

Bilateral Gestational Gigantomastia Complicated by Severe Sepsis; Case Report of a Preventable Mortality

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ABSTRACT

Gestational gigantomastia is characterized by a rapid and disproportionate growth of the breasts with a homogeneous increase in breast volume. Enlargement of the breasts with infection, ulceration and sepsis are potentially fatal for the patient and her fetus. The management is challenging, however the need for appropriate surgical intervention is rarely in question. A multidisciplinary approach regarding the decision for surgery and the timing of surgery is crucial to outcome. We report a case of gestational gigantomastia complicated by severe sepsis in a low resource setting to highlight the peculiar challenges of management.

Key words: Consent, gigantomastia, gestational, late presentation, mastectomy, severe sepsis

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INTRODUCTION

Breast enlargement in pregnancy is a physiological response to hormonal stimulation in preparation for lactation after parturition.^[1,2] Thus, in the normal gravid state, breast volume may double without adverse sequelae.^[3] This natural phenomenon, in rare instances, becomes grossly exaggerated resulting in the development of crippling gigantomastia.^[2] It is characterized by a rapid and disproportionate growth of the breasts with a homogeneous increase in breast volume.^[3-8] Breast tissue may enlarge as much as 10-20 times.^[3] The rapid increase in breast size can lead to pain, skin ulceration, infection, bleeding and necrosis as well as difficulty with ambulation and performance of activities of daily living.^[3] In addition to the physical and emotional stress, gestational gigantomastia may cause maternal and fetal death. The management of gigantomastia in pregnancy is challenging and the need for appropriate surgical intervention is rarely in question.^[6,9] However, surgery in Sub-Saharan Africa is widely known to be done against a background of late presentation with complicated pathologies. Furthermore, endemic cultural practices poses significant challenges as patients try alternative treatments that may be more harmful. A delay in referral and diagnosis secondary to these intrinsic difficulties

continues to contribute unfortunately, to the increased morbidity and mortality in surgical care.^[1] We report a case of bilateral gestational gigantomastia complicated by severe sepsis to highlight the peculiar management challenges in a low resource setting.

CASE REPORT

A 27-year-old woman, gravida 2 para 1 + 0 presented at 20 weeks gestation with bilateral gestational gigantomastia and extensive necrosis of the lower pole of the breasts. The patient noticed bilateral enlargement of the breasts during the first trimester, which rapidly increased in size as the pregnancy progressed. She had previously sought unorthodox medical treatment for the progressively enlarging breasts. These included the use of traditional remedies in the form of oral herbal medications and application of herbal pastes to the breast. She noted some degree of enlargement of her breasts during her first pregnancy which started at 30 weeks gestation and was well-tolerated until

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delivery. After delivery, her breasts returned to normal size. Menarche was at the age of 14 years and she had no family history of breast disease.

On examination, she was anxious, pale and dehydrated. The breasts were markedly enlarged and exquisitely tender with massive ulcerations and sepsis of the distal third of the breasts [Figure 1] The overlying skin showed marked hyperpigmentation with prominent dilated superficial veins. The skin was edematous with a *peau d'orange* appearance. There were multiple, tender, discrete, axillary lymphadenopathy. She was severely limited by the intense pain and size of her breasts. She needed help mobilizing, feeding and bathing. The only position she could sleep was supine in order to rest her breasts beside her. The examinations by the obstetrician and ultrasound of the pelvis at admission revealed a viable, progressing normal fetus. The patient had a complete laboratory work-up, including a complete blood count, chemistry and hormonal assay. Her hemoglobin was 8 g/dL, platelet count of 380 g/L and WBC was 17.2×10^9 L. A review of the peripheral blood smear revealed Neutrophilia with toxic granulation. Blood chemistry was within normal range. A swab for culture and sensitivity yielded a growth of proteus species. Hormone assays showed elevated prolactin and luteinizing hormone. She was scheduled to have an ultrasonography of the breast, but this was not done because of unavailability of a mobile unit. Tissue biopsies were performed bilaterally. Histologically, the biopsies were consistent with chronic non-specific inflammation of the breasts. Blood culture subsequently returned positive for *Staphylococcus aureus*. The patient was commenced on parenteral antibiotics and daily dressings of the ulcers with honey.

At a multidisciplinary team consultation, involving the Breast and Endocrine Surgeon, Plastic and Reconstructive Surgeon, Obstetricians, Pediatricians and Anesthesiologist, it was decided that the patient



Figure 1: A 27-year-old woman at 22 week gestation with massive breast hypertrophy and extensive septic ulceration

should have a simple mastectomy immediately. This was declined by the patient. The patient and family had problems with accepting bilateral simple mastectomy as a treatment option for her benign breast lesion. She gave birth to a fresh still birth at 22 weeks gestation. Her general condition deteriorated within 48 hours of delivery with severe sepsis. She was dyspneic with orthostatic pneumonia. She was resuscitated with crystalloids, blood transfusion and given intravenous antibiotics. The patient succumbed to overwhelming sepsis despite rigorous and intensive medical therapy. An autopsy was not performed. (The family declined the request for an autopsy)

DISCUSSION

The first case of gestational gigantomastia was reported by Palmuth in 1648 and since then less than 150 cases have been reported in the literature.^[6,10] The rarity of the disease has created difficulties in assessing the incidence, with estimates ranging between 1 per 28 000 pregnancies and 1 per 100 000 pregnancies.^[11] Risk factors for gestational gigantomastia are not fully appreciated. However, it most commonly occurs in multiparous women (although primiparous women may be affected). Caucasian women are more likely to be affected than blacks (9:4).^[12] Fetal gender and maternal age do not appear to be significant risks, paternal risk factors are yet to be identified.^[4] Despite the ambiguity in etiology, it is generally agreed that there is an abnormal end-organ response to the hormones of pregnancy.^[13] Breast enlargement is typically observed during the first trimester and occurs in 64% of women who develop gestational macromastia.^[3,8] Gigantomastia need not occur in the first pregnancy. However, having occurred in one pregnancy, gigantomastia complicates almost all subsequent pregnancies. This condition, whatever its cause, is a progressive disease.^[10,14] Patients with gestational gigantomastia will at first consult their general practitioner or obstetrician. It is important that they consult early with other disciplines to determine a treatment plan.^[5]

Our patient fell into the same age-range as described as in most reports and the onset was in the first trimester.^[4-6,15] She also had a history of breast enlargement in the first pregnancy. Antenatal care was at a peripheral clinic but presented to the hospital with breast ulcerations. The majority of pregnant women in developing countries including Nigeria, do not have access to antenatal care by qualified obstetricians.^[16] This is further aggravated by a health care delivery system that is not based on a standardized evidence-based protocol. Rather, the provision of care is based on the knowledge, competence and decisions of individual practitioners, which are obviously highly variable. Periodic updating and the regular

monitoring of adherence to health care delivery protocols are important tools that will facilitate prompt and appropriate referrals.^[16] Late presentation in the hospital is indeed a common phenomenon in developing countries. This is well demonstrated in this study. The problem is multi-factorial in nature, ranging from lack of awareness, religious belief and an ever-present socioeconomic problem. There are readily available and accessible herbal and spiritual treatment options and ill-informed perceptions about breast diseases in pregnancy. There is also the fear of mastectomy.^[17]

The management of gestational gigantomastia requires a multidisciplinary approach regarding the decision for surgery and the timing of surgery. Involved disciplines include plastic surgery, obstetrics, anesthesia and neonatology.^[5] Surgical treatment becomes mandatory once complications occur. Enormous enlargement of the breasts with infection, ulceration and hemorrhage are potentially fatal for the patient and her fetus and are absolute indications.^[10,13] The treatment of choice is a simple mastectomy with delayed reconstruction. Intra-operative and peri-operative management issues must be considered during mastectomy. In patients with gestational gigantomastia, intraoperative blood loss is substantially greater because of the severely engorged and friable vessels.^[4,5] Consideration has to be given to the large blood loss during surgery.^[12] Thus, in performing bilateral mastectomy, speed and accuracy are important factors in reducing blood loss. Reducing blood loss and the duration of the operation, diminishes the risk of spontaneous abortion as well as exposing the mother and fetus to less of the potentially teratogenic anesthetic agents.^[4,5] In the absence of complications, bilateral mastectomy with delayed reconstruction and breast reduction are the most common surgical procedures performed. The decision to perform one versus the other depends on multiple factors. Breast reduction reduces the volume of breast tissue but does not eliminate the risk of recurrence with subsequent pregnancies. Patients who underwent breast reduction either before pregnancy, during pregnancy, or after delivery and again became pregnant had a 100% (four of four) chance of recurrence. In addition, breast reduction does not eliminate the risk of further hypertrophy during the existing pregnancy.^[3,4] Therefore, if the patient is planning on future pregnancies and wishes to reduce the risk of recurrence, then bilateral mastectomy with delayed reconstruction is her best option. If she is not planning on future pregnancies, is clinically stable, and understands the associated risk of increased operative blood loss and continued hypertrophy during the existing pregnancy, then breast reduction is a viable option.^[4]

Our case exhibited a few unusual features. She demonstrated most of the complications of gigantomastia; ulceration, sepsis and spontaneous

termination of pregnancy. Swelstad *et al.*,^[4] in their review of the natural history of gestational gigantomastia, reported the occurrence of spontaneous termination of pregnancy in seven (10%) out of 69 women. In pregnancies carried to at least 8 months, two infants died in the peripartum period. Two women died while pregnant. One of these women died after her breasts turned black in the fourth month of her pregnancy. Hence, bilateral simple mastectomy was the treatment of choice as a life-saving procedure for this patient. The peculiar feature here is that the patient and her family declined to give consent for simple mastectomy. The moral obligation for Nigerian surgeons towards their patients is to be able to secure first-person voluntary informed consent, while at the same time remaining conscious of the attendant cultural implications.^[18] This is because a woman concedes part of (if not all) her autonomy to her husband and husband's family members on the payment of the bride-price on her 'head'. This makes it difficult, especially in emergency cases, to obtain consent for surgery of a married woman, as the surgeon has to wait for the husband or senior male members of the family to be present before surgery can be undertaken.^[18] The dictum that a pregnant woman should never be penalized because of her pregnancy has universal applicability. Necessary treatment should not be restricted or unduly delayed because of the pregnancy.^[10]

Just as public-health interventions and educational projects have greatly improved outcome in communicable diseases in sub-Saharan Africa, so might analogous efforts in patient education and advocacy in surgery, improve surgical outcome and quality of care. All patients who are identified as high risks for surgical intervention should have processes of care that will increase the probabilities of a good outcome. These should commence at the initial pre-operative assessment to the final post-operative discharge clinic and should be communicated clearly to the patient and his family.^[19] Patients need to know the specific implications for care so they can make informed decisions based on the assessment of the surgeon.

CONCLUSION

Gestational gigantomastia is a rare disorder with remarkable changes in the mammary glands in response to hormonal stimulation. It is associated with physical and emotional distress, and when complicated by sepsis may cause maternal and fetal death. The management requires a multidisciplinary approach regarding the decision for surgery and the timing of surgery. There is a need for an awareness of the attendant cultural implications, especially as regards voluntary informed consent. This will improve both quality of care and surgical outcome.

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