



Basal Cell Carcinoma of Vulva with Situs Inversus Totalis: A Rarest of the Rare Phenomenon

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Introduction

Basal Cell Carcinoma, known earlier as basal cell epithelioma, is the commonest cancer in Humans. It arises primarily on sun-damaged skin, in head-neck region (85%) and rarely develops on the mucous membranes or palms and soles, but can involve the trunk and extremities too. BCC is associated with damage caused by exposure to sunlight and radiation, arsenic ingestion, immunosuppression and inherited conditions, such as xeroderma pigmentosum and nevoid BCC syndrome [1,2]. Usually it's a slow-growing tumor with negligible chance of distant metastasis via lymphatic or hematogenous route. Although

rarely fatal, BCC has the tendency to be highly destructive, locally infiltrative, hence also known as Rodent Ulcer, and disfigure local tissues when it's not adequately and timely treated. On examination, BCC usually appears as pink coloured, pearly papules with overlying ulceration or telangiectatic vessels [1].

However, BCC of the vulva accounts for less than 1% of all BCCs [2]. Basal Cell Carcinoma of Vulva, or of any site in-fact, as they seldom spread to lymph nodes, are appropriately treated with surgery alone. The current mainstay of BCC treatment in-

Abstract

Basal Cell Carcinoma, primarily occurring due to sunlight exposure, radiation, arsenic, immunosuppression or inherited conditions like Xeroderma pigmentosum, mostly present in sun-exposed areas of the head and neck but can also present elsewhere. It's a slow growing tumour with negligible chance of distant metastasis via lymphatic or hematogenous route, but highly destructive and locally infiltrative. Basal Cell Carcinoma of Vulva is rarely seen (1%). A 57 years old postmenopausal female presented with such a rare case of Vulval Basal Cell Carcinoma at the Gynae OPD, with complaints of vaginal dryness, progressively developing a fungating growth in the anterior aspect of right vulva (4x3cm) with no other significant complaints or clinical findings. Biopsy proved it as a Basal Cell Carcinoma, but on metastatic workup we discovered that the patient is a highly rare case of Situs Inversus Totalis (Mirror image like transportation of both thoracoabdominal organs). It's found in one out of every 25000 individuals. Typical findings include dextrocardia, left-sided liver and right-sided stomach and spleen (with polysplenia occasionally). The Patient underwent Right Hemivulvectomy with adequate margins, which was histopathologically proven to be a Basal Cell Carcinoma, (BER EP4 Positive) and is currently on Close follow up from our side with no complications post-operative or otherwise.



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volves surgical modalities such as excision, Electrodesiccation and Curettage (EDC), cryosurgery, and Mohs micrographic surgery. Such methods are typically reserved for localized BCC and offer high 5-year cure rates, generally over 95% [1].

Case presentation

The patient, a P2L2 Post menopausal female, aged 57 years, presented to Gynaecology OPD with symptoms of chronic Vulval pruritus which was primarily thought to be due to estrogen deprived vaginal mucosal atrophy and dryness. A month or so later, she complained of a growth on the right labial folds. On examination, a fungating mass was detected in the anterior aspect of the right vulva, at the junction of labia majora and minora. It was 4 x 3 cm in size, and was not apparently involving or extending to the vaginal mucosa. Urethral involvement was also not noted. Inguinal Lymph Nodes were not palpable on either side. On Per-rectal examination, the rectal mucosa was free. Clinical Stage IB.



Figure 1: T1w contrast MRI (axial sections) showing the lesion at right vulva (arrow).

There was no significant family history, no history of addiction, and no history of arsenic exposure as per patient and patient party. She was a known hypertensive on medications and on salt-restricted diet.

Diagnosis, Therapeutic interventions, and outcomes: -

Biopsy from the fungating growth revealed the tumour to be a Basaloid Neoplasm. (30/12/24) Metastatic workup revealed no mets on CECT Thorax and Whole Abdomen but incidentally we discovered an umbilical hernia, and the rather astonishing fact that the patient is a highly rare case of Situs Inversus Totalis. (11/12/24)

Situs Inversus Totalis is a rare congenital abnormality which is characterized by a mirror image like transposition of both the abdominal and the thoracic organs. It is typically found in one in 25,000 cases [3].

It's an anomaly known since ancient times, but most health care professionals do not have adequate experience regarding this. Even a highly busy surgeon wouldn't expect to encounter this anomaly more than once or twice in a lifetime. Though situs inversus may be detected by a careful physical examination, medical imaging and routine screening programs are necessary to confirm the findings and look for further related pathologies, for example, syndromic Left-Right defects like immotile cilia syndrome, (also known as primary ciliary dyskinesia currently). Traditional and modern medical imaging modalities like ultrasonography, X-rays, CT, MRI scans are usual choices of diagnostic imaging for Situs Inversus Totalis, and are essential before any further invasive therapy. Typical findings include dextrocardia, left-sided liver and right-sided stomach and spleen (rarely poly-

splenia may be detected) [3].

The patient underwent Right Hemi-vulvectomy under spinal anesthesia (on 24/02/25). After proper antiseptic dressing and draping, in Lithotomy position, the entire mass was resected with a 2-3 cm margin all around, and the skin margins were opposed with 2-0 vicryl interrupted sutures. Postoperative phase was uneventful. Patient was catheterised by foley's catheter. Wound healed well before the patient was discharged from the hospital. There was no significant delay in recovery due to any reason.

Post-operative specimen was sent for further histopathological examination. (10/04/2025) It revealed large basaloid lobules with peripheral nuclear palisading with epidermal attachment in a fibromyxoid stroma, and with cleft formation between tumour lobules and stroma. Mitotic activity was High, with Full Thickness Depth of Invasion (> 10 mm). Lymphovascular and perineural invasions were not seen, and the circumferential resection margins were free from tumour. No Lymph Nodes were identified microscopically.

Immunohistochemistry (11/3/25) revealed BER EP4 positivity, and CK20 Negativity, which further confirmed the diagnosis as Basal Cell Carcinoma. Ber EP4, a highly sensitive and specific marker, is an antibody against CD326 (EpCAM) which is expressed in most basal cell carcinomas. CK20 is not associated with Basal Cell Carcinoma typically. It's associated with Merkel Cell Carcinoma, transitional cell carcinoma, etc. Hence, it's presence would have indicated a different diagnosis than Basal Cell Carcinoma [4].

Hence the postoperative specimen confirmed the diagnosis to be Basal Cell Carcinoma of Vulva, with pathological stage T2NxMx.

This was reviewed by the Oncopathology unit, of our institute (on 14/05/25), and was further confirmed to be a case of Basal Cell Carcinoma.

No other forms of cytotoxic therapy, like chemotherapy or radiation therapy, was offered to this patient. She was kept under close follow up.

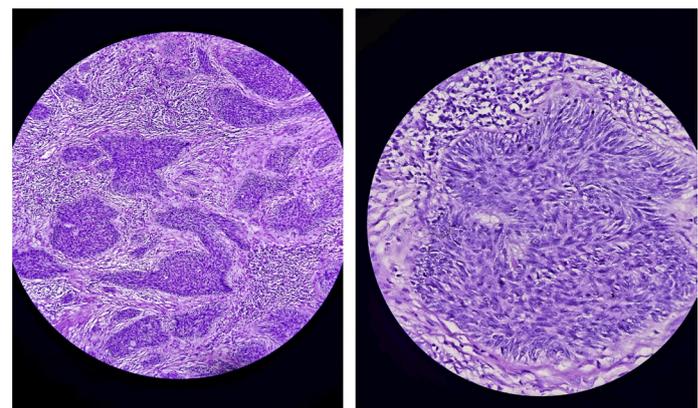


Figure 2: Microscopic view of the lesion showing large basaloid lobules with peripheral nuclear palisading with epidermal attachment in a fibromyxoid stroma, features diagnostic of basal cell carcinoma.

Postoperative CECT Thorax and Whole Abdomen also revealed no abnormalities except Situs Inversus Totalis which was already known by now. (22/05/2025). A Bulky heterogenous thyroid gland with multiple hypodense nodules was also incidentally detected which would be followed up at an Endocrine OPD in the future.

MRI Pelvis Post-operatively done (on 29/04/2025) revealed No Residual/Recurrence in vulva, only Inflammatory postoperative edema. Endometrial polyp of indeterminate etiology, was detected, sized 4.7×1.4 cm. This endometrial polyp was also investigated by Endometrial tissue sampling under Total Intravenous Anesthesia (? on 14/05/25), in lithotomy position on OT table after dilatation of cervical os, and the subsequent sample obtained was very scanty in quantity due to age-related endometrial atrophy.

Histopathological examination of this specimen (on 17/06/2025) revealed scanty Stratified Squamous Epithelium, in a background of acellular eosinophilic material, and few fragmented mucin secreting glands. There was no endometrial gland or stroma, and no evidence of any granuloma or atypia.

The patient is hence kept on close follow-up. She has been advised to undergo clinical gynaecological examination every 4-8 weeks, MRI Pelvis every 6 months, and PAP Smear every 3-6 months as advised by Gynaecology Department.

She's doing well at present, and there are no long-term post-operative complications at present.

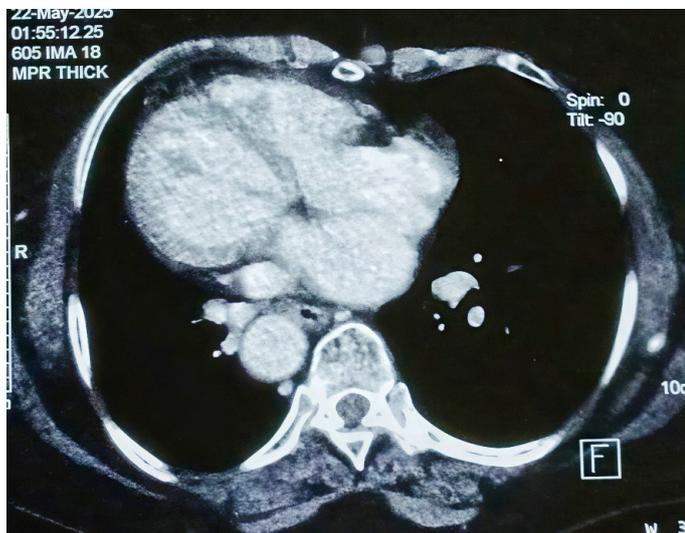


Figure 3: CECT scan of thorax (axial view) showing Cardiac Apex towards right side.

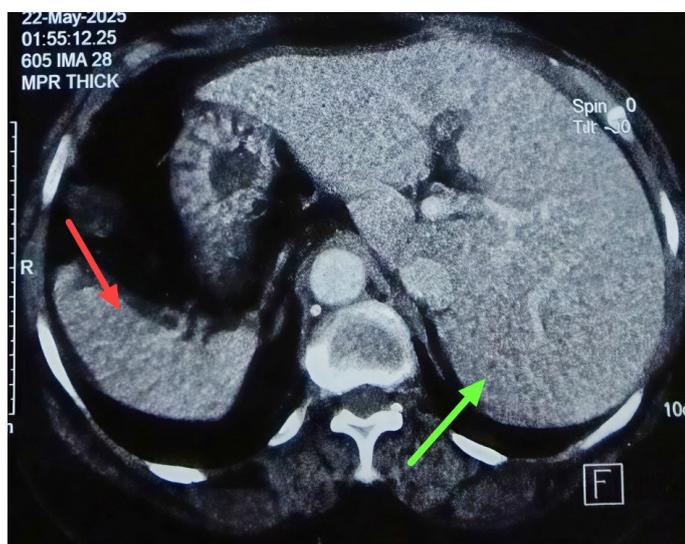


Figure 4: CECT scan of abdomen (axial view) showing Liver on the left side (green arrow) and spleen on the right (red arrow).

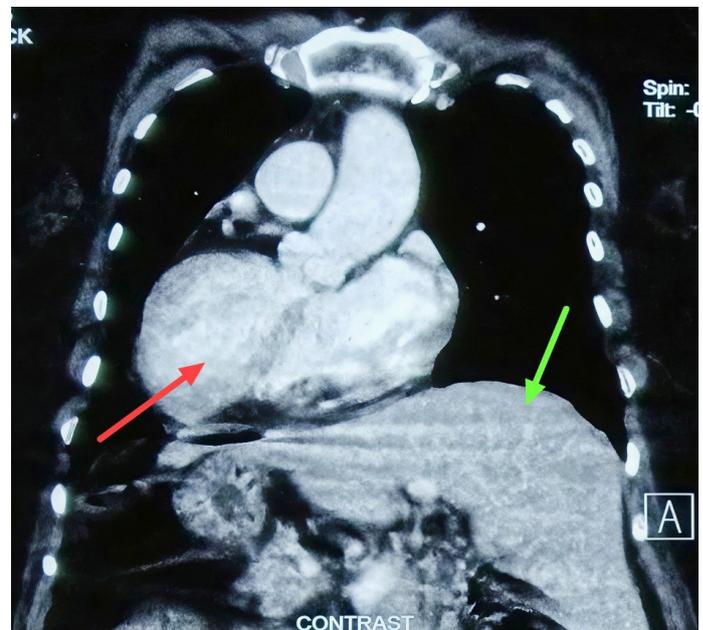


Figure 5: CECT scan Thorax (coronal view) showing left sided Liver (green arrow) and right sided heart (red arrow).

Discussion

De Giorgi et al, in 2005, did a retrospective study and literature review of all BCCs diagnosed between the year 1995 and 2003 in the University of Florence, Italy, Dept. of Human Pathology and Oncology, and reported that 63 out of 3604 cases were located in the vulva, with average age of patients being 70.35 years.

Hence, they concluded that vulval BCCs, though rare, might not be as rare as earlier thought, and should be suspected in non-healing lesions, believed to be inflammatory, and not responding to usual treatments [5].

Cömert Asuman et al, in 2008, reported a case of Vulval BCC in a 78-year-old woman who complained of painful vulval ulceration and bleeding per vaginum, whose biopsy revealed Solid-type BCC, with no lymph nodal metastasis, and was treated with wide local excision and clitoral amputation, and was completely disease free at post-surgical follow up after 1.5 years [6].

Arash Mohammadi Tofigh et al. published a list of three cases of Situs Inversus Totalis in 2023, all incidentally detected during management of other diseases like Cholecystitis (in a left-sided gall-bladder in vicinity of left-sided liver as revealed by Ultrasonography), Perforated peptic ulcer (Erect Posture Straight Xray Abdomen showed Subdiaphragmatic free gas as well as transposition of visceral organs), and sigmoid volvulus (detected on spiral CT scan) [7].

Lakshman VB et al. in 2013 reported a case of Situs Inversus Totalis in an 84-year-old patient who went through most of their life without knowing about this astonishing detail about their own body [8].

Nana Ama Amankwa et al. in 2023 reported an incidentally found case of Situs Inversus Totalis in a 34-year-old diabetic female during work-up and management of Costochondritis [9].

Vulvar BCC is uncommon and accounts for 2 to 4% of all vulvar cancers and <1% of all BCCs. In vulvar area BCC is characterized by poor pigmentation and a clinical appearance often mimicking other dermatological pathologies like eczema or psoriasis. Vulvar BCC exhibits a more aggressive clinical behavior

compared with BCC arising in sun exposed areas [10].

In our case, the did not know that she has such an astonishing finding in herself. The Situs inversus Totalis was diagnosed incidentally during the investigations. The patient finally underwent Right Hemi-vulvectomy under spinal anesthesia (on 24/02/25). After proper antiseptic dressing and draping, in Lithotomy position, the entire mass was resected with a 2-3 cm margin all around, and the skin margins were opposed with 2-0 vicryl interrupted sutures.

Postoperative phase was uneventful. Patient was catheterised by foley's catheter. Wound healed well before the patient was discharged from the hospital. There was no significant delay in recovery due to any reason.

Conclusions

Hence, we can conclude in brief that this was a case of Basal Cell Carcinoma Right Vulva, in a 57-year-old postmenopausal P2L2 hypertensive female who was also incidentally found to be a rare case of Situs Inversus Totalis. She was diagnosed by clinical examination, histopathology from the growth, and underwent Right Hemivulvectomy and the tumour was excised with adequate margins. The Histopathology and Immunohistochemistry reports confirmed this to be a case of Ber EP4 positive Basal Cell Carcinoma of Vulva. An endometrial polyp was also detected on MRI but endometrial biopsy revealed no granuloma, atypia or malignancy. No further cytotoxic therapy was given to this patient and the patient is currently doing well, and has been kept on close follow-up with serial clinical examinations 4-8 weeks, 3-6 monthly PAP Smears and planned for 6 monthly MRI evaluation.

Author declarations

Human subjects

Informed consent for treatment and open access publication was obtained or waived by all participants in this study. No patient particular has been disclosed.

Conflicts of interest

In compliance with the ICMJE uniform disclosure form, all authors declare the following.

Payment/services info

All authors have declared that no financial support was received from any organization for the submitted work.

Financial relationships

All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.

Other relationships

All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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