal hypertrophy, and other rare differentials such as lipoma, hamartoma, breast abscess, macrocyst, adenocarcinoma, and pseudoangiomatous stromal hyperplasia. Treatment modalities and prognoses differ in these conditions. Some will require only local excision, aspiration, or conservative management while carcinoma may need wide local excision or even mastectomy.[5] FNAC and biopsy can be employed in the diagnosis of these conditions.[2] FNAC was done in our case and found to be fibroadenoma. Being a benign process, the optimal and most common treatment for JFs is surgical excision. However, cases managed with mastectomy and breast reconstruction are amply documented in the literature.[3,4,6,9,10] Mastectomy has been employed when GJF replaces the entire breast and is extensively ulcerative or multifocally florid.[4,10] Our case was quite extensive with total replacement of as yet undeveloped breast tissue, thus subtotal mastectomy was carried out to achieve complete excision. Subtotal mastectomy (breast-conserving surgery) was well received by the patient and accepted by the parents as it gave relief for a huge discomforting and distressing mass. Delayed reconstruction is planned when the patient might have achieved adulthood.

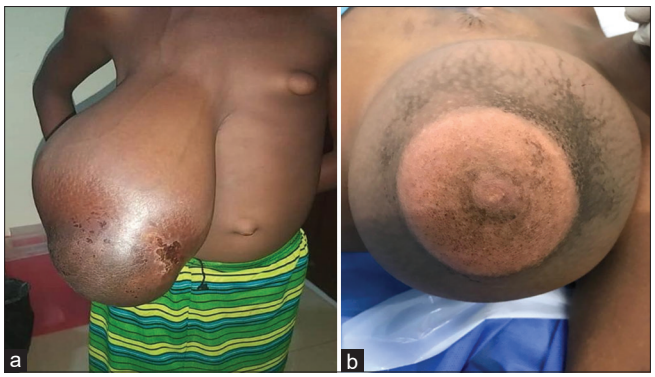

Figure 1: (a) Photograph of right breast mass showing tense skin, engorged vessels, and skin ulcerations. Mass grew to the size of 2.5 kg in six months. The left breast was normal. (b) Photograph of left breast mass. The left breast mass was noticed two weeks after excision of the right mass. It grew rapidly to the size of 3 kg in three months

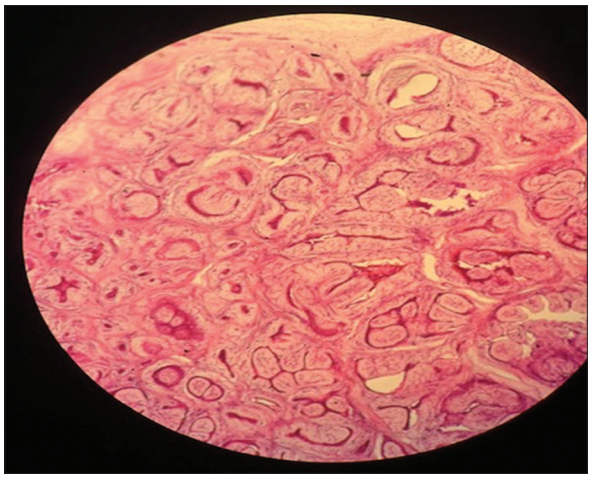

Figure 2: Photomicrograph histologic section of breast tissue showing classic features of fibroadenoma

It is worthy to note that GJFs may recur after complete excision and the chance of recurrence becomes less after the third decade.[11] The possibility of bilateral GJFs being a component of Carneys complex-comprising multiple myxoid fibroadenomas, endocrine hyperactivity, cardiac myxoma, cutaneous hyperpigmentation, and other abnormalities-should always be explored.[12] In our case, detailed examination revealed only fibroadenomas, other components of the Carneys complex being absent.

In conclusion, we report an extremely rare case of metachronous bilateral giant fibroadenomas of breasts in a 10-year-old premenarcheal girl. Large tumours effacing normal breast tissue or tumours complicated by extensive ulceration are treated with subtotal mastectomy to ensure complete resection. Adequate follow-up and breast reconstruction are needed to ameliorate the psychological effect of treatment.

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Conflicts of interest

There are no conflicts of interest.

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