

MAMMOGRAPHIC AND ULTRASOUND FINDINGS IN A CASE OF UNILATERAL CONGENITAL AMAZIA

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Abstract: Scenario: Congenital amazia is a very rare anomaly in which the mammary gland is absent unilaterally or bilaterally in the presence of the nipple-areolar complex (NAC). When one or both breasts are not developed during female puberty, the referenced treatment is mammoplasty for aesthetic purposes (silicone). **Goals:** We reported the case and mammographic and ultrasound images of a woman with unilateral amazema who underwent control mammography. The images were analyzed by a radiologist from a diagnostic imaging clinic in the city of Manaus/AM (Brazil). **Methods:** This is a 41-year-old, healthy, asymptomatic patient who underwent her first screening mammogram at the Clínica de Diagnóstico por Imagem - Sensumed. The analysis and report of the images were performed by a radiologist linked to the site. **Conclusion:** The literature presents few reported cases of unilateral amazia without association with other abnormalities, therefore it is a rare condition of uncertain incidence in the general population. In these cases, the patient must be instructed to undergo screening mammography for breast cancer from the age of 40 onwards in the breast with parenchyma present. More studies on this entity are interesting for the correct management of these patients and a timely surgical approach. **Keywords:** Unilateral breast tenderness, congenital breast anomaly, breast implants, breast cancer screening.

INTRODUCTION

Congenital amazia is characterized by the absence of one or both mammary glands but with both nipple-areolar complexes present (OZSOY et al, 2007). Congenital breast abnormalities can be isolated, associated with genetic syndromes, such as Poland's syndrome, or together with other malformations (PAPADIMITROU et al., 2009).

Cases of amazia associated with facial anomalies including cleft palate, high arched palate, saddle nose, piriformis hypoplasia, acute nasolabial angle and hypertelorism have been reported. However, few reports of isolated amazia have been recorded. Therefore, the incidence of this condition is unknown because it is a very rare or misdiagnosed entity (OLUWAYEMI; AGAJA, 2016).

In this case report, we present a case of unilateral amazonia without other associated malformations and its mammographic and ultrasound findings.

CASE REPORT

A 41-year-old female noticed growth of only the right breast during puberty even though both nipple-areolar complexes were present. Due to breast asymmetry on clinical examination, at the age of 15 she underwent surgery to place a silicone implant bilaterally. At the age of 32, she underwent a new surgical procedure to replace the implants for aesthetic reasons. Asymptomatic, denies history of trauma, other congenital anomalies or comorbidities. She had two children, the first being at age 28 and the second at age 34. Breastfeeding was normally done through the right breast.

The patient sought a diagnostic imaging clinic (Sensumed), located in the city of Manaus/AM (Brazil), in order to perform her first screening mammogram. Digital mammography and tomosynthesis images showed the presence of bilateral breast implants without signs of rupture, normal skin thickness, normal nipple-areolar complexes, predominantly dense right breast and fatty left breast with complete absence of breast parenchyma. Examination evaluated as category 2 (benign findings) according to the BI-RADS® classification.

(Figures 1-6).

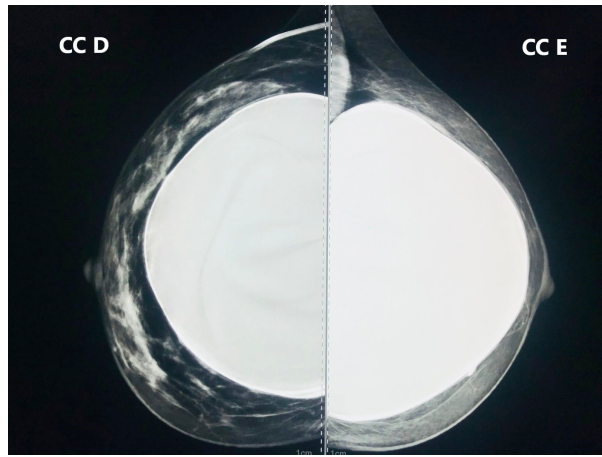


Figure 1: Digital mammography in the craniocaudal view. Breast implants bilaterally. Right breast contains breast parenchyma density. Adipose left breast.

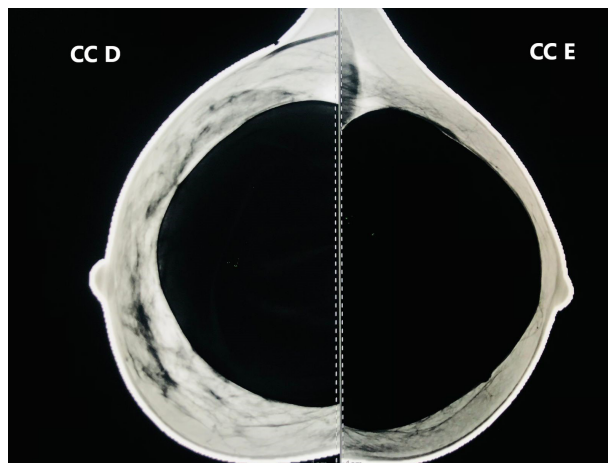


Figure 2: Digital mammography in craniocaudal view with negative image. Breast implants bilaterally. Right breast contains breast parenchyma density. Adipose left breast.

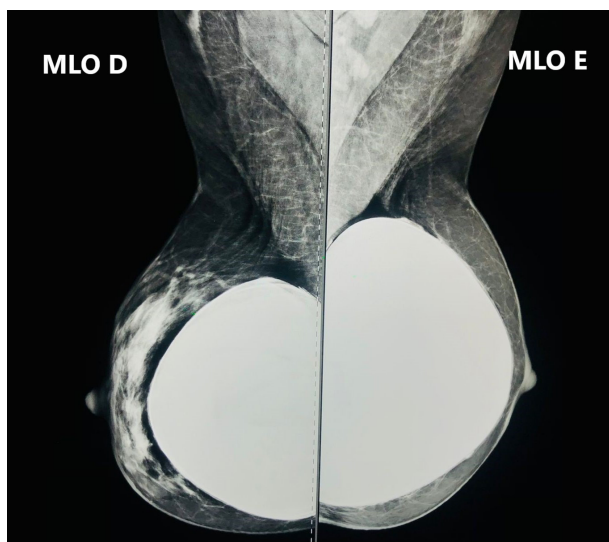


Figure 3: Digital mammography in the oblique medial lateral view. Breast implants bilaterally. Right breast contains breast parenchyma density. Adipose left breast.

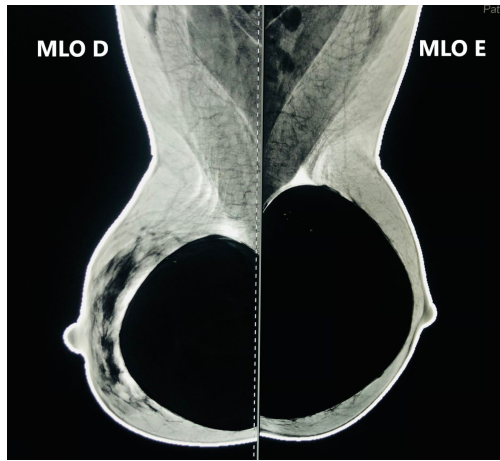


Figure 4: Digital mammography in the oblique mid-lateral view with a negative image. Breast implants bilaterally. Right breast contains breast parenchyma density. Adipose left breast.

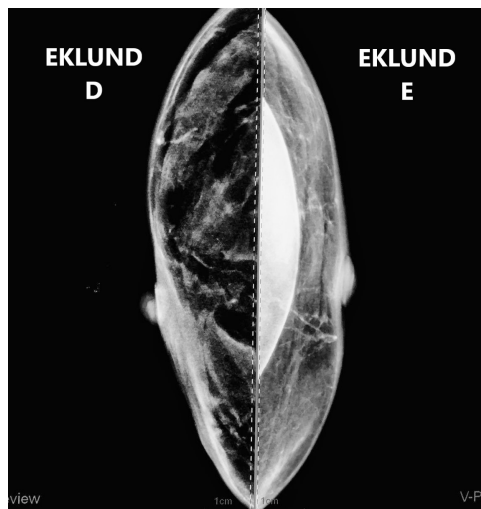


Figure 5: Digital mammography. Eklund maneuver. Right breast contains breast parenchyma density. Adipose left breast.

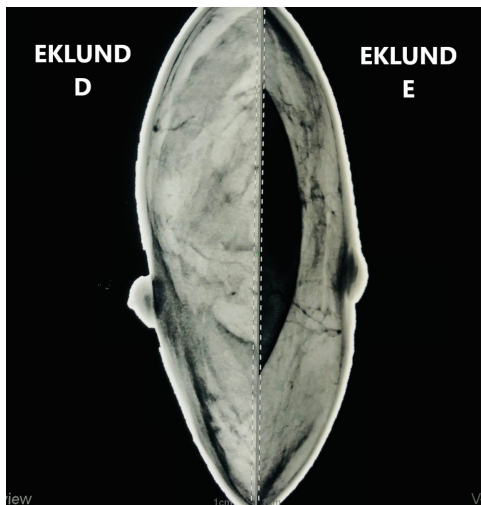


Figure 6: Digital mammography. Eklund maneuver with negative image. Right breast contains breast parenchyma density. Adipose left breast.

Likewise, complementary breast ultrasound showed the presence and integrity of breast implants, also not showing left breast parenchyma. In addition, anechoic cystic images with regular contours and thin walls were seen, located in the superior lateral

quadrant of the right breast, measuring 0.5 x 0.3 cm and two in the inferior lateral quadrant, smaller than 0.3 x 0.2 cm. Examination evaluated as category 2 (benign findings) according to the BI-RADS® classification. (Figures 7-10).

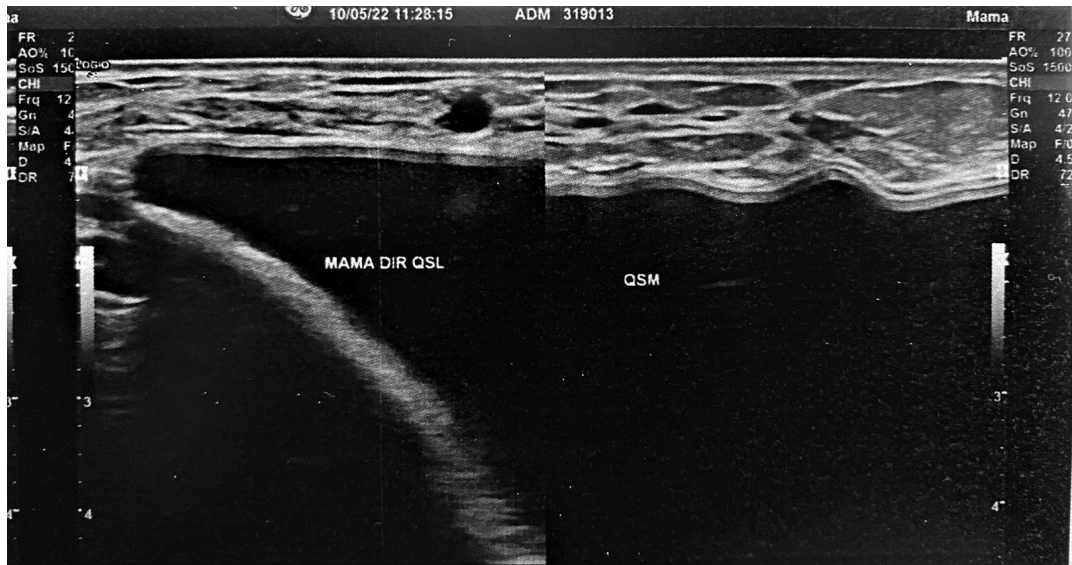


Figure 7: Right breast ultrasound. Breast parenchyma presenting a homogeneous background echotexture with a predominance of fibroglandular tissue in the midst of adipose tissue. Intact breast implant with regular contours, with anechoic homogeneous content. Cystic image in the lateral superior quadrant, measuring 0.5 x 0.3 cm.

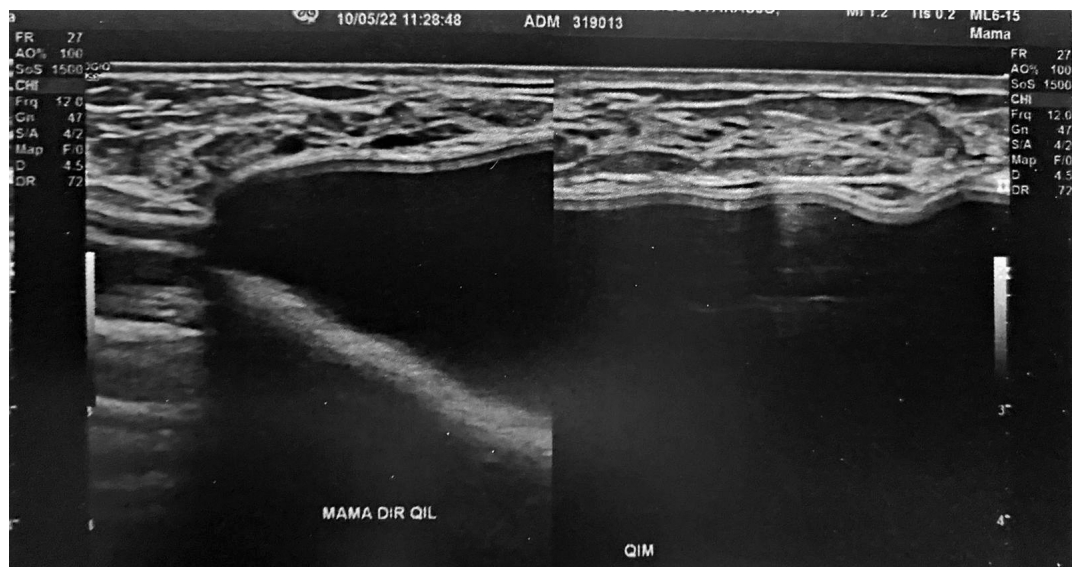


Figure 8: Right breast ultrasound. Breast parenchyma presenting a homogeneous background echotexture with a predominance of fibroglandular tissue in the midst of adipose tissue. Intact breast implant with regular contours, with anechoic homogeneous content. Two cystic images in the inferior lateral quadrant, smaller than 0.3 x 0.2 cm.

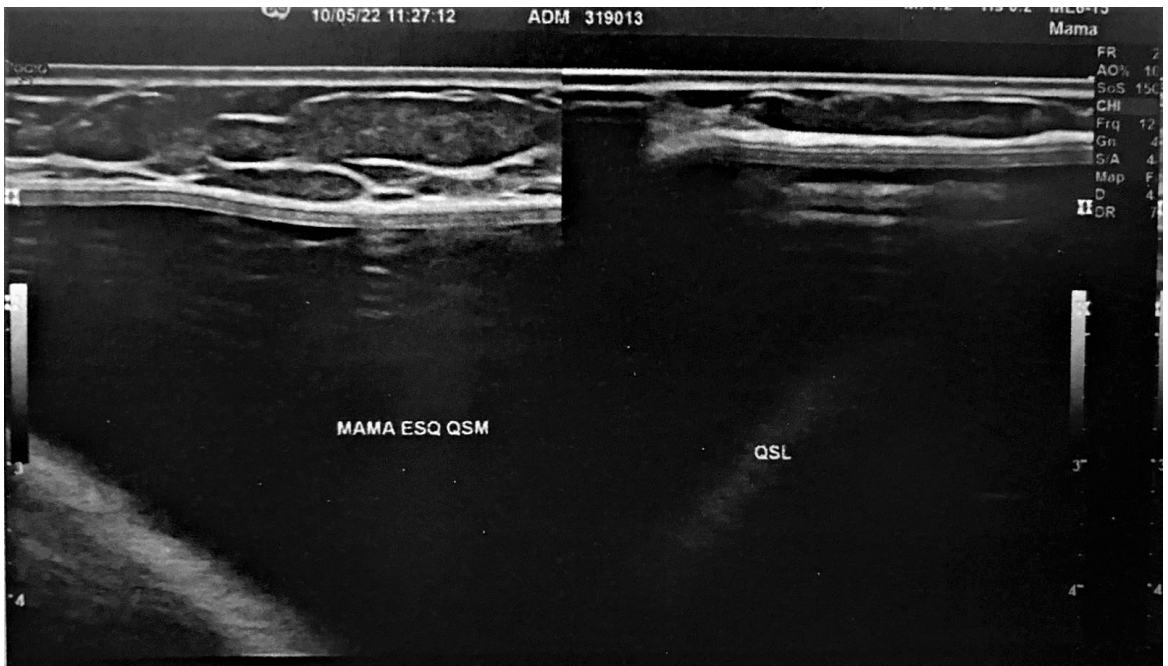


Figure 9: Left breast ultrasound. Intact breast implant with regular contours, with anechoic homogeneous content. Subcutaneous cellular tissue is observed, with no evidence of breast parenchyma.

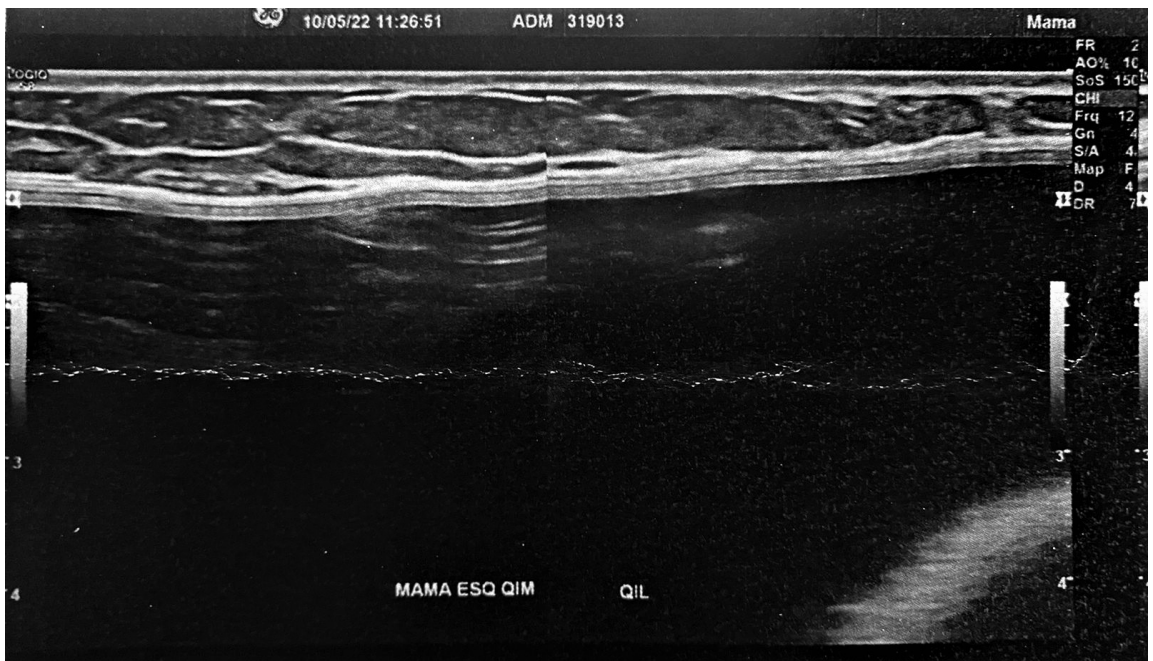


Figure 10: Left breast ultrasound. Intact breast implant with regular contours, with anechoic homogeneous content. Subcutaneous cellular tissue is observed, with no evidence of breast parenchyma.

DISCUSSION

Breast development begins between 4 to 6 weeks of gestation, when specific progenitor cells can be seen. The proliferation of paired areas of epithelial cells in the epidermis of the thoracic region occurs at the 35th week of gestation. Such areas extend in a line from the axillary region to the inguinal region, forming two ridges, which are called mammary ridges or milk lines. Most of these breast lines atrophy, except for paired solid epithelial masses in the fourth intercostal space, which form the primary breast bud. Areolas appear in the fifth month of gestational age as areas of skin devoid of hair follicles surrounding the future nipples. By the sixth month of gestation, the basic structure of the mammary gland is established and breast tissue may be apparent. Amazia and other breast anomalies are often associated with cephalic, cervical and upper limb malformations. For this reason, this spectrum of mammary anomalies is believed to be secondary to a disruptive sequence involving the brachiocephalic arterial system early in embryonic life, leading to a failure in the development of the primitive mammary gland. (OZSOY et al, 2007; NSO-ROCA; AGUIRRE-BALSALOBRE; JUSTE RUIZ, 2012; OLUWAYEMI; AGAJA, 2016).

Congenital hypoplastic breast abnormalities include atelia (absence of NAC), amastia (lack of breast tissue and NAC), hypoplastic breasts (deficiency in the amount of breast parenchyma) and amazia (absence of breast tissue with presence of NAC). These abnormalities may occur alone or be associated with a syndrome, such as Poland syndrome, where there is absence of the breast and the ipsilateral pectoralis major and minor muscles, syndactyly, lack of ribs, chest wall depression, and limited subcutaneous fat (NSO). -ROCA; AGUIRRE-BALSALOBRE; JUSTE RUIZ, 2012). Reports show that amazia may be associated with choanal atresia,

hemiface, limbs and vertebrae abnormalities (OZSOY et al, 2007; PAPADIMITROU et al., 2009). On the other hand, there are reports showing cases of unilateral amathia with no other findings (KACHEWAR, 2010; NSO-ROCA; AGUIRRE-BALSALOBRE; JUSTE RUIZ, 2012; RAO, 2015).

Usually, amazia is diagnosed during female puberty, when one or both breasts do not develop in the presence and evolution of other secondary sexual characteristics (PAPADIMITROU et al., 2009; NSO-ROCA; AGUIRRE-BALSALOBRE; JUSTE RUIZ, 2012).

Congenital breast anomalies can harm the mental health of affected women by generating concerns about body image and the ability to breastfeed, mainly because they are noticed during an emotionally vulnerable period such as adolescence (NSO-ROCA; AGUIRRE-BALSALOBRE; JUSTE RUIZ, 2012). ; RAO, 2015; OLUWAYEMI; AGAJA, 2016;).

The management of the amazia is done with surgery to place a breast implant, as in the case reported. In cases of unilateral hypoplasia, such reconstruction can be performed after full growth of the contralateral breast (NSO-ROCA; AGUIRRE-BALSALOBRE; JUSTE RUIZ, 2012). Another possible surgical approach is the use of adjustable implants/expanders to maintain the size of the breasts suitable for the adolescent's growth and development phase, making it possible to increase the breast volume as the patient grows. (DREIFUSS; MACISAAC; GRUNWALDT, 2014).

Breastfeeding with a functioning breast can normally occur in cases of unilateral breast tenderness, but in bilateral cases, these women must resort to milk banks or formulas to feed their children. (KACHEWAR, 2010).

Annual mammographic screening is recommended for women between 40 and 74 years old, according to the Brazilian College

of Radiology and Diagnostic Imaging, the Brazilian Society of Mastology and the Brazilian Federation of Gynecology and Obstetrics Associations. (URBAN et al., 2017). For this reason, the patient presented had her first mammogram at age 41. Since amazia is a condition where there is no breast parenchyma, we do not suggest performing an annual mammogram on the affected side, only on the breast with fibroglandular tissue present. Therefore, cases of bilateral amazia do not require screening for breast cancer. However, we emphasize that ultrasound and magnetic resonance imaging of the breasts are essential for the evaluation of the integrity of breast implants, in addition to being good complementary exams for the identification of suspicious lesions for breast malignancy.

The Food and Drugs Administration (2020) recommends that breast implant rupture screening be performed with MRI, starting between five to six years after surgery and every two to three years after the first examination. Ultrasonography is recognized as a good alternative method for this type of screening. Therefore, regular monitoring of patients with breast implants is essential, as is the case portrayed.

CONCLUSION

Congenital amazia is a very rare breast anomaly, especially when it presents unilaterally and is not associated with other findings. In addition, it is a condition rarely reported in the literature and of unknown incidence. In these cases, imaging methods show the absence of breast parenchyma on the affected side and are capable of helping to define the correct diagnosis. Patients with unilateral breast enlargement must undergo screening mammography in the contralateral breast from the age of 40, which may be complemented with ultrasound and/or magnetic resonance imaging. Further studies

on this anomaly and its genesis are interesting for the correct management and follow-up of these patients.

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