Review

Basal cell carcinoma of the vulva: a case report and systematic review of the literature

Sruthi Renati¹, MD, Christopher Henderson², MD, MPhil, Ashley Aluko³, MD, and Susan Burgin⁴, MD

¹Department of Dermatology, University of Michigan, Ann Arbor, MI, USA, ²Department of Dermatology, Duke University Hospital, Durham, NC, USA, ³Department of Obstetrics and Gynecology, Beth Israel Deaconess Medical Center, Boston, MA, USA, and ⁴Department of Dermatology, Beth Israel Deaconess Medical Center, Boston, MA, USA

Correspondence

Christopher Henderson, MD, MPHIL Department of Dermatology Duke University Hospital Durham, NC USA

E-mail: christopher.r.henderson@duke.edu

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Introduction

While basal cell carcinomas (BCC) are the most common human malignancy, the vulva is an unusual location, with vulvar BCCs accounting for <1% of all BCCs. Additionally, BCC of the vulva accounts for <5% of all vulvar malignancies. BCC of the vulva most commonly affects elderly women. Presenting symptoms include pruritus, a lump, bleeding, and pain. The proposed risk factors for BCC occurring in non sun-exposed sites such as the vulva include radiation therapy and basal cell nevus syndrome. Treatment options for vulvar BCCs include radical and simple vulvectomy, wide local excision, simple surgical (local) excision, and Mohs micrographic surgery (MMS). We report the case of an 83-year-old woman who presented with a BCC of the vulva. We also review the characteristics of the reported cases of vulvar BCCs in the literature, including treatment approach.

Case Report

An 83-year-old woman with a 40-year history of treated lichen sclerosus presented with a tender growth in the genital area for

Abstract

The vulva is an unusual site for basal cell carcinoma (BCC). Vulvar BCC accounts for <1% of all BCCs and <5% of all vulvar malignancies. We report the case of an 83 year-old woman who presented with a 2-month history of a tender labial growth, with histopathology confirming nodular BCC. We conducted a systematic literature review of the characteristics of reported cases of vulvar BCCs. A comprehensive systematic review of articles indexed for MEDLINE and Embase yielded 96 reports describing 437 patients with 446 BCCs of the vulva. The mean age at presentation was 70 (range 20–100). Most women had no underlying vulvar disease. Approximately 60% of cases were of the nodular subtype. Treatment approach varied widely with over half of cases treated with wide local or local excision. Mohs micrographic surgery (MMS) for vulvar BCC was first reported in 1988 with seven total MMS cases reported. Twenty-three cases of recurrence have been reported; 21 of these cases after local excision but none following MMS. Vulvar BCC is a rarely reported cancer that affects older women predominantly. MMS represents a promising treatment for BCC in this anatomic location.

2 months. Genital examination revealed a 5-mm eroded papule on the right inferolateral labium majus arising on normal skin, and a white, sclerotic appearance of the labia minora and clitoris (Fig. 1). Biopsy of the papule revealed a nodular BCC. The patient underwent MMS. She has no evidence of disease recurrence at 3 years.

Literature Review: Materials and Methods

A comprehensive systematic review of articles indexed for MEDLINE and Embase using the terms basal cell carcinoma or basal cell cancer or basal cell epithelioma or basal cell tumor or ba

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Figure 1 BCC arising from the labium majus. Changes of treated lichen sclerosus also seen

titles and abstracts for relevance. Two reviewers assessed the remaining articles for inclusion, with disagreements going to a third reviewer. Only published case reports or case series in peer-reviewed journals were included in this review, with publication years ranging from 1950 to 2015. Case series that presented case reports in aggregate form were included even if these reports did not specify lesion details beyond location and histopathology. In most case reports, lesion size, specific anatomic site, patient age, treatment, recurrence, associated metastases, and provider specialty were available. These data were noted and tabulated yielding 96 reports in the English literature describing 437 patients with BCCs of the vulva. 1-95 Of the 96 reports, 41 were published in the gynecology or gynecologic oncology literature, 29 in the dermatology literature, and 26 in nongynecology, nondermatology literature (pathology, surgery, oncology, medicine).

Results

The mean age at presentation was 70 years (range 20–100). Of the 437 patients, seven patients had more than one vulvar tumor (two of whom had three primary tumors) resulting in a total of 446 cases of vulvar BCC.^{5,6,32,38,53,54} Risk factors for the seven

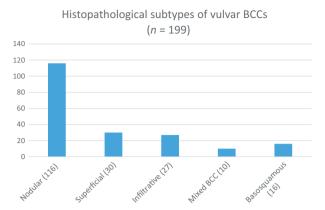


Figure 2 Histopathologic subtypes of vulvar BCCs (n = 199)

patients with multiple BCC of the vulva included four patients with a previous history of multiple BCC on sun-exposed sites, ^{32,38,53,54} two patients with history of radiation to pelvis, ^{5,54} and three patients with Gorlin's syndrome. ^{6,14} Overall, most women did not have underlying vulvar disease; however, in three patients, BCCs arose in contiguity with extramammary Paget disease. ^{7,44,63} Two BCCs arose within an area of lichen sclerosus ^{57,59} while in a third case, a BCC arose adjacent to the area affected by lichen sclerosus. ³⁴ as was seen in our patient.

Anatomic site

The exact site on the vulva was documented in 212 cases. The labium majus was the site of origin in 162 cases, the clitoris in 10, labium minus in eight, perineum in four, vagina/surrounding introitus and posterior fourchette in three each, and one BCC arose from the periurethral mucosa. Twenty-one further cases involved more than one anatomic location due to the larger size of tumor at presentation. Potential risk factors in these patients included underlying extramammary Paget disease in two women, 7,44 five patients with aggressive histology such as infiltrative, morpheaform, and infundibulocystic BCC patterns, 7,56,61,71,91 two patients with a history of previous BCC on sun-exposed sites, 71,76 and seven patients with a documented late presentation, with either lesion ulceration, large size, and/or metastases. 36,60-62,71,91

Histopathologic subtype

Specific histopathologic subtype of the BCC was documented in 199 cases (Fig. 2) and included 116 (58%) cases that were nodular, 30 (15%) superficial, and 27 (14%) infiltrative. There were 10 documented cases displaying more than one BCC histopathologic subtype – six were nodular, infiltrative, and superficial; three were nodular and superficial; and one was nodular and infiltrative and contiguous with a squamous cell carcinoma (SCC). Other reported histopathologic diagnoses were four cases of mixed BCC and SCC, three total cases of BCC contiguous to SCC (one mentioned above), and 10 cases of

Table 1 BCC recurrences

100		O HOLO			Recurrence		
recurrence rate	o N	patient history	Primary BCC* size (cm), extent	Primary treatment	Months to recurrence	Features	Recurrence treatment/outcome
Siegler <i>et</i> <i>al.</i> , 1951	-	Age 49	Size: 3	X-ray therapy, followed by radium applied directly to the lesion	12	Local recurrence	Radical vulvectomy with removal of posterior and lateral vaginal walls, postoperative radiation. Patient died 1 year later of local spread and infaction
Abell <i>et</i>	2	Age 44	Size: 2.5	Local excision (complete)	~156	Local recurrence	or rocal spread and infection Vulvectomy and bilateral inguinal lymph node
ar., 1969 al., 1969	ю	Age 39	I	Local excision	24 then 72	Local recurrence	Local re-excision both times. Patient died of squamous cell CA of lung 11 years after
	4	Age 69	Size: 1.5	Local excision	36	Local recurrence	biopsy of primary boor resion Local re-excision. Disease-free 6 years following original excision
Yamagami <i>et al.</i> , 1974	ß	Age 53	ı	Local excision	13 then 22	Local recurrence *At the time of 3rd recurrence, patient was diagnosed with	conowing original excision. Chemotherapy followed by local radiation. Patient's lungs deteriorated and she died of respiratory failure due to lung metastases
Deppisch	9	Age 56	I	Local excision	9	metastatic BCC Local recurrence	Local re-excision. Disease-free at 5-year
et al., 1970 Zerner et al., 1979 Simonsen et al., 1985	7 8 9–10	Age 87 Age 92 -	Size 3 × 5	Local excision (complete) Local excision (complete) Local excision (complete)	09~	Local recurrence Local recurrence Local recurrence	Ionow-up - - Local surgical extirpation
9.5% (2/21) Copas <i>et al.</i> , 1996	11 2	Age 87 Renal cell CA Age 89 2 BCC	1 1	Local excision Local excision (complete)	60 120 *time to 3rd	Local recurrence (3 × 5 cm) Local recurrence (three lesions, largest	Local re-excision (complete). Patient died 1 month later of MI Local re-excision (complete). Disease-free at 6 months follow-up
Feakins <i>et al.,</i> 1997 8.9% (4/45)	£1 4.	recurrences	Size: >5, nodal metastases	Radical vulvectomy, inguinal lymphadenectomy (complete)	recurrence 23	4 × 7 mm) Local recurrence	Tumor eroded into inguinofemoral blood vessels causing hemorrhage and septicemia. Patient died from BCC
Benedet <i>et al.</i> , 1997 10.7% (3/28)	19 19 19	- Age 80	Size 2 × 1	Wide local excision (incomplete) Wide local excision (incomplete), followed by immediate re-excision (complete)	24 72	Local recurrence *After primary treatment, prior to local recurrence, patient presented with metastatic BCC in supraclavicular node	Local re-excision (complete) Irradiated supraclavicular nodal metastasis (apparently complete resolution). Patient died of renal failure 2 years later, due to pelvic and abdominal disease

		old etc. N			Recurrence		
recurrence	8	patient history	Primary BCC* size (cm), extent	Primary treatment	Months to recurrence	Features	Recurrence treatment/outcome
Piura <i>et al.</i> , 1999	20	Age 62	Size: 2	Wide local excision (complete)	65	Local recurrence	Local excision (complete). Disease-free at 15 months follow-up
	21	Age 72	Size: 1	Excisional biopsy with tumor	16	Local recurrence	Right hemivulvectomy (complete). Disease-
		Breast CA		free margins	*time to 2nd		free at 143 months follow-up
				(required repeat excisional	recurrence		
				biopsy at			
				6 months due to			
				first recurrence)			
Gibson et	22	I	I	Wide excision	84	I	I
<i>al.</i> , 2001 Sakai <i>et</i>	23	Ane 87	Size: 5 × 3.7	l ocal excision	55	Presented with ulcerated	Surgical excision then palliative treatment
<i>al.</i> , 2011				(complete)		indurated left inguinal lesion.	Patient alive and well at 14 months follow-
						mass in left inguinal region	L.
						(recurrence vs. lymph node	
						metastasis); MRI showed	
						metastasis to pubic bone	

BCC, basal cell carcinoma. indicates the qualifier of the time cited to mean the time until the second or third recurrence.

Table 1 Continued

Table 2 Basal cell carcinoma primary treatment preceding local recurrence

Primary treatment	Case number	Local recurrence (n = 23)	Avg. size (cm) if recurrent
Local or wide local excision	261	21	3.0 cm (n = 7)
Bilateral simple vulvectomy	40	0	_
Radical vulvectomy	23	0	_
Hemivulvectomy	9	0	-
Excisional biopsy	8	1	1 cm $(n = 1)$
Radiation therapy	4	1	3 cm (n = 1)
Electrocautery	3	0	_
Mohs micrographic surgery	7	0	_

basosquamous cell carcinoma (BSC). Of the 10 cases of BSC, one case of BSC was infiltrative and metastatic to bone. This patient also had a history of basal cell nevus syndrome.¹⁴

Therapeutic approach

Surgical treatment approaches varied widely and were documented in 353 cases. The most common surgical treatment was wide local excision in 88 (25%) cases and simple surgical (local) excision in 173 (49%). Vulvectomy was also reported with 40 (11%) cases of bilateral simple vulvectomy, 23 (7%) cases of radical vulvectomy of which 17 had concomitant lymph node dissection, and nine (3%) cases of hemi- or partial vulvectomy of which two cases had concomitant lymph node dissection. The indication for lymph node dissection in these 19 patients included seven patients with palpable inquinal lymphadenopathy and three patients whose biopsy revealed BSC histology. 15,53,56 Other reasons for lymph node dissection included three cases with initial diagnosis of SCC or adenocystic carcinoma^{15,55} and one case due to large tumor size and concern for involvement of the clitoral lymphatic network.²³ Indications for lymph node dissection were not specified in the remaining five cases. Eight cases were treated with excisional biopsy alone, four cases were treated with radiation therapy, and three cases were treated with electrocautery.

Seven (2%) total cases of MMS for the treatment of vulvar BCC have been reported. The first reported cases of vulvar BCC treated with MMS were in 1988, when two patients with BCC located on the perineum were successfully treated with MMS.²⁰ An additional five cases of vulvar BCC have since reported treatment with MMS.^{2,65,76} A study by Gibson *et al.* found eight cases of perianal and genital BCC successfully treated with MMS; however, this report does not specify whether these BCC were located on the vulva.¹ Our case documents an additional vulvar BCC successfully treated with MMS.

Local recurrence

Twenty-three cases of local recurrence have been documented in the literature (5.1% of 446 cases) (Table 1). Twenty-one of

these cases occurred following local excision (seven completely excised, seven incompletely excised, six unspecified). The remaining cases recurred after radical vulvectomy and lymphadenectomy, x-ray therapy followed by direct radium application, and excisional biopsy with tumor-free margins. Of the 14 recurrent cases in which age was reported, patients were with an average of 69 years old (range 44–92). The histopathological characteristics of primary BCC lesions that recurred were specified in 10 cases. ^{10,34,61,69,77,94} There was notable variation, including cases with both low ³⁴ and high ^{34,77} mitotic counts. Squamous change was evident in five primary tumors. ^{34,61,69,94}

Recurrent lesions were most often treated with local excision. Four patients died from recurrent disease – one from local tumor invasion leading to hemorrhage and septicemia, another from local spread and infection, a third from renal failure due to pelvic and abdominal disease, and a fourth from respiratory distress secondary to lung metastases. ^{15,34,80,94} Multiple recurrences were not uncommon, occurring in four BCC cases. Of the cases involving a single recurrence, time to recurrence ranged from 6 months to 13 years. Three series of BCC cases revealed a local recurrence rate of 8.9–10.7%. ^{15,34,82} No recurrence after MMS has been reported (Table 2).

Metastatic BCC

Twelve cases in this series of 446 were metastatic (2.7%). Table 3 presents key features of these cases. Of the 11 metastatic cases in which age was reported, patients were an average of 74 years old (range 41–87). BCC was metastatic to inguinal lymph nodes in nine of the 12 cases. Palpable lymphadenopathy on the side of histologically confirmed metastases was appreciated in only three of these cases. There was one reported case of distant nodal metastasis (supraclavicular lymph node)¹⁵ and four cases of hematogenous spread (right thigh skin, pubic bone, femoral head, lung). 61,62,77,94 Metastatic BCC lesions tended to be large and deeply infiltrative. The depth of involvement was reported in four cases, 34,62,71,86 with the deepest penetration measuring 6 cm. 62 In seven cases, the primary BCC invaded subcutaneous tissue or fat, and in two cases, the tumor extended to the pubic bone.

Perrone *et al.* compared their single case of metastatic BCC to 10 cases of nonmetastatic BCC. They found that the metastatic lesion was distinguished by extensive local involvement of the urethra and vagina, clinical history of vaginal bleeding secondary to tumor infiltration, deep penetration into subcutaneous fat, and a morphea-like growth pattern.⁷¹ Two cases of metastatic BCC were notable for delayed presentation – one patient noticed "something growing" on her vulva for approximately 5 years before seeking treatment, and another patient presented with a 14-year history of a lump on the mons pubis.^{86,91}

Most patients with metastatic disease were treated with radical vulvectomy and bilateral inguinal lymphadenectomy. The patient with femoral head metastasis did not undergo inguinal node dissection because of known hematogenous spread and a

Table 3 BCC metastases

			Primary		Palpable		
Article	8	Age	Size (cm)/extent	Histologic features	nodes?	Site of metastasis	Treatment/outcome
Yamagami et al., 1974	-	53		Mixed solid and adenoid type	O _N	Lung	Chemotherapy followed by local radiation. Patient's lungs deteriorated, and she died of respiratory failure due to lung metastases.
Jimenez et al., 1975	0	4	5 × 5 × 2.5 Infiltrated deep and lateral subculaneous fissues	Infiltrative, small foci of squamoid cells	Yes (left)	Inguinal lymph nodes (left, both superficial and deep)	Radical vulvectomy, bilateral lymph node dissection
Sworn et al., 1979 Perrone	ω 4	71	8 × 4.5 Extended 0.5 cm into adjacent fat "Large"	Solid and adenoid, many mitotic figures Adenoid, morphea-like,	0	Lymph nodes in subcutaneous fat (5 total) Inguinal lymph nodes	Lesion completely excised Radical vulvectomy, bilateral inguinal
et al., 1987			Inflitrated rateful wall of vagina, urethra, subcutaneous fat (thickness = 1.2)	pilat-type keraunizauon		(s rignt, 4 iert, 1 Croquet)	ymphadenectorny, postoperative radiation to pelvis and inguinal areas. Patient alive and well at 1 year and 2 months follow-up
Hoffman <i>et al.</i> , 1988	Ŋ	74	6×4 , 1 (two lesions) Infiltrated subcutaneous fat, extended to perineal body and approaching anal skin	Solid, adenoid, areas of keratinization, infrequent mitoses	ON.	Inguinal lymph nodes (2/12 left, superficial)	Radical vulvectomy, bilateral inguinal lymphadenectomy
Winklemann et al., 1990	Ø	7	6×15 Ulceration and necrosis to the depth of the symphysis pubis	Morpheic	Yes (bilateral)	Inguinal lymph node (1/6 right, 0/8 left)	Preoperative radiation to vulva, symphysis pubis, and perineal body (complete resolution), radical vulvectomy and proetectomy with removal of the anus with transverse colostomy, inguinal lymphadenectomy
Gleeson <i>et al.</i> , 1994	_	98	2.5×4 Shallow ulcer	Nodular, adenoid, few mitotic figures, hyperkeratotic squamous epithelium	Yes (left)	Inguinal lymph node (right)	Radical vulvectomy, bilateral inguinofemoral lymphadenectomy. Patient disease free at 1-year followup
Mizushima et al., 1995	ω	62	10 × 8 Infiltrated deep and lateral subcutaneous tissues	Adenoid, squamoid tumor cells, mitotic figures observed, vascular invasions, tumor embolizations	Yes (bilateral)	Inguinal lymph nodes (10/26) Skin (right thigh)	The skin metastasis on the right thigh was slightly reduced by radiation before patient was lost to follow-up. (Patient had undergone radical excision of vulvar BCC and bilateral lymph node dissection 6 months prior). Patient's breast CA recurred, and she died the following year

Table 3 Continued	tinue	p					
			Primary		Palpable		
Article	8	Age	Size (cm)/extent	Histologic features	nodes?	Site of metastasis	Treatment/outcome
Feakins et al., 1997	σ	1	Infiltrated subcutaneous fat (depth = 1.12)	Infiltrative, compact lobular, low mitotic count, focal benign squamous change	1	Inguinal lymph node	Radical vulvectomy and inguinal lymphadenectomy (complete). Local recurrence at 23 months, tumor eroded into inguinofemoral blood vessels causing hemorrhage and septicemia. Patient died from BCC
Benedet et al., 1997	10	80	2 × 1 Infiltrated subcutaneous tissue, perineural invasion	1	1	Supraclavicular Iymph node	Treated supraclavicular nodal met with irradiation, with apparently complete resolution. Patient developed a local recurrence 2 years after this. Patient died of renal failure 2 years later (thought 2/2 pelvic and abdominal disease)
Mulayim et al., 2002	Ξ	98	13 × 8 Infiltrated to pubic bone, (max thickness = 6), involved perineal body and the lower vagina.	Solid, adenoid, mitoses common	O _Z	Femoral head (left)	Radical resection of the vulva bilaterally, mons anteriorly, perineum posteriorly. Radiation to the femoral head metastasis. At 6 months, she progressed with multiple bony (right femur, skull) and intraperitoneal metastases and died with disease
Sakai <i>et</i> <i>al.</i> , 2011	12	87	5 × 3.7	Infiltrative, high mitotic figures	ı	Inguinal lymph node (left, vs. local recurrence) Pubic bone	Surgical excision then palliative treatment. Patient was alive and well at 14 months follow-up

BCC, basal cell carcinoma.

primary goal of palliation.⁶² Radiation therapy was performed on five patients. Winkelmann *et al.* presented a case of a large and locally invasive tumor that rendered the patient an inoperable candidate. This patient was treated with preoperative radiation that resulted in complete resolution of the tumor prior to radical vulvectomy and proctectomy with inguinal lymphadenectomy.⁹¹ Four patients with metastatic BCC died from the disease. ^{15,34,62,94}

Discussion

Although BCC is a common malignancy, the vulva is an unusual anatomic location for BCC and accounts for <1% of all BCC. We reviewed over 400 published cases of vulvar BCC in the literature and found that vulvar BCCs most often affect postmenopausal females over the age of 70. The most common location is the labium majus. Vulvar BCC is rarely associated with underlying vulvar disease such as extramammary Paget disease or lichen sclerosus.

Vulvar BCC is histopathologically similar to BCC occurring in other anatomic locations. ¹⁹ Certain histologic types such as basosquamous carcinoma, adenocystic, and infiltrative types tend to be more aggressive and may explain cases of recurrence. ⁹⁶ Lymph node involvement and hematogenous metastasis to distant sites are rare but can occur. Treatment approaches varied widely in reported literature. Most cases were treated with wide local excision. A minority of patients underwent simple or radical vulvectomy. Seven cases treated with MMS have been well documented in the literature.

In our review of 437 patients with vulvar BCC, 23 (5.1%) recurred and 12 (2.7%) metastasized. Studies estimate a recurrence rate of 4.2% (follow-up <5 years) and 8.7% (follow-up >5 years) for all BCCs,97 and the reported incidence of metastatic BCC for all BCCs ranges from 0.0028 to 0.55%. 5,65 These vulvar recurrence and metastatic rates are likely underestimated, as patient follow-up was limited in most case reports. Because of the low incidence of vulvar BCC, it remains difficult to define factors associated with increased risk of recurrent or metastatic disease. Several reported cases of local recurrence followed incomplete excision, though the presence or absence of positive surgical margins does not appear predictive of recurrence. High recurrence rates have been reported in the literature with local recurrence after complete local excision in about 10-20% of cases, possibly due to inadequate surgical margins. 69 Advanced vulvar BCC often involves metastasis to inguinal lymph nodes, though distant nodal metastasis and hematogenous spread are also possible. Based on our review, potential risk factors for metastatic disease include large size (including from late presentation), depth of involvement, and aggressive histologic features. Though uncommon, both recurrent and metastatic BCC can be fatal.

MMS has been associated with improved cosmetic and functional outcomes.^{5,65} MMS may be the best chance of cure,

especially with increased tumor size, aggressive histology, and when maximum uninvolved tissue preservation is desired.²⁰ The use of MMS for treatment of vulvar BCC may lead to improved rates of local recurrence. MMS represents a promising treatment for BCC in this anatomic location; however, further experience with MMS for vulvar BCC is needed to confirm this observation.

Questions (answers found after references)

- 1 In which decade do most patients experience vulvar BCC?
 - (a) Fifth
 - (b) Sixth
 - (c) Seventh
 - (d) Eighth
- 2 Which of the following is not a potential risk factors for BCCs developing in non sun-exposed areas?
 - (a) Radiation therapy
 - (b) Basal cell nevus syndrome
 - (c) Recurrent infection
 - (d) Lichen sclerosus
- **3** The majority of the articles reviewed in this manuscript come from which literature sources?
 - (a) Gynecology
 - (b) Gynecological oncology
 - (c) Dermatology
 - (d) Surgical oncology
- 4 What is the most common site for vulvar BCCs?
 - (a) Introitus
 - (b) Posterior fourchette
 - (c) Periurethral
 - (d) Labium majus
- 5 What is the most common histological subtype of vulvar BCC?
 - (a) Superficial
 - (b) Nodular
 - (c) Infiltrative
 - (d) Basosquamous
- 6 What is the recurrence rate of vulvar BCCs?
 - (a) 1.4%
 - (b) 3.5%
 - (c) 5.1%
 - (d) 8.5%
- 7 After which treatment modality did most vulvar BCC recurrences occur?
 - (a) Mohs micrographic surgery
 - (b) Electrodessication and curettage
 - (c) Local excision
- 8 What percentage of vulvar BCCs result in metastasis?
 - (a) 1.5%
 - (b) 2.7%
 - (c) 5.1%
 - (d) 7%

- 9 What is the most common location for vulvar BCC metastasis?
 - (a) Inguinal lymph node
 - (b) Retroperitoneal lymph node
 - (c) Distant site
 - (d) Uterus
- 10 In the author's opinion, what is the preferred method for vulvar BCC treatment?
 - (a) Local excision
 - (b) Electrodessication & curettage
 - (c) Mohs micrographic surgery

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Answers to questions

1, c; 2, c; 3, a, b, & c; 4, d; 5, b; 6, c; 7, c; 8, b; 9, a; 10, c