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CASE REPORT
Breast

A Rare Case of Persistent Unilateral Gestational Gigantomastia

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Summary: A 34-year-old para 2+0 Indonesian woman presented with persistent right-sided gestational gigantomastia some 24 months following delivery. This was severely debilitating her activities of daily living, including caring for her children. On examination, she was found to have extreme hypertrophy of her right breast, which was nodular throughout on palpation. Biochemical investigations were unremarkable and revealed no obvious etiology. Magnetic resonance imaging identified grossly enlarged right breast tissue with prominent vessels. Given the minimal involution of her breast over the 24 months postpartum, she elected for a breast reduction with free nipple grafting following appropriate counseling. This was performed through excision of breast parenchyma preserving superior-medial tissue, followed by a free nipple graft. *(Plast Reconstr Surg Glob Open 2019;7:e2372; doi: 10.1097/GOX.0000000000002372; Published online 19 August 2019.)*

This is a rare case of persistent unilateral gestational gigantomastia. A 34-year-old Indonesian woman presented with persistent right-sided gestational gigantomastia almost 24 months postpartum. She had initially presented while 32 weeks pregnant with her second child. On consultation, she reported a significant increase in size of her right breast during her first pregnancy 4 years before initial presentation.

Although there had been some involution of her right breast postpartum, this was further exacerbated during her second pregnancy, during which time she initially presented. She was unable to lactate from her right breast. The decision was made to adopt a watchful wait approach to allow for potential involution postpartum; however, there was minimal change.

On examination, her left breast was a B cup. Her right breast was significantly enlarged, with ptosis of tissue to her iliac crest. Her nipple–areolar complex was significantly enlarged due to stretching of the skin and was displaced laterally and inferiorly. The skin over the upper pole of the right breast and thorax had multiple prominent dilated veins. To compensate for the increase in weight, there was evidence of muscle hypertrophy around the shoulder, neck, and back. There was no evidence of ulceration or infection. On palpation, the left breast was unremarkable. The right breast was dense and nodular throughout. There were no sinister features, or evidence of axillary or supraclavicular lymphadenopathy.

INVESTIGATIONS

Laboratory investigations were unremarkable. Hormonal screening and autoimmune screening were not performed.

Ultrasound scan of both breasts was performed during her initial presentation. This was technically challenging; however, no evidence of an underlying mass was noted.

MRI was subsequently performed on follow-up at 24 months postpartum. This identified an encapsulated heterogeneous right breast mass measuring 16.9 cm × 14.7 cm × 20.6 cm, consisting of multiple well-defined lobules, with a mass effect displacing normal breast parenchyma laterally. The fat plane between the mass and pectoralis major was noted to be preserved. Prominent vessels were noted in the periphery of the mass, arising from the right internal thoracic artery.

DIFFERENTIAL DIAGNOSIS

In the setting of rapid and extreme breast hypertrophy during pregnancy, the most likely cause is gestational gigantomastia. However, it is important to exclude other underlying causes such as breast cancer, inflammatory breast cancer, fibroadenoma, Paget's, Phyllodes tumor, and lymphoma of the breast.[1]

TREATMENT

Following a 24-month period of watchful waiting postpartum to allow for potential breast tissue involution,

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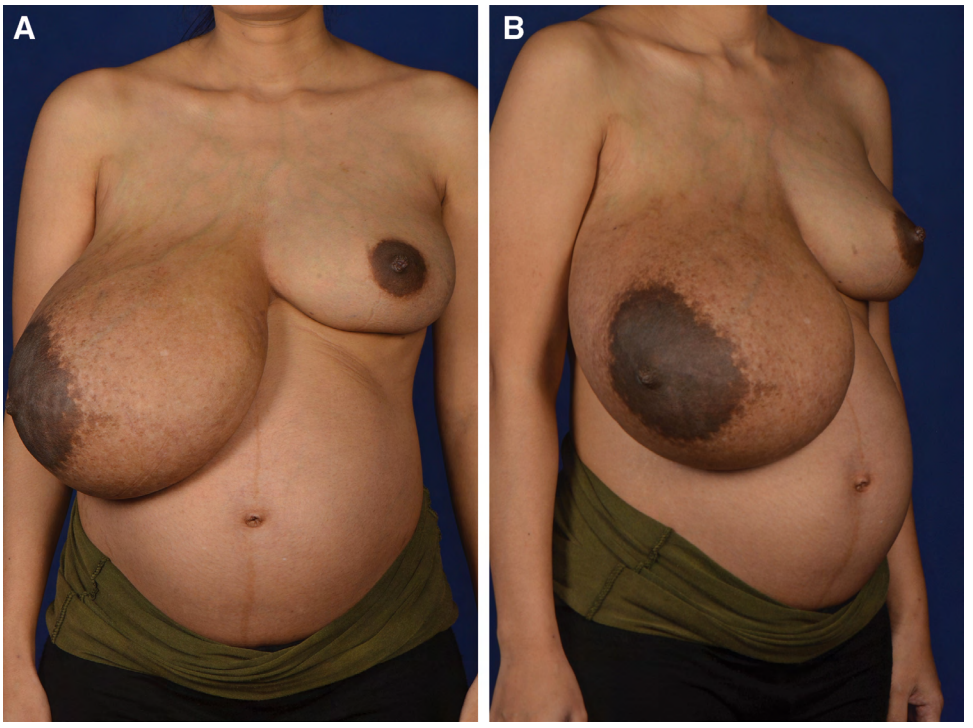

Fig. 1. A, Frontal and B, right lateral view at 24 months post partum after delivery of her second child. Note the minimal involution of the right hypertrophied breast.

there was found to be minimal change (Fig. 1). She had completed her family. The patient therefore elected to undergo a breast reduction and free nipple grafting. A Wise-pattern approach was performed through parenchymal excision preserving superior-medial tissue, followed by free nipple grafting. The total breast tissue excised was 2.25 kg. Histopathology showed that benign breast tissue with increased collagenization and fibrosis of stroma. There was no evidence of atypia or malignancy.

OUTCOME AND FOLLOW-UP

Following surgery, the patient reported instant relief of her back and shoulder ache. Initial follow-up at 1 and 6 weeks were satisfactory, with good wound healing, intact nipple graft, and no evidence of complications. At 1-year postoperatively, there was good symmetry of the shape and size. The nipple areolar complex was at the same level on the breast mound and the patient scarred well (Fig. 2). She has no further lower back pain and was able to take care of her children with ease. The patient was extremely pleased with the outcome.

DISCUSSION

Definition and Background

Gigantomastia, also known as macromastia, is a rare condition characterized by excessive hypertrophy of the connective tissues of the breast. Although there is no universally accepted definition of gigantomastia, it generally refers to extremely rapid breast hypertrophy of >1.5 kg. Gigantomastia is almost universally bilateral in occurrence, however may rarely be unilateral.[2–5]

Gestational gigantomastia is a rare, benign condition of unknown etiology characterized by diffuse, extreme hypertrophy of 1 or both breasts during pregnancy. It is estimated that 1 in every 28,000–100,000 pregnancies are affected to some extent by gestational gigantomastia. There have been approximately 150 cases of gestational gigantomastia reported, because it was first described in 1648.[6] However, there have only been 4 other cases of unilateral gestational gigantomastia previously reported.[2–5] Hypertrophy usually commences in the first trimester, or early in the second, and progresses until delivery. Although, in some cases, breasts recede to their postpartum state, with subsequent pregnancies, they enlarge again, usually more extensively.[7] Although this condition can be associated with considerable morbidity for the mother, it is generally associated with good fetal outcome.[8]

Etiology

The etiology of gigantomastia remains uncertain, and many possible theories have been postulated. Although benign in nature, it may cause severe morbidity and even mortality. The most common associations appear to be obesity, puberty, pregnancy, multiparous women, and Caucasian ethnicity.[8] The pathogenesis appears to be linked to excessive production of estrogen and prolactin; however, several cases of gigantomastia have been reported in the setting of normal hormone levels; therefore, another hypothesis is increased hormone receptor sensitivity.

It has also been reported in the setting of autoimmune disease such as systemic lupus erythematosus, myasthenia

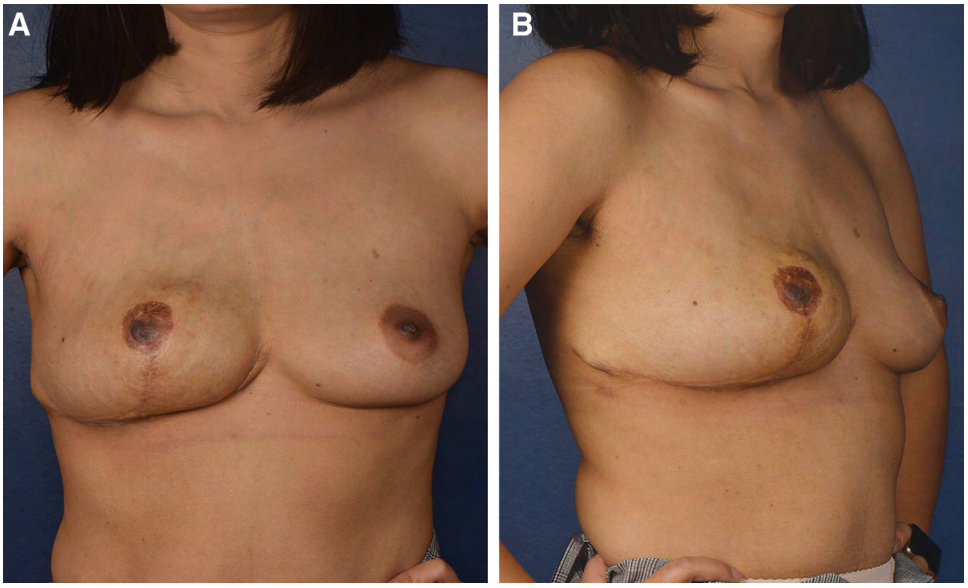

Fig. 2. One year postoperative follow-up. A, Frontal and B, lateral view with good size and symmetry match.

gravis, Grave's disease, chronic arthritis, psoriasis, Hashimoto's thyroiditis, antiphospholipid syndrome, and undifferentiated connective tissue disease.[9,10] It has also been reported in association with hypercalcemia due to pseudohyperparathyroidism.[7]

Gigantomastia has been a reported side effect with particular medication, including the oral contraceptive pill, steroids, and D-penicillamine,[11] with subsequent treatment with danazol.[12,13]

Clinical Features

Features of gigantomastia include rapid growth of the breasts resulting in muscular pain, mastalgia, and incapacity. Enlargement and displacement of the nipple–areolar complex is common, resulting in potential loss of nipple sensation due to chronic traction on the fourth, fifth, or sixth intercostal nerves.[10] Overstretching of the breast skin envelope in extreme cases may result in ulceration, skin necrosis, and subsequent infection and hemorrhage. In the setting of pregnancy, lactation may be inhibited due to pinching of the ducts, resulting in mastitis. This may also have implications on breastfeeding, which is of great importance nutritionally and immunologically in the neonate.

On palpation, the breasts are generally firm, with varying degrees of nodularity. It is also important to consider the emotional and social impact that this condition poses.

Diagnosis and Imaging

Laboratory investigations include full hematological and biochemical profiling, including calcium, thyroid function, and parathyroid function. Hormonal investigations include prolactin, progesterone, and estrogen levels.

Serological autoantibody screening includes antinuclear antibodies (ANA).

Imaging with ultrasound or mammogram may be challenging, or technically not possible; therefore, MRI may be the best modality. Regardless, it is of the utmost importance to have a baseline of breast appearance before undergoing any treatment and to ensure that there is no underlying malignant process. If there is any uncertainty, a core biopsy should be performed under image guidance, taking particular care due to vessel engorgement.

Management

Management of gigantomastia may be divided into conservative and surgical approaches. Following cessation or management of the presumed causative stimulating factor, such as medication and pregnancy, a watch-and-wait approach may be adopted, due to potential involution of the breast tissue. Invariably, however, once significant hypertrophy has occurred, it does not appear to regress.[14] Binders can be used to support the breasts. Infection and ulceration can be managed with antibiotics and dressings. Medical management includes treatment with danazol in the setting of D-penicillamine. Dopamine 2 receptor agonists such as bromocriptine, or the less toxic cabergoline, may be used in the setting of hyperprolactinemia to halt progression and may cause regression.[1] If taken during pregnancy, fetal growth should be closely monitored due to the potential risk of intrauterine growth retardation.[10] Other medications, which have been tried without success, include diuretics and androgens.

Surgical management tends to be the mainstay of treatment. Ideally this should be postponed until postpartum or until a viable fetus can be delivered. Options generally include reduction mammoplasty, or mastectomy, with or without reconstruction. Reduction has the advantage of allowing for postoperative breastfeeding; however, it is extremely important that patients are adequately informed as to the risks of recurrent gigantomastia of residual breast

tissue, particularly in the setting of further pregnancies. Free nipple grafting may be required in massive gigantomastia but is associated with reduced nipple projection and sensation and destroys lactation potential; therefore, mothers must be carefully counseled.[7]

Mastectomy is usually reserved for those who have recurrent gigantomastia following reduction. Mastectomy may also be necessary in the setting of life-threatening complications of gigantomastia, such as infection and hemorrhage.

CONCLUSIONS

Persistent unilateral gigantomastia is extremely rare. Although benign, with no agreed etiology, it has the potential to cause significant maternal morbidity, and even mortality. Although breast parenchyma may involute following delivery, the mainstay of treatment remains surgical management with breast reduction, or mastectomy in the setting of recurrence or life-threatening complications.

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