

Ectopic Breast Tissue in the Vulvar Region: A Case Report and Review of Literature

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Abstract

Case Report

Ectopic breast tissue (EBT) is a rare congenital anomaly. It typically develops along the embryonic mammary ridge, with common sites including the axillae, while its presence in the vulvar region is exceptionally rare. This condition results from abnormal migration of breast tissue during embryonic development when the primordial mammary lines fail to regress completely. Diagnosis is challenging and requires clinical examination, imaging, and histopathological analysis to exclude malignancy. Treatment generally involves surgical excision to alleviate symptoms and reduce the risk of malignant transformation. We report a case of vulvar supernumerary breast tissue in a 46-year-old woman, confirmed through imaging and histopathological analysis following excision. This case highlights the importance of early diagnosis and surgical management to prevent complications.

Keywords: Breast, Ectopic Breast Tissue, Vulva.

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INTRODUCTION

Ectopic breast tissue (EBT) is a rare congenital anomaly characterized by the presence of mammary glands outside their usual anatomical location, occurring in approximately 1-3% of women (Famá *et al.*, 2016, Lapid, 2013). While it is most commonly found in the axillary region, its vulvar localization remains extremely rare and presents significant diagnostic and therapeutic challenges. This condition results from incomplete regression of the embryonic mammary ridges during fetal development, leading to the persistence of ectopic mammary structures. Due to its rarity, vulvar EBT is often misdiagnosed, as it can mimic various benign and malignant vulvar lesions, including Bartholin gland cysts, epidermal inclusion cysts, and vulvar carcinoma. Accurate diagnosis requires a thorough clinical evaluation, often supplemented by imaging and histopathological analysis to rule out malignancy. Given its potential for malignant transformation, early recognition and appropriate treatment, primarily through surgical excision, are crucial to alleviate symptoms and prevent complications. In this article, we report a rare case of vulvar supernumerary breast tissue in a 46-year-old woman, contributing to the limited body of literature on this condition and highlighting the importance of

considering this diagnosis when evaluating vulvar masses.

CASE REPORT

A 46-year-old Tunisian woman presented with a progressively enlarging swelling of the right labium majus, evolving over two years with worsening symptoms in the past six months. Her medical history included the excision of axillary supernumerary breast tissue at age 17, appendectomy, and right salpingectomy for ectopic pregnancy in 2016. Gynecologically, she had menarche at 13, regular cycles, and menopause at 40. Clinical examination revealed normal breasts without palpable masses, nipple discharge, or lymphadenopathy, but bilateral axillary supernumerary breast tissue was noted. A 4 cm mobile, elastic, mildly tender subcutaneous mass was palpated in the right labium majus. Complementary investigations included mammography, which showed benign bilateral macrocalcifications (BI-RADS 2), and soft tissue ultrasound of the Right Labium Majus revealing a 47 × 25 mm hypoechoic, mildly vascularized mass resembling glandular tissue. Abdominopelvic CT scan identified a fatty density in the right inguinal canal with mild vulvar prolapse, and perineal MRI confirmed a Subcutaneous mass of the right labium majus, well-

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defined, encapsulated fatty mass with glandular components ($63 \times 27 \times 70$ mm), suggestive of ectopic breast tissue. Surgical management involved an elliptical incision for complete excision of the vulvar mass with preservation of underlying structures, meticulous hemostasis, and two-layer closure for optimal healing. Bilateral axillary supernumerary breast tissue was also

excised through careful subcutaneous dissection while preserving neurovascular elements. Histopathological examination confirmed the benign nature of the excised tissue. Ethical approval was obtained from the institutional review board, and written informed consent was secured from the patient for case publication.



Figure 1: Intraoperative View: Incision of the Right Labium Majus Tumor



Figure 2: Perioperative View of the Perivulvar Tumorectomy

DISCUSSION

Supernumerary breast tissue, or ectopic mammary tissue, is a rare anomaly most commonly observed in women, though it has also been reported in men. These abnormalities are generally sporadic, though familial occurrences have been documented. While ectopic tissue can be found in various locations, the axilla remains the most common site, accounting for 60-70% of cases. Vulvar involvement remains exceptional, with only a few cases reported to date (Godoy-Gijón *et al.*, 2012), as evidenced by the case presented in our study.

The embryonic development of mammary glands provides a fundamental explanatory basis for this anomaly. During the early weeks of gestation, the mammary glands originate from the ectoderm. The thickening of this layer gives rise to the primitive milk line, also known as the mammary ridge. This structure extends from the axillary regions to the inguinal folds. Several mammary primordia appear along this ridge during embryonic development (Teixeira *et al.*, 2020). In humans, most of these primordia spontaneously disappear, leaving only one pair of functional mammary glands located on either side of the thorax. This process is marked by the persistence of two pectoral buds, while the remainder of the mammary ridge disappears around

the 6th week of gestation. In rare cases, a mammary primordium persists outside its usual location, such as in the vulvar region.

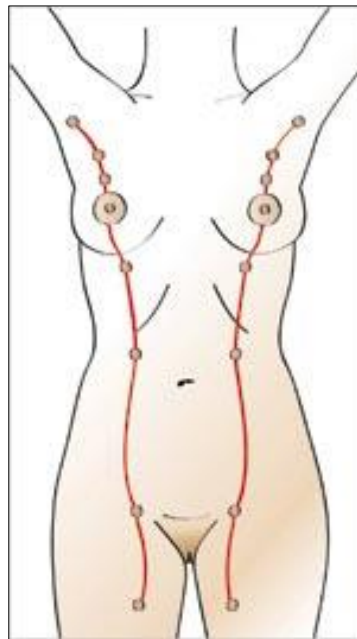


Figure 3: Natural milk lines in the human body (B. Chaput, M. Courtade-Saïdi, 2017)

Two main hypotheses explain the appearance of ectopic vulvar mammary tissue (Zhang *et al.*, 2023): The Mammary Ridge Theory which suggests that ectopic mammary tissue originates from an embryonic mammary primordium that did not regress during the 5th and 6th weeks of embryonic development. Under the influence of placental hormones, this tissue can proliferate, particularly during hormonally significant periods such as pregnancy (Voerman *et al.*, 2019); and the Independent Gland Theory proposing that ectopic tissue results from the development of a genital glands mimicking mammary tissue. This tissue has the potential to develop benign lesions such as fibroadenomas or malignant lesions like invasive adenocarcinoma (Kalyani *et al.*, 2014).

Ectopic breast tissue often referred to as supernumerary nipple or accessory breast, encompasses a much broader spectrum. (Ghosn *et al.*, 2007). This concept can be classified into two main categories: Supernumerary breast, characterized by the presence of a persistent or atrophic mammary gland accompanied by a nipple and/or areola and aberrant breast, defined by the exclusive presence of mammary tissue, without an associated nipple or areola (Nihon-Yanagi *et al.*, 2011, Toman *et al.*, 2008). A classification proposed by Kajava details all the clinical characteristics based on tissue composition and anatomical location (Surcel *et al.*, 2023). According to this classification, our case corresponds to class IV ectopic breast tissue at the level of the right labium majus and class V at the axillary level on both sides (Table 1).

Table 1: Kajava classification for supernumerary breast tissue

Class	Description
Class I (Polymastia)	Complete breast(s) with the nipple, areola, and glandular tissue
Class II (Supernumerary breast without areola)	Nipple and glandular tissue but no areola
Class III (Supernumerary breast without nipple)	Areola and glandular tissue but no nipple
Class IV (Mamma aberrata)	Glandular tissue only
Class V (Pseudomamma)	Nipple and areola but without glandular tissue (replaced by fat)
Class VI (Polythelia)	Nipple only
Class VII (Polythelia areolaris)	Areola only
Class VIII (Polythelia pilosis)	Patch of hair only

Ectopic breast tissue in the vulva presents significant diagnostic and therapeutic challenges, as this anomaly can easily be mistaken for other benign or malignant vulvar masses. The differential diagnosis notably includes vulvar carcinoma, Bartholin gland disorders, epidermal cysts, and femoral hernias.

Ectopic breast tissue lesions become symptomatic primarily at menarche, during pregnancy, or lactation. They may exhibit cyclic size fluctuations synchronized with the menstrual cycle. During pregnancy and lactation, these lesions tend to enlarge, sometimes accompanied by lacteal secretions. These cyclic variations or the onset of specific symptoms during these periods strongly support the diagnosis of accessory breast tissue.

Benign pathological changes, such as fibroadenomas, fibrocystic changes, phyllodes tumors, and intraductal papillomas, have been reported. Although rare, malignant lesions such as carcinomas, adenocarcinomas, and sarcomatous degeneration have also been observed, with no evidence of an increased frequency in accessory breasts (Hong *et al.*, 2009). Breast cancer arising in ectopic breast tissue is a rare tumor, accounting for 0.2% to 0.6% of all breast cancers (Haddad *et al.*, 2012). The malignant degeneration of ectopic breast tissue can present both diagnostic and therapeutic challenges.

A histopathological study is essential to confirm the clinical diagnosis and exclude the presence of a neoplasm. Due to their potential for malignant transformation, these lesions require early surgical management to ensure optimal treatment and minimize risks.

In our case, surgical resection provided effective management and confirmed the nature of the mass through histopathological examination. However, the long-term risk of recurrence remains uncertain, highlighting the importance of regular clinical follow-up (Nihon-Yanagi *et al.*, 2011).

Future Research Prospects

As future work, we aim to elucidate the factors driving the persistence and growth of ectopic breast tissue in unusual locations, such as the vulva. We plan to conduct longitudinal studies with larger cohorts to enhance our understanding of disease progression, recurrence, and malignant risk.

CONCLUSION

In conclusion, this case highlights the rarity and clinical implications of vulvar ectopic breast tissue. A thorough understanding of embryology and the underlying pathophysiological hypotheses is essential for accurate diagnosis. Due to its potential for malignant

transformation, treatment primarily relies on surgical resection. This work contributes to the existing knowledge of this atypical location and emphasizes the importance of long-term follow-up. It is crucial to integrate these factors into clinical management to ensure appropriate care and minimize the risks of recurrence or future complications.

REFERENCES

- Chaput, B., & Courtade-Saïdi, M. (2017). 1: Embryologie et histologie mammaire | Medicine Key. <https://clemedicine.com/1-embryologie-et-histologie-mammaire/>
- Famá, F., Cicciú, M., Sindoni, A., Scarfó, P., Pollicino, A., Giacobbe, G., Buccheri, G., Taranto, F., Palella, J., & Gioffré-Florio, M. (2016). Prevalence of ectopic breast tissue and tumor: A 20-year single center experience. *Clinical Breast Cancer*, 16(4), 107–112. <https://doi.org/10.1016/j.clbc.2016.03.004>
- Ghosn, S. H., Khatri, K. A., & Bhawan, J. (2007). Bilateral aberrant axillary breast tissue mimicking lipomas: Report of a case and review of the literature. *Journal of Cutaneous Pathology*, 34(s1), 9–13. <https://doi.org/10.1111/j.1600-0560.2006.00713.x>
- Godoy-Gijón, E., Yuste-Chaves, M., Santos-Briz, Á., Esteban-Velasco, C., & De Unamuno-Pérez, P. (2012). Mama ectópica vulvar. *Actas Dermo-Sifiliográficas*, 103(3), 229–232. <https://doi.org/10.1016/j.ad.2011.02.015>
- Haddad, H., Bourhaleb, Z., El Harroudi, T., Mezouar, L., & El Hfid, M. (2012). Breast cancer in ectopic breast tissue: Report of 2 cases. *The Pan African Medical Journal*, 13, 50–50.
- Hong, J. H., Oh, M. J., Hur, J. Y., & Lee, J. K. (2009). Accessory breast tissue presenting as a vulvar mass in an adolescent girl. *Archives of Gynecology and Obstetrics*, 280(2), 317–320. <https://doi.org/10.1007/s00404-008-0905-z>
- Kalyani, R., Srinivas, M. V., & Veda, P. (2014). Vulval fibroadenoma—A report of two cases with review of literature. *International Journal of Biomedical Science: IJBS*, 10(2), 143–145.
- Lapid, O. (2013). Breast reconstruction after mastectomy for gestational gigantomastia. *Aesthetic Plastic Surgery*, 37(2), 388–391. <https://doi.org/10.1007/s00266-012-0054-4>
- Nihon-Yanagi, Y., Ueda, T., Kameda, N., & Okazumi, S. (2011). A case of ectopic breast cancer with a literature review. *Surgical Oncology*, 20(1), 35–42. <https://doi.org/10.1016/j.suronc.2009.09.005>
- Surcel, E., & Koljonen, V. (2023). Kajava classification: The person and the research. *Aesthetic Plastic Surgery*, 47(5), 2177–2178. <https://doi.org/10.1007/s00266-023-03451-7>

- Teixeira, I. P., Azzopardi, C., Chowdhury, R., Zsolt, O., Castro, M., & Teh, J. (2020). Ectopic breast tissue presenting as an enlarging abdominal mass. *Radiology Case Reports*, 15(6), 733–740. <https://doi.org/10.1016/j.radcr.2020.02.028>
- Toman, N., Buschmann, A., & Muehlberger, T. (2008). Ectopic breast cancer without breast parenchyma. *The Breast*, 17(1), 3–4. <https://doi.org/10.1016/j.breast.2007.07.043>
- Voerman, E., Santos, S., Inskip, H., Amiano, P., Barros, H., Charles, M.-A., Chatzi, L., Chrousos, G. P., Corpeleijn, E., Crozier, S., Doyon, M., Eggesbø, M., Fantini, M. P., Farchi, S., Forastiere, F., Georgiu, V., Gori, D., Hanke, W., Hertz-Picciotto, I., Gaillard, R. (2019). Association of gestational weight gain with adverse maternal and infant outcomes. *JAMA*, 321(17), 1702-1702. <https://doi.org/10.1001/jama.2019.3820>
- Zhang, L., Yao, X. L., Zhu, X., Fu, Y., & Li, S. (2023). Ectopic breast tissue of the vulva. *Polish Journal of Pathology*, 74(2), 141–143. <https://doi.org/10.5114/pjp.2023.128787>