

ISSN 2374-216X

Gigantomastia: A case report with review of literature

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Abstract

Gigantomastia is a rare disabling condition characterized by excessive breast enlargement. We report a case of gigantomastia in a 20-year old woman. The patient presented with complain of progressive bilateral breasts enlargement starting during pregnancy which continued even after spontaneous abortion in early third trimester. The patient was known to have lupus nephritis which may be a precipitating factor as autoimmune disorders have been implicated as a cause. The patient underwent bilateral mastectomy but failed to survive secondary to lupus induced severe renal failure.

Keywords

gigantomastia; massive hypertrophy of breast; virginal hyperplasia; gestational macromastia

Introduction

Gigantomastia is a rare condition characterized by massive enlargement of breasts with per breast weights of 4-6 kilograms, leading to severe morbidity and sometimes even mortality. It can either occur during adolescence or puberty called virginal gigantomastia or during pregnancy known as gravidic gigantomastia [1]. Few drug induced cases have also been described in literature. Gestational gigantomastia complicates about one of every 28,000-100,000 pregnancies [2] and less than 100 cases have been described in literature [1]. Medical treatment is based on bromocriptine administration, but in most cases surgical intervention in the form of reduction mammoplasty or simple mastectomy with posterior reconstruction is required if the disorder progresses [3,4].

We report a case of gigantomastia in a 20-year old woman. This is the first case of gigantomastia that we encountered in our practice. The severity of the problem was psychologically and physically disablitating leading to dyspnoea and significant difficulty in standing or walking.

Case Presentation

A 20-year-old married woman, weighing 60 kg, presented with complains of progressive enlargement of bilateral breasts after the onset of pregnancy along with pain, discomfort and difficulty breathing as well as standing up or walking (figure 1). She was known to have lupus nephritis leading to renal failure and was dialysis dependent. This enlargement started at 24 weeks of gestation and continued to increase rapidly as the pregnancy progressed. She had spontaneous abortion at 28 weeks gestation due to intrauterine fetal death of unknown etiology. Her breasts however continued to increase in size. The patient underwent laboratory analysis which was inconclusive.

Owing to her critical condition the gynecological team recommended radiological evaluation. Contrast enhanced CT scan with axial images was obtained from lung apices up to adrenal glands acquiring lung and mediastinal window. The examination was technically compromised because of massively enlarged breasts which could not be placed into the field of imaging. Bilateral breasts were larger than CT scan machine gantry resulting in streak artifacts obscuring fine anatomical details. As the breast enlargement had occurred during pregnancy thus termed gestational macromastia or gigantomastia. The CT scan report mentioned mild pulmonary infiltrates bilaterally and mild pleural effusions, unfortunately the films were not available for review.

The patient consulted a breast surgeon who advised bilateral breasts ultrasound to rule out any occult lesion. Both breasts were examined using high as well as low frequency probe, which showed significant stromal proliferation with variable size cysts. There were multiple bizarre shaped anechoic areas with subtle mobile low level internal echoes as well as incomplete septations suggestive of gross ductal dilatation (figure 2a). Few cystic spaces showed low velocity venous flow suggesting them to be venous spaces (figure 2b). No solid mass was seen on either side. Bilateral axillae showed benign appearing lymph nodes. The patient underwent bilateral reduction mastectomy with nipple areolar graft because of gross breast hypertrophy. The excised right breast tissue weighed 4600 grams while the excised left breast tissue weighed 4100 grams. Bilateral resected specimens revealed breast parenchyma exhibiting fibrosis, adenosis and foci of intraductal microcalcification. The skin was unremarkable. No focus of in-situ or invasive malignancy was seen.

Patient recovered well after the surgery and was discharged home in stable condition, however, she did not survive due to severe renal failure.

Discussion

Gigantomastia is a rare condition characterized by excessive breast enlargement, first reported by Palmuth in 1648 [1]. The rarity of this disorder has led to difficulties in determining its true incidence. It may occur spontaneously, during puberty or pregnancy or can be drug induced [5].

Gigantomastia can present as mastalgia, ulceration, infection, postural problems and back pain along with difficulty in breathing and movement as well as social and psychological problems [3]. It may also be associated with fetal growth retardation, if this condition presents during pregnancy [5].

There is controversy regarding the exact definition for gigantomastia, however gigantomastia is considered if there is breast enlargement requiring reduction of over 0.8 - 2 kg of breast tissue [3].

The condition is seen exclusively in females and there is no reported case of gigantomastia in males, best to our knowledge.

The exact etiology of gigantomastia is not known, however most studies speculate either hormonal oversensitivity or increased production [4,5,6]. This usually results in excessive breast growth at puberty or during pregnancy. Adverse effect of certain medications like D-pencillamine have also been proposed as a likely cause in few cases [5]. There is still another group of patients in whom no precipitating cause or drug is found and these constitute the idiopathic group. The patients in this group may or may not be overweight and the presentation is usually in third to fifth decade [5]. There are also case reports in which autoimmune diseases such as SLE and rheumatoid arthritis have been reported in

association with gigantomastia [7]. Dancey et al. found three idiopathic cases with concomitant immunological diseases in their literature review, out of which one was subsequently found to have some features of SLE indicating an autoimmune etiology [5].

Our patient fell in the same age-range as described as in most reports and onset was in the second trimester. She was known to have lupus nephritis and this may be the precipitating factor for breast hypertrophy during pregnancy.

Risk factors for gestational gigantomastia are not fully understood. Caucasian, multiparous women are more likely to be affected although it may occur in primiparous women as well [7]. Prior history of gestational gigantomastia also increases the risk [3,7] in subsequent pregnancies.

The breast enlargement can occur any time in pregnancy however Shoma et al. indicated it to be more common in first trimester [8].

Unfortunately, majority of pregnant women in developing countries do not have access to antenatal care by qualified obstetricians. Late presentation is indeed a common phenomenon in underdeveloped countries. The problem is multi-factorial in nature, ranging from lack of awareness, religious beliefs and an ever-present socioeconomic problem.

The disease is usually progressive and spontaneous regression rarely occurs even if the enlargement ceases. There are few reports of gestational gigantomastia resolving spontaneously postpartum, however majority require surgical treatment [7].

Although gestational gigantomastia is a benign entity, imaging workup should be done to rule out mastitis or underlying tumour such fibroadenoma, phyllodes, primary breast neoplasm or lymphoma [7].

The management of progressive gestational gigantomastia requires a multidisciplinary approach regarding the decision for surgery and the timing of surgery. Involved disciplines include plastic surgery, obstetrics, anaesthesia and neonatology. There are also several reports of attempts to treat gigantomastia conservatively with various hormones [2,9] as majority believe in hormonal etiology. Bromocriptine is recommended as the initial drug of choice in gestational gigantomastia with addition of progesterone [10,11] although there is no definite evidence of increased prolactin levels [1]. Bromocriptine has been shown to arrest as well as decrease breast growth [5] and can be continued until the baby delivers following which surgery can be planned. Tamoxifen and medroxyprogesterone are more suitable for juvenile/virginal gigantomastia [5].

Surgical treatment is recommended for cases non responsive to medical treatment or when there is haemorrhage, necrosis, ulceration and sepsis as these may be fatal [7]. Surgical options include reduction mammoplasty, subcutaneous mastectomy or simple mastectomy, with or without reconstruction [12]. Simple mastectomy is preferred over reduction mammoplasty as there is less blood loss and subsequent pregnancies are expected to be uneventful [1].

Figures



Figure 1: Massively enlarged breasts with scarring in the peri-areolar region due to previous ulcerations. (A, B) patient in lying position, (C) patient is sitting.

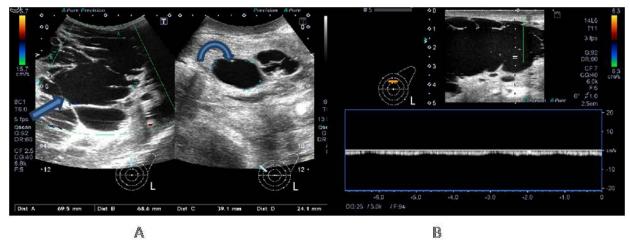


Figure 2: (A) Bizzare shaped anechoic spaces with sepatations (straight arrow) representing dilated ducts and cysts (curved arrow). (B) One of the cystic space shows low velocity continuous flow indicating venous space.

Conclusion

Gestational gigantomastia is an exceptionally rare disorder with remarkable changes in the mammary glands in response to hormonal stimulation resulting in physical and emotional distress. Massive enlargement of the breasts leads to infection, ulceration and hemorrhage which are potentially lethal, both for the patient and fetus. The management requires a multidisciplinary approach regarding the decision for surgery and its timing.

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