

# Benign phyllodes tumor of the vulva: a case report and literature review

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## Abstract

Phyllodes tumor is an uncommon breast lesion with characteristic histologic appearance when examined by hematoxylin and eosin staining: leaf-like fronds projecting into cystic spaces on low-power microscopy, and biphasic (epithelial and stromal) components on high-power microscopy. We report a rare primary case of this tumor arising within the vulva. A 34-year old African American female presented with a 3 cm slow-growing vulvar mass initially thought to be an inclusion cyst. The lesion was excised and histologic examination demonstrated this lesion to be a rare case of benign phyllodes tumor with morphologic features similar to those arising from breast tissue. Patient received no further treatment and did not exhibit any recurrence or metastasis. Nearly two years after excision, the patient died due to an unrelated medical cause. This rare tumor should be considered in the differential diagnosis for women presenting with a slow-growing vulvar mass.

## Introduction

In 1872, Hartung first reported benign mammary glands in the vulva in a 30-year-old woman.<sup>1,2</sup> Since then, a variety of lesions similar to breast lesions have been described in the vulva, including sclerosing adenosis, intraductal papilloma, fibroadenoma, adenocarcinoma, lactating adenoma, fibrocystic change, hidrocystoma, hidradenoma papilliferum, and phyllodes tumor.<sup>2-8</sup> In addition to the vulva, mammary lesions have been found in other extra-mammary sites such as the axilla and anus.<sup>9,10</sup> Both benign and malignant lesions have been described with varying prognoses.<sup>1</sup>

## Case Report

### Presentation

A 34-year-old African American woman with

a medical history of sickle cell anemia and heterozygous factor V Leiden mutation was referred due to an enlarging vulvar mass causing discomfort. There was no history of prior malignancy or breast disease. Physical exam showed an approximately 3-cm mass located in the lower right labia minora, which initially was thought to be an inclusion cyst. No radiological studies were obtained. Simple excision of the lesion was performed and submitted for pathologic examination. The patient tolerated the procedure without any complications.

### Pathologic findings

Gross examination revealed a 1.5×1.5×0.7 cm polypoid fragment that on serial sectioning disclosed a cystic cavity filled with multiple papillary excrescences. Microscopic examination showed a well circumscribed lesion composed of broad, leaf-like papillary fronds projecting into a cystic space (Figure 1A). Covering the fronds was a cuboidal epithelium with a subjacent layer of presumably myoepithelial cells (Figure 1B). Within the invaginations of the fronds, the epithelium demonstrated apocrine metaplasia and rare areas of secretory change. The stroma was paucicellular, composed of sparse cytologically bland cells with ovoid nuclei and spindled or stellate cytoplasm set within a densely collagenized extracellular matrix. There was no cytologic atypia, necrosis, or hemorrhage. No mitotic figures were seen. No immunohistochemical stains were performed. Based on the histologic features, the lesion was diagnosed as a benign phyllodes tumor arising in the vulva according to the WHO criteria for grading phyllodes breast lesion.<sup>11</sup>

### Follow-up information

Patient did not exhibit any recurrence or metastasis of the phyllodes tumor. Nearly two years later she died due to an episode of sickle cell crisis. Autopsy examination was performed and did not reveal any evidence of local recurrence (based on external examination) or metastasis of the phyllodes tumor.

## Discussion and Conclusions

First described in the breast by Johannes Muller in 1838, phyllodes tumors comprise less than 1% of primary breast tumors.<sup>2,4,12</sup> Containing both epithelial and cellular stromal components, phyllodes tumors are known for their characteristic leaf-like architecture.<sup>13</sup> Breast phyllodes tumors may be classified as benign, borderline or malignant based on several histological features using various classification systems. WHO classification employs the following histologic features: amount of

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stromal cellularity, degree of cellular pleomorphism, number of mitoses (malignant >10/10 HPF), infiltration of margins, and presence or absence of heterologous differentiation.<sup>11</sup> While commonly found in the breast, phyllodes tumor of the vulva have been reported rarely, with only 14 cases found upon review of the current literature.<sup>2-4,7,10,12-15</sup>

Phyllodes tumor of the vulva usually presents as a unilateral, painless mass, the most common locations being labia majora, labia minora, and interlabial cleft.<sup>2,13</sup> Microscopically, the vulvar tumors contain the same classic biphasic epithelial and stromal phenotype of the breast tumor.<sup>2</sup> While recurrence is rare, there have been 3 out of 14 cases where recurrence was reported 8 months after initial excision in one case and 2 years later in the other two cases.<sup>3,5,13</sup> After re-excision, no further recurrence occurred. Because phyl-

lodes tumor of the vulva is a rare tumor, the behavior and prognosis of this lesion is difficult to predict. Currently, there is no specific classification or grading system. Using criteria for breast phyllodes, three cases were graded as borderline (or low-grade malignant)<sup>5,12</sup> and one malignant,<sup>15</sup> with all other cases being classified as benign. No cases of metastasis have been reported. We report here a case of phyllodes tumor of the vulva that was classified as benign based on WHO grading system for breast phyllodes.<sup>11</sup> Available clinical follow-up

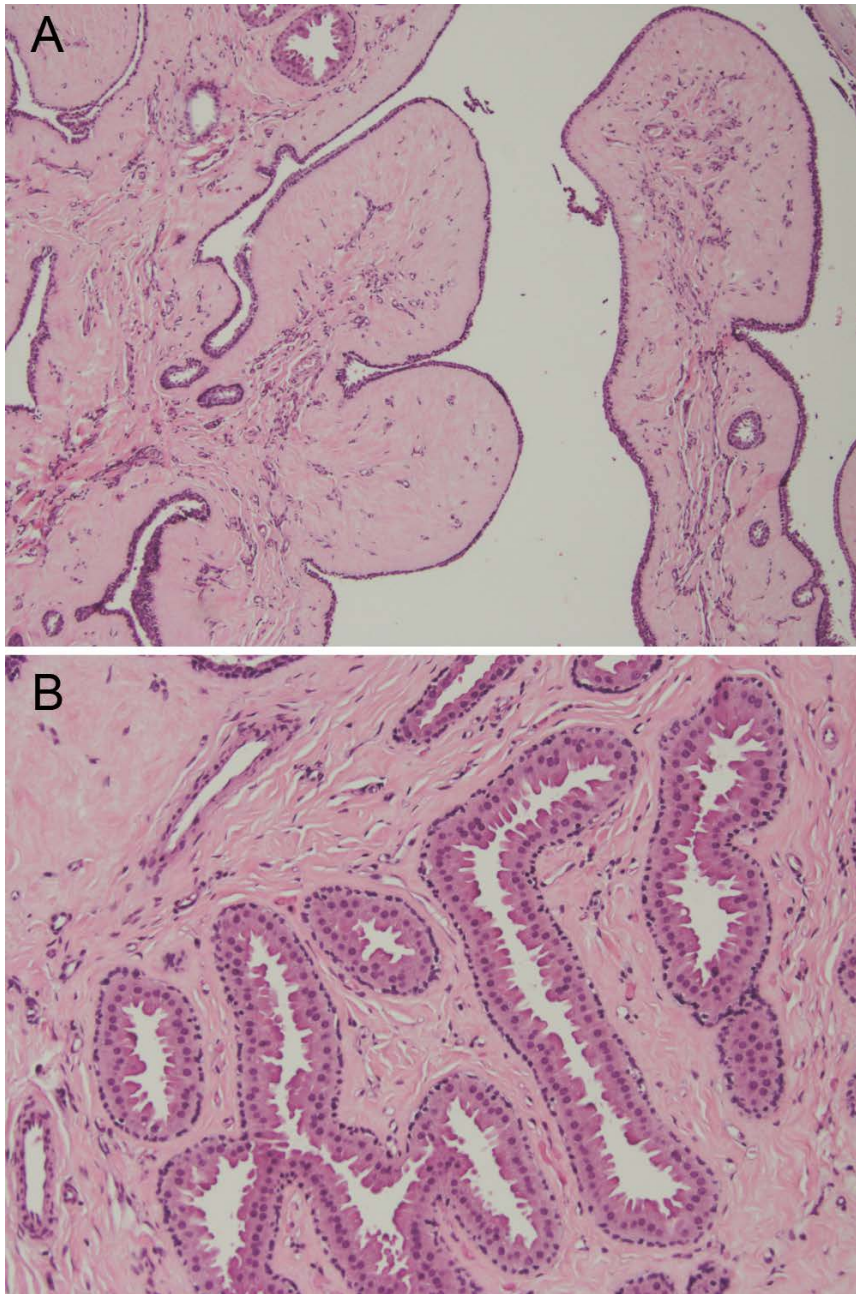
and autopsy findings showed no evidence of recurrence and were thus consistent with the assigned grade of benign.

Differential diagnosis for phyllodes tumor of the vulva include other biphasic tumors, including fibroadenoma, papillary hidradenoma and chondroid syringoma. Distinction between phyllodes tumor and benign or borderline phyllodes tumor can be difficult due to overlapping morphological features. The characteristic leaf-like architecture seen in phyllodes tumor may be seen rarely in fibroadeno-

ma. The stroma in fibroadenoma is generally less cellular without cytologic atypia and more homogeneous compared to phyllodes.<sup>5,6</sup> Microscopically, papillary hidradenoma tumors have a complex papillary growth pattern similar to phyllodes tumors; however, these growths are lined by a double layer of epithelial cells and focal apocrine snouts.<sup>4</sup> Papillary hidradenomas also lack a prominent stromal component.<sup>4</sup> Chondroid syringomas have a fibromyxoid stroma with cartilaginous islands. In contrast to phyllodes tumors, chondroid syringomas may have areas of sebaceous and squamous differentiation.<sup>4</sup> A rare case of Mullerian adenosarcoma of the cervix secondarily involving the vulva has been reported that may mimic a phyllodes tumor due to a similar leaf-like architecture and biphasic pattern. Mullerian adenosarcoma also display periglandular condensation that distinguishes them from phyllodes tumor.<sup>16</sup>

There is controversy regarding the origin of proliferative processes mimicking breast lesions in the anogenital region. In the past, the generally accepted theory regarding these lesions was that they arose from ectopic mammary glands as remnants of the mammary ridges.<sup>1</sup> First studied in pigs, the mammary ridges are areas of mammary glands that develop from the axilla to the inguinal regions of the medial thigh. During embryologic development, the glands regress from the inguinal regions upwards, resulting in a single pair of breasts in the pectoral region. Supernumerary mammary tissue was thought to occur when regions of the original mammary ridges failed to regress.<sup>1</sup> van der Putte challenged this theory of supernumerary mammary tissue, arguing that mammary ridges in human embryos do not involve the anogenital region and therefore cannot be attributed to the origin of breast mimicking lesions.<sup>1</sup> Instead, in 1991 van der Putte described mammary-like glands (MLG) found in the anogenital region, mimicking mammary glands but still distinguishable from eccrine and apocrine glands.<sup>17</sup> van der Putte also suggested that MLG are a normal part of the vulvar epithelium and are the source for anogenital lesions mimicking proliferative processes of the breast.<sup>1,17</sup>

Mammary-like glands vary in morphology, from simple tubular structures to branching glands to well-formed lobuli.<sup>17</sup> The secretory epithelium consists of a luminal layer of tall columnar cells with apical *snouts* in the ductuli and cuboidal cells in the acini, resting on a peripheral layer of flat basal myoepithelium.<sup>1,15</sup> The glands often exhibit ER and PR expression and are concentrated in the sulcus between the labia minora and labia majora. van der Putte refuted that these glands were from ectopic mammary tissue due to the following considerations: i) while similar to mammary glands, MLG have a more simple configuration



**Figure 1.** Hematoxylin & Eosin images of phyllodes tumor of the vulva: A) Low-power view (10×) showing characteristic leaf-like projections into cystic spaces; B) High-power view (20×) showing biphasic pattern consisting of epithelial and stromal components, and apocrine metaplasia.

and a different acinar epithelium, ii) the number of MLG is much higher than would be expected from remnants of the mammary ridges, and the rows of glands suggest a relationship to cloaca-derived tissue rather than mammary ridges, iii) MLG show a direct relationship to eccrine glands of the anogenital region, iv) even if mammary ridges run to the inguinal folds, they do not involve the anogenital region.<sup>1</sup> van der Putte concluded that these lesions, previously thought to be associated and derived from supernumerary mammary glands, are actually derived from MLG.<sup>1</sup> Recent literature concludes that the concept of vulvar mammary tissue as a derivative of mammary ridges is no longer tenable and instead, MLG are a normal constituent of the anogenital region.<sup>1,15</sup> Some argue whether *phyllodes tumor of the vulva* is justified simply because of its morphological similarity to mammary phyllodes tumor, when recent evidence is more favorable for MLG rather than ectopic breast tissue, suggesting *phyllodes-like tumor of the anogenital mammary-like glands* reflect a more appropriate terminology.<sup>2</sup>

## References

1. van der Putte SC. Mammary-like glands of the vulva and their disorders. *Int J Gynecol Pathol* 1994;13:150-60.
2. Mannan AA, Kahvic M, Aziz AH. Phyllodes tumor of the vulva: report of a rare case and review of the literature. *Am J Dermatopathol* 2010;32:384-6.
3. Tbakhi A, Cowan DF, Kumar D, Kyle D. Recurring phyllodes tumor in aberrant breast tissue of the vulva. *Am J Surg Pathol* 1993;17:946-50.
4. Chulia MT, Paya A, Niveiro M, et al. Phyllodes tumor in ectopic breast tissue of the vulva. *Int J Surg Pathol* 2001;9:81-3.
5. Kazakov DV, Spagnolo DV, Stewart CJ, et al. Fibroadenoma and phyllodes tumors of anogenital mammary-like glands: a series of 13 neoplasms in 12 cases, including mammary-type juvenile fibroadenoma, fibroadenoma with lactation changes, and neurofibromatosis-associated pseudoangiomatous stromal hyperplasia with multinucleated giant cells. *Am J Surg Pathol* 2010;34:95-103.
6. Kazakov DV, Spagnolo DV, Kacerovska D, Michal M. Lesions of anogenital mammary-like glands: an update. *Adv Anat Pathol* 2011;18:1-28.
7. Mariappan MR, Lagera JE, Fadare O, Sibley RK. A 69-year-old woman with a vulvar lesion. Phyllodes tumor of the vulva. *Arch Pathol Lab Med* 2006;130:e11-2.
8. Assylbekova B, Yan P, Yang M. Phyllodes tumour of vulva with prominent pseudoangiomatous stromal hyperplasia features: a case report and review of literature. *J Obstet Gynaecol* 2015:1-2.
9. Oshida K, Miyauchi M, Yamamoto N, et al. Phyllodes tumor arising in ectopic breast tissue of the axilla. *Breast Cancer* 2003;10:82-4.
10. Ozbudak IH, Akkaya H, Akkaya B, et al. Phyllodes tumor of the vulva: report of two cases. *Turk Patoloji Derg* 2013;29:73-6.
11. Bellocq JP, Margo G. Fibroepithelial tumours. In: Tavassoli FA, Devilee P, eds. *Pathology and genetics of tumours of the breast and female genital organs World Health Organization classification of tumours*. Lyon: IARC; 2003. pp 100-103.
12. Giger OT, Lacoste E, Honegger C, et al. Expression of the breast differentiation antigen NY-BR-1 in a phyllodes tumor of the vulva. *Virchows Arch* 2007;450:471-4.
13. Heffernan TP, Sarode VR, Hoffman B, Lea J. Recurrent phyllodes tumor of the vulva: a case report with review of diagnostic criteria and differential diagnosis. *Int J Gynecol Pathol* 2010;29:294-7.
14. Tresserra F, Grases PJ, Izquierdo M, et al. Fibroadenoma phyllodes arising in vulvar supernumerary breast tissue: report of two cases. *Int J Gynecol Pathol* 1998;17:171-3.
15. Fu L, Lau S, Roy I, Ferenczy A. Phyllodes tumor with malignant stromal morphology of the vulva: a case report and review of the literature. *Int J Gynecol Pathol* 2011;30:198-202.
16. Garcia-Rostan y PeGM, Troyas RG, Bercero EA. Mullerian adenosarcoma of the cervix: differential diagnosis, histogenesis and review of the literature. *Pathol Int* 1995;45:890-4.
17. van der Putte SC. Anogenital sweat glands. Histology and pathology of a gland that may mimic mammary glands. *Am J Dermatopathol* 1991;13:557-67.