

# A Rare Case of Solitary Unilateral Ectopic Breast Tissue in Vulva Region in a Young Female Patient: Case Report of Youngest Female Patient from Indian Subcontinent and Case Series Analysis

## Case report

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### Abstract

Ectopic Breast tissue (EBT) is a rare entity. It occurs due to the incomplete regression of the milk line which extends from axilla to groin and occurs commonest in axilla and rarest in vulva. EBT has the potential to turn malignant, thus the condition needs to be followed up including cytology if required. However, rarity of the condition frequently delays the diagnosis which can lead to morbidity and mortality in a patient. There is urgency of formulation of guidelines for management of such patients which should be addressed.

We report the youngest patient of healthy EBT from the Indian subcontinent in a 25-year-old woman and discuss review of literature.

**Keywords:** Ectopic Breast Tissue; Vulva; Estrogen Receptor; Progesterone Receptor; Immuno histochemistry

### Introduction

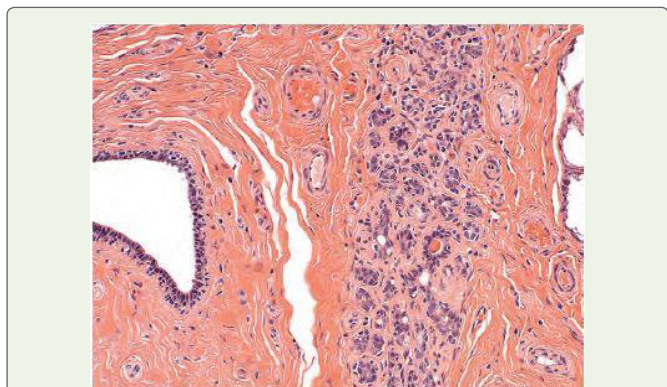
Ectopic breast tissue (EBT) is a rare entity with an incidence of 2-6% in women & 1-3% in men, with women to men ratio of 2:1 [1]. It is a developmental anomaly due to incomplete regression of milk line and presents as residual breast tissue with or without nipple areolar complex in milk ridge line in addition to normal pectoral tissue. It can be solitary or multiple [2,3]. EBT in vulva, including benign and malignant pathology, are reported in literature in 129 patients in age group of 18-89 years [4], of which 20 patients with healthy breast tissue in EBT is reported from 29-69 years. From the Indian subcontinent only 4 such patients are reported in women from 28-60 years. [5-8]. We report the youngest female patient of 25 years old of healthy breast tissue (HBT) in EBT from the Indian subcontinent and discuss review of literature, management dilemmas and urgency to formulate guidelines in such patients.

### Case History

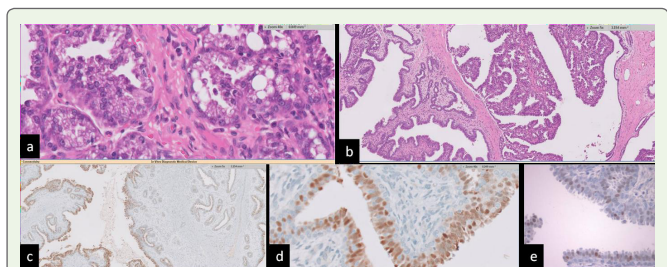
A 25-year-old pregnant lady reported with non-painful swelling on left side of vulva at eight month of gestation which progressively increased after delivery. A soft mobile mass of 3x2x1cm was palpated in the vulva. Excisional biopsy was done under local anesthesia. Cut surface revealed grey white and lobulated mass. Differential diagnosis of lipoma, Bartholin Cyst, and Hidradenitis Papilliferum was made. On histologic evaluation on staining with hematoxylin and eosin, lobules of healthy breast tissue with lactational changes were seen. Immunohistochemistry revealed estrogen receptor (ER) positive (ALL RED Score 8) (Figure 2 c-d) and focally positive for progesterone receptor (PR) (Figure 2e). [9] Diagnosis of EBT (Figure 2a) (Figure 2b) was made.

### Discussion

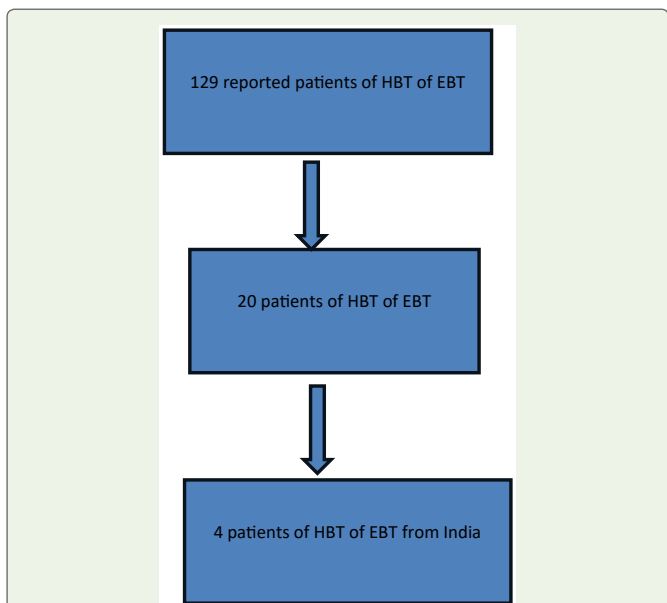
The present patient is the youngest reported patient (25 years



**Figure 1:** Ectopic Breast tissue resembling Normal Breast tissue (Source: Benign Cysts, Rests, and Adnexal Tumors of the Vulva. In: Crum Christopher P, Nucci Marisa R, Lee Kenneth R, editors. Diagnostic gynaecologic and obstetric pathology, 2nd ed. Philadelphia: Elsevier Saunders; 2011. p. 89.



**Figure 2:** Histopathological analysis of present patient (a & b) Ectopic breast with Adenosis, Epitheliosis and Lactational Changes (a) (H and E 5X), (b) (H and E 40X). (c & d) Estrogen Receptor Showing diffuse and intense positivity (ALL RED Score 8) (c) (IHC 5X), (d)(IHC 40X). (e) Progesterone Receptor showing focal positivity (IHC 40x)



**Figure 3:** PRISMA representation of systemic review of HBT of EBT HBT: Healthy Breast Tissue; EBT: Ectopic Breast Tissue

old) of HBT of EBT in vulva from the Indian subcontinent. Previous literature reveals 126 patients of EBT of which HBT of EBT was reported in 20 patients in the age group from 29-69 years including, 4 such patients in age group of 28-60 years from the Indian subcontinent are reported. [4-8].

Primary milk line develops in human embryo at 6 week gestation and extends from axilla, through thoracic (pectoral, chest area), abdominal, inguinal, to pubic area, where it traverses bilaterally and atrophies by 9 wks of gestation except in chest where normal breast tissue develops. Incomplete regression of primary milk line gives rise to EBT anywhere in milk line and appears as smooth well circumscribed mass with/ without nipple and /or areola. EBT may not be clinically apparent unless it becomes site of pathological process or physiological changes. The commonest site of EBT is axilla, however it is reported in unusual sites like vulva, chest, face, ear, abdomen, inguinal region, perineum, perianal region, heart and foot. EBT on vulva is rarely reported and may present as an abscess, Bartholin abscess, fibroadenoma, recurrent vulval Fibroadenoma, phyllodes tumors and atypical hyperplasia [10-16]. EBT in vagina was reported at a rare site of median episiotomy [17]. Extra vulval EBT was reported in infraclavicular chest wall, in perineal region of a gentleman, in inguinal region and as an enlarging abdominal mass [18-21]. Usually EBT occurs without nipple and/or nipple areolar complex. However, EBT with nipple areola in vulva with lactation from vulva and perianal mass is also reported including Indian subcontinent [22-23].

Lactational changes in EBT vulva & lactational adenoma of EBT have also been documented in literature [24,25]. Rarely, EBT may present outside ventrally placed milk ridge such as on the face or foot which can be explained by displacement or migration of milk ridge anlage [26]. Heterotopic breast type epithelial inclusion was seen in atrial tissue in surgically explanted native heart of 72-year-old transplant recipient [27]. EBT occurs commonly in reproductive age group, however rarely it's reported in postmenopausal women [28]. The youngest reported patient of HBT of EBT of vulva was reported in an adolescent girl of 18 years [29]. In literature HBT in EBT vulva has been reported in 20 patients with 4 from Indian subcontinent [30-45]. The management of EBT is similar to a growth seen in healthy breast. Previously, patients of EBT which revealed fibroadenoma and HBT, local excision of EBT was done. [2, 3,5,7,8,30-43] [Table 1] as in the present patient. If EBT reveals carcinoma, then according to the stage of carcinoma, neoadjuvant chemotherapy may be added. In a patient of EBT in infraclavicular area, cytology revealed invasive duct carcinoma and patient was treated with chemotherapy and hormone therapy.

The differential diagnosis of EBT is lactational breast, Hidradenoma papilliferum, Bartholin Cyst. Lactational breast is diagnosed by Immunohistochemical (IHC) demonstration of ER and PRs in the present patient. Hidradenoma papilliferum is a well circumscribed mass lesion with papillary glandular epithelium with associated myoepithelial cells and a fibrovascular core forming true papillae. Bartholin cyst show epithelial lining as squamous, transitional or mucinous.

EBT is under same hormonal influences as normal breast and

**Table 1:** Healthy Ectopic Breast Tissue (EBT) reported in literature

Author	Country	Design	Age	Lesion Size (cm)	Location	Hormonal status	Diagnosis	Management
*D Sanchita et al 2023[6]	India	Case report	43 F	5*3 cm Healthy EBT	RLM	Reproductive age	Healthy breast tissue (HBT)	Local Excision
De La Vegas et al 2020[30]	USA	Case report	31F	5*3*3	LLM	Postpartum	Lactational tissue HBT	Local Excision
Ardell S et al 2019[31]	Canada	Case report	30F	7cm	Vulva	Pregnancy	HBT (Benign)	Local Excision
Mayor RB et al 2019[32]	Austria	Case report	29F	6cm	Vulva	Post partum	Lactational tissue (Benign)	Local Excision
Baradwan S et al 2018[2]	Saudi Arabia	Case report	27F	6*4	RLM	Postpartum	HBT	Local Excision
Zhou M et al 2017[33]	China	Case report	26F	6*5	Vulva	Pregnancy	HBT	Local Excision
*Kalyani et al 2014[5]	India	2 case reports	26 F 45 F	4.5*2.4*1.2 FA 12*8*6.5 FA	RLM LLM)	Reproductive age	Benign	Excision biopsy
Zhang et al 2014[34]	China	Case report	25F	6*5	Vulva	Pregnancy	HBT	Local Excision
Al- Badani IA 2014[35]	Saudi Arabia	IN THE	48F	7*4	RLM	Perimenopausal	HBT	Local Excision
Wagner ID et al [36] 2013	USA	Case report	23F	6 cm	Vulva	Pregnancy	HBT	Local Excision
*Deshmukh SN et al 2012[7]	India	Case report	45 F	4*4 cm Healthy normal EBT	L L M	Perimenopausal	Healthy breast tissue	Local Excision
Godoy-Gojon 2012[37]	Spain	Case report	24F	4*2	LLM	Reproductive age	HBT	Local Excision
Hanief et al 2011[38]	UK	Case report	40F	3.5*4.5	Perineum	Reproductive age	HBT	Local Excision
Sundaram SS et al 2009[39]	UK	Case report	37F	1 cm	Clitoris	Reproductive age	HBT	Local Excision
Dordevic M et al 2008[40]	Serbia	Case report	27F	5*3	LLM	Reproductive age	HBT	Local Excision
Duvuur S et al 2007[41]	UK	Case report	41F	1*1	Vulva	Reproductive age	HBT	Local Excision
England E 2007[42]	UK	Case report	45F	1.5*5	LLM	Postmenopausal	HBT	Local Excision
Baldsley M et al 2004[43]	Australia	Case report	31F	7*4	Vulva	Postpartum	HBT	Local Excision
Kapila K et al 1998[53]	Kuwait	Case report	30F	2*1	Multiple sites	Pregnancy	HBT	Local Excision
*Prasad KR et al 1995 [8]	India	Case report	30 F	Fibroadenoma	Vulva	Reproductive Age	Benign	Excision biopsy

RLM: Right labia majora/LLM: Left labia majora/HBT : Healthy Breast tissue

undergoes same physiological and pathological changes as of normal breast and thus may transform to malignant variant. Most cases of EBT are benign however few malignant cases of EBT are also reported which includes papillomas, intraductal carcinoma, invasive ductal carcinoma, metachronous vulval EBT adenocarcinoma, invasive lobular carcinoma and mucinous Adenocarcinoma [46-51]. Primary carcinoma should be differentiated from metastatic breast carcinoma by initial diagnosis by fine needle aspiration cytology and confirmation by histopathological examination [52-53]. Benign EBT may convert to malignant morphology.

The entity of HBT presenting as EBT in vulva is rare. However, it may convert to malignant variant anytime during the course of the lifespan of the patient. Thus, it is recommended that a treating physician on encountering any mass in vulva should be vigilant in its diagnosis including cytology and regular follow ups to rule out malignancy. The mass maybe removed if the patient desires so due to discomfort or cosmetic reasons as in the present patient. There

are no guidelines in literature on management of HBT in EBT vulva and thus requires urgency to formulate guidelines on screening and follow up of EBT.

### Conclusion

EBT should be suspected in any subcutaneous mass along milk line and should be regularly followed up due to its malignant potential. Due to scarcity of such patients in reported literature, every case of EBT should be reported and documented to formulate guidelines.

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