

A RECURRENT LACTATING VULVAR BREAST AND GIGANTOMASTIA DEVELOPING DURING PREGNANCY: A CASE REPORT

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ABSTRACT

Gigantomastia during pregnancy is a rare disorder that is characterized by diffuse, extreme, and incapacitating breast hypertrophy. Ectopic breast tissue is defined as glands of breast tissue located outside of the normal anatomic breasts. The factors controlling breast growth are complex and not completely understood. Breast tissue including ectopic displays all of the characteristics of normal breast tissue including growth during puberty, pregnancy and lactation. Cases of ectopic vulvar lactating breast with gigantomastia have been described in only few cases in the literature. Hence, we present this rare case of a gravid patient with recurrent lactating vulvar breast and gigantomastia. This was managed conservatively with postpartum resolution of gigantomastia and excision of the vulvar mass.

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INTRODUCTION

Ectopic breast is any type of breast tissue found outside of two normally situated pectoral breasts and found along the primitive embryonic milk lines which extend from the axilla to the groin¹. Incomplete involution anywhere along the primitive milk streak can result in ectopic mammary tissue². The condition ranges from a fully developed breast with a nipple to only accessory nipples or only breast glands¹.

Although the incidence of ectopic breast tissue is reported to occur in 2-6% of women, surprisingly there are only a few case reports in the literature³. Different parts of the body can be involved but the axilla is the most common site accounting for 60%-70% of cases and the most common manifestation is accessory nipple^{1,4}. Vulvar or suprapubic involvement is very rare and few cases have been reported to date⁴. The ectopic breast tissue displays all of the characteristics of normal breast tissue including growth during puberty and pregnancy, lactation, fibrocystic disease and growth of fibroadenoma, phylloides tumors, extramammary Paget's disease and any of the various histological malignancies⁵.

Gestational gigantomastia is a very rare condition and only about 100 cases have been reported in the literature. Physiological enlargement of the breasts occurs at puberty and during pregnancy, when it starts very early and is sustained until delivery. The factors controlling breast growth are complex and not completely understood, although oestrogens, progesterones, prolactin, growth hormone and adrenal steroids are all known to play a role. Sometimes this process goes wrong result-

ing in an excessively large and painful breast called gestational gigantomastia (gravidic gigantomastia, mammary hyperplasia of pregnancy) or virginal hyperplasia when it occurs at puberty. This rare but important condition of the breast not only interferes with breast feeding but may cause severe maternal morbidity and even mortality⁶. True gigantomastia develops rapidly during pregnancy, undergoes regression after delivery, and recurs with subsequent pregnancy⁷.

The aim of this study was to report two extremely rare conditions occurring in a single patient- a case of recurrent lactating ectopic breast in bilateral vulva and gigantomastia in pregnancy of 28-year-old woman. Thus, practicing gynecologists and obstetricians become more confident in managing such a rare condition.

CASE

A 28-year-old Gravida II Para I at 22 weeks dated by her last menstrual period presented to antenatal care clinic. Like the first pregnancy she complains of an increase in the size of both breasts and vulvar mass that appeared at the beginning of the fourth month of pregnancy. During the remaining follow up of pregnancy the size and discomfort breasts became bothersome and sometimes had to ask support during standing from supine and sitting position. With the same rate the vulvar mass became voluminous and caused walking difficult. Previous pregnancy had similar events but with the lesser degree and she gave birth by caesarean section. She has no specific medical, surgical or family history. Routine antenatal tests were normal.

On physical examination both breasts were grossly enlarged and extended to the level of the umbilicus (Fig. 1).



Figure 1: Gigantomastia in a term pregnant lady

The left breast was slightly bigger than the right. Both breasts had dilated veins and exhibited peau d'orange, and a milky discharge could be expressed from each nipple. There are seven firm masses ranging from 5x6 cm to 8x10cm in the labia majora and clitoral region (See fig 2).



Figure 2: Seven nodular masses located in the bilateral labia majora and clitoral region in a term pregnant lady. Four masses were present in the left labium majora and two in the left labium majora and one in periclitoral region.

The routine prenatal laboratories as well as hormone studies were all within normal parameters. With an indication of previous caesarean section scar and vulvar mass, a healthy neonate was delivered by elective caesarean section at term. Few weeks postpartum the vulvar mass started to ooze out whitish non-offensive fluid which later ceased spontaneously three months later when the lady stops breastfeeding. The size of the vulvar masses has decreased during annual examination to 2x3cm to 6x7cm (See fig 3).



Figure 3: Seven vulvar nodular masses one year postpartum.

Surgical excision of vulvar masses revealed seven encapsulated lobules of variable sizes measuring from 2x3cm to 5x8cm. The gross finding of the cross sectioned specimen showed solid areas with multiple cysts having whitish mucinous secretions. Microscopically, the masses were composed of glandular tissue with well organized ducts and lobular structures divided by thin fibrous stroma. The glands showed proliferation of regular looking duct epithelium with some areas of cystic dilatation containing granular or amorphous proteinaceous material. The cytoplasm of the luminal cells was

vacuolated with apical snouts typical of the lactating breast (See fig 4).

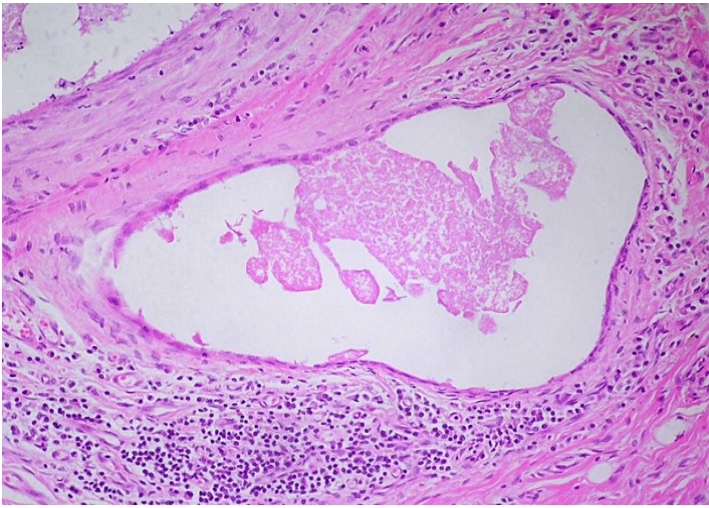


Figure 4: The glands are lined by columnar epithelial cells and an underlying myoepithelial cell layer. The cytoplasm of the luminal cells is vacuolated, and the tumor cells show reactive nuclear changes, including a uniform increase in size, chromatin clearing, and prominent nucleoli.

DISCUSSION

The first case of vulvar ectopic breast is in 1875 by Hartung⁸. Vulvar ectopic breast usually takes the form of breast glandular tissue only; fully developed breasts with nipples are extremely rare. It complete with nipple and have been reported only four times. Vulvar ectopic breasts that consist only of breast ducts and lobules are reported much more frequently⁽¹⁾. Ectopic breast tissue located in vulva may present with growth of the mass and cause pain⁵.

Gigantomastia during pregnancy is characterized by diffuse, extreme, and incapacitating breast hypertrophy⁽⁹⁾. To date, there is no universally accepted definition

for gigantomastia; however, Dancey *et al.* (2007) state that a review of the medical literature suggests that definitions range from a D-cup bra size to breast modification requiring reduction of over 0.8 - 2 kg¹⁰. Massive hypertrophy of the breasts in pregnancy is a very rare (1 in 100 000) condition¹¹.

The aetiology of Gestational Gigantomastia is unknown but various factors have been proposed. These include over-sensitivity to or over-production of hormones such as oestrogen, human chorionic gonadotrophin, human placental lactogen and prolactin⁶. Auto-immune disorders, high IGF-1 and decreased catabolism of hormones have also been proposed^{10, 12}. In our case hormone studies were normal.

Clinical features of gestational gigantomastia may include mastalgia, ulceration, posture problems, back pain and chronic traction injury to 4th/5th/6th intercostal nerves with resultant loss of nipple sensation and obstetrical complications such as decreased fetal growth¹⁰. Lactational change develops mostly in later gestations and often persists after the period of lactation¹.

Treatment of gigantomastia is based on the person's symptoms and may include conservative management, breast reduction, mastectomy with or without reconstruction or a combination of treatments^{10,11}.

Since Bell first reported vulvar lactating breast tissue in 1926, two cases of lactating ectopic breast with breast abnormality have been reported. The patients in these two cases had gigantomastia with myasthenia in one patient and hypertrophy and chronic cystic mastitis of

breast and axillary supernumerary breast tissue in the other patient¹. Similarly, the patient in our case has lactating vulvar breast tissue and breast abnormality-gigantomastia, showing exaggerated hormonal response of the involved Heterotropic Breast.

There are two theories to account for ectopic breast in the vulva, and these are not mutually exclusive. In the traditional theory, normal breast tissue development takes place during the 4th and 5th weeks of gestation—the embryogenic period. Two band-like strips of ectoderm, the mammary ridges, occur along the sides of the embryo during the 6th week of gestation and extend bilaterally from the mid axillae through the normal breasts inferiorly to the medial groin and vulva. Normally, the mammary ridges disappear except for in the pectoral region, where the primary and secondary buds develop initially and mammary glands with lactiferous ducts develop later. As a result of an embryologic abnormality, breast tissue may remain anywhere from the axilla to the vulva and may form any part of or a complete, functioning breast.

The other theory is that vulvar ectopic breast derives from mammary-like glands of the anogenital region. Mammary-like glands are a newly described skin appendage normally present in the interlabial sulcus of the vulva, perineal, and perianal skin. The mammary-like glands not only have a close histologic resemblance to eutopic breast ducts, but, unlike all other skin appendages, are estrogen and progesterone hormone receptor-positive. Furthermore, the proponent of the mammary-like gland theory of vulvar ectopic breast lesions states that there is no convincing evidence for

caudal extension of the milk ridge beyond the pectoral region in humans and that, even if there were, the milk ridge theory could not account for perianal breast lesions¹³.

The ectopic breast tissue is subject to hormone responses and may develop benign and malignant pathologic processes similar to those seen in normally located breast tissue⁽¹⁴⁾. The most common changes are fibrocystic disease, fibroadenoma, and lactation changes. Cystosarcoma phyllodes and primary vulvar breast carcinoma also have been reported¹.

Therefore, although the incidence of ectopic mammary tissue of the vulva is low, this possibility should be considered when evaluating patients with mass lesions of the vulva. Because of the chance of pathological change of ectopic breast tissue, such vulvar masses of breast tissue should be excised.

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