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The utility of high-frequency ultrasonography in preoperative assessment of vulvar Paget's disease – a case report

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Abstract

Paget's disease of the vulva is a rare skin cancer accounting for less than 1% of all vulvar neoplasias. Surgery is the first treatment of choice. Unfortunately, it is insufficient in many cases, which require further treatment, and the risk of recurrence is high. We present a case of an 85-year-old woman who underwent surgery due to Paget's disease of the vulva. A preoperative skin imaging using a high-frequency 48 MHz mechanical probe was used to assess the lesion margins. Typical high-frequency ultrasonographic features of vulvar Paget's disease were identified and analyzed. This is the first report of high-frequency ultrasonography imaging of vulvar Paget's disease.

Introduction

Paget's disease of the vulva (PDV) is a rare superficial skin cancer accounting for less than 1% of all vulvar neoplasias⁽¹⁾. Extramammary manifestation occurs in the apocrine glands and is a form of intraepidermal adenocarcinoma^(2,3). The most commonly affected regions are the vulva, perineum, perianal region, scrotum, penis, or pubic area⁽⁴⁾. Although surgery is the most effective treatment, a complete excision is challenging due to PDV intraepithelial reticular spread⁽²⁾. According to recent data, the rate of involved margins after primary surgery is up to 72%⁽⁵⁾. High-frequency ultrasonography (HFUS) is a technological advance that has been used for over 30 years in dermatology⁽⁶⁻⁸⁾. Although the value of HFUS as a method for differential diagnosis of skin cancer is still debatable, it gives a clear picture of the size and depth of the tumor, playing an important role in preoperative evaluation⁽⁹⁾. Therefore, we would like to present the utility of HFUS in PDV.

Case report

An eighty-five-year-old patient was referred to our unit with a diagnosis of PDV after biopsy. The main symptoms were discomfort and itching for over 3 months. The patient had arterial hypertension and glaucoma. A few years ago she suffered from encephalitis complicated with right facial paralysis. Physical examination revealed acetowhite areas in the left labia minora and majora with spread to left thigh (Fig. 1). The patient was qualified for partial left sided vulvectomy. Before the surgical procedure, we used DermaView high-frequency mechanical probe scanner (Dramiński S.A., Poland) to evaluate the PDV margins. The characteristic ultrasonographic features of malignant lesion were hypoechoic lesion and altered normal HFUS image of the vulva⁽⁷⁾ as compared to adjacent tissue, that is, the dermis with hypoechoic subepidermal band (Fig. 2, Fig. 3, Fig. 4). The lesion was within the dermis with blurred irregular outer margins. Additionally, we noticed



Fig. 1. Paget's disease of the vulva on the left labia majora and thigh erythematous or eczematous, focally eroded or crusted plaques (arrows)

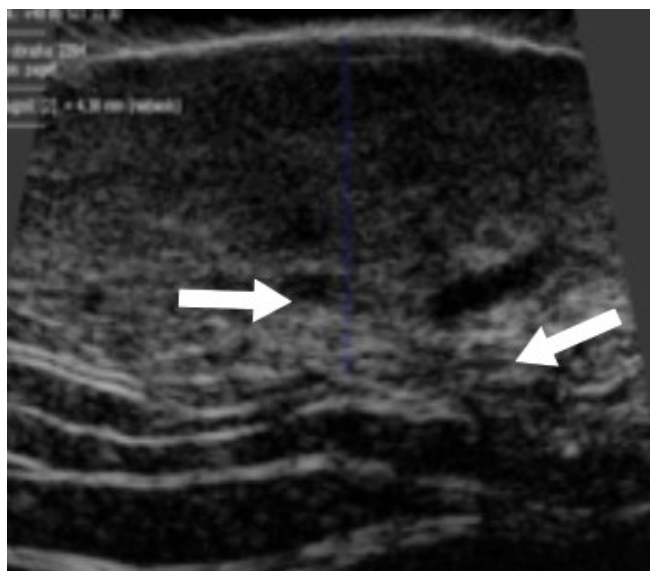


Fig. 2. Hypoechoic lesion with edema and enlarged blood vessels (arrow). The margin is blurred. The disease extends to the subcutaneous tissue (long arrow)

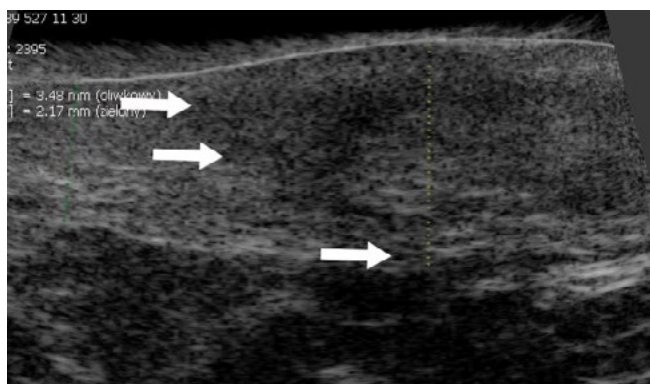


Fig. 3. Paget's disease of the vulva – blurred margin (long arrows) and reaches the subcutaneous tissue

edematous and enlarged blood vessels (Fig. 2, Fig. 4). We also measured skin thickness at the PDV site, margin and healthy skin, which were 4.38 mm, 3.48 mm and 2.17 mm, respectively. The margin was usually irregular, but we found no deeper invasion into the subcutaneous tissue. Compression with the ultrasound probe did not change the shape of the tissue. Although we could assess the margin of PDV, it was not possible to excise the entire lesion for technical reasons. Immunohistochemistry was positive for cytokeratin 7 in neoplastic cells (Fig. 5). The margin was involved, confirming incomplete excision.

Discussion

From all cases reported in the literature, this is the first one with a preoperative assessment of PDV using HFUS. Since it is a rare cutaneous intraepithelial adenocarcinoma that originates from the apocrine glands, the depth of invasion is the most important prognostic factor. Tumors with invasion of >1 mm spreading outside the skin or into lymph nodes or other tissues are associated with poorer prognosis⁽¹⁰⁾. The optimal management of PDV remains unclear. Surgical excision is usually the primary therapy⁽¹¹⁾.

As in our case, surgical excision often results in positive margins due to anatomical limitations. In addition, the disease is often multifocal and many patients require multiple excisions resulting in significant morbidity⁽¹²⁾. Currently, researchers have gained the possibility to examine skin with novel high resolution imaging technique known as HFUS. The possibility of real-time imaging and measurement, mobility and the lack of contraindications make it a helpful tool in skin diagnosis. In dermatological oncology, HFUS is used to assess the depth of invasion in melanomas and basal cell carcinomas (BCC)⁽¹³⁾. Similarly to PDV, preoperative determination of surgical margins determines future management and patient's prognosis⁽¹²⁾. Polańska *et al.* reported that melanomas and BCC have a similar presentation in the form of hypoechoic circular or oval structures surrounded by hyperechoic areas⁽¹³⁾. In the case of PDV, the high-frequency image is also hypoechoic with irregular shape. Additional features included a sub-epidermal low-echogenicity band and blurred outer edges with increased blood vessel diameter and edema. In our opinion, these features could be used for better preoperative assessment and planning. In our case, HFUS imaging indicated that the margin would be PDV positive, which was later confirmed by histology. Hopefully, in cases of PDV with more favorable location or smaller size, the HFUS could improve surgical outcome in terms of negative margins. Unfortunately, in most PDV cases reported in the literature, significant delay in the diagnosis leads to quite extensive vulvar lesions and more unfavorable outcome⁽¹⁴⁾.

Conclusions

HFUS may serve as a useful tool in the diagnosis of Paget's disease of the vulva. We were first to present

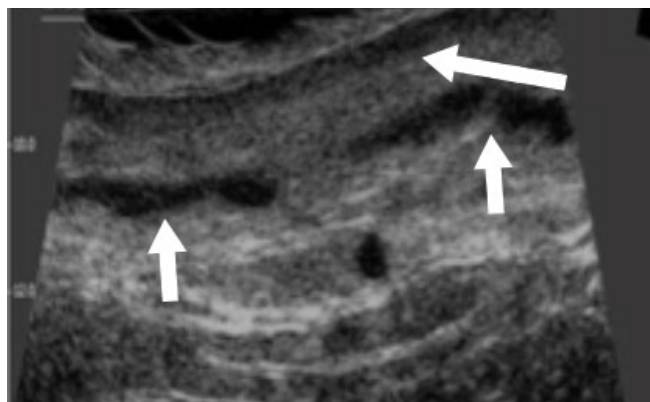


Fig. 4. Subepidermal low-echogenicity band (long arrow) with enlarged blood vessels (thick arrows)

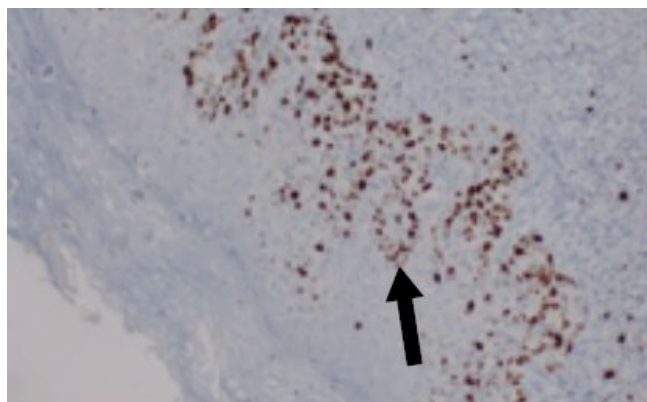


Fig. 5. Immunohistochemistry with positive cytokeratine 7 in neoplastic cells (arrow)

the characteristic ultrasonographic features of PDV. It seems that the routine use of HFUS in preoperative assessment and planning is possible due to its mobility and general acceptance with no contraindications. Further research in this area is recommended to confirm our report.

Conflict of interest

Authors do not report any financial or personal connections with other persons or organizations, which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

References

1. Barmon D, Katakai AC, Imchen L, Sharma JD: Extra mammary Paget's disease of the vulva. *J Midlife Health* 2012; 3: 100–102.
2. Dogan A, Hilal Z, Krentel H, Cetin C, Hefler LA, Grimm C *et al.*: Paget's disease of the vulva treated with imiquimod: case report and systematic review of the literature. *Gynecol Obstet Invest* 2017; 82: 1–7.
3. Gonçalves Amorim A, Batista Fraga Mendes B, Neves Ferreira R, Chambô Filho A: Paget disease of the vulva: diagnosis by immunohistochemistry. *Case Rep Dermatol Med* 2015; 2015: 162483.
4. Kanitakis J: Mammary and extramammary Paget's disease. *J Eur Acad Dermatol Venereol* 2007; 21: 581–590.
5. Roh HJ, Kim DY, Kim JH, Kim YM, Kim YT, Nam JH: Paget's disease of the vulva: evaluation of recurrence relative to symptom duration, volumetric excision of lesion, and surgical margin status. *Acta Obstet Gynecol Scand* 2010; 89: 962–965.
6. Lassau N, Spatz A, Avril MF, Tardivon A, Margulis A, Mamelle G *et al.*: Value of high-frequency US for preoperative assessment of skin tumors. *Radiographics* 1997; 17: 1559–1565.
7. Migda MS, Migda M, Migda B, Slapa RZ, Mlosek RK: Feasibility of using high-frequency skin ultrasound (HFSU) in vulvar skin assessment – initial report with the description of HFSU anatomy. *Ginekol Pol* 2016; 87: 19–25.
8. Mlosek RK, Malinowska S: Ultrasound image of the skin, apparatus and imaging basics. *J Ultrason* 2013; 13: 212–221.
9. Warszawik-Hendzel O, Olszewska M, Maj M, Rakowska A, Czuwara J, Rudnicka L: Non-invasive diagnostic techniques in the diagnosis of squamous cell carcinoma. *J Dermatol Case Rep* 2015; 9: 89–97.
10. Carbotta G, Sallustio P, Prestera A, Laforgia R, Lobascio P, Palasciano N: Perineal Paget's disease: A rare disorder and review of literature. *Ann Med Surg (Lond)* 2016; 9: 50–52.
11. Edey KA, Allan E, Murdoch JB, Cooper S, Bryant A: Interventions for the treatment of Paget's disease of the vulva. *Cochrane Database Syst Rev*. 2013: CD009245.
12. Onaiwu CO, Salcedo MP, Pessini SA, Munsell MF, Euscher EE, Reed KE *et al.*: Paget's disease of the vulva: A review of 89 cases. *Gynecol Oncol Rep* 2017; 19: 46–49.
13. Polańska A, Dańczak-Pazdrowska A, Jałowska M, Żaba R, Adamski Z: Current applications of high-frequency ultrasonography in dermatology. *Postepy Dermatol Alergol* 2017; 34: 535–542.
14. Hillmann BR, Pereira AA, Sommacal LF: Extramammary Paget disease of the vulva – case report. *Rev Bras Ginecol Obstet* 2016; 38: 524–528.