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 neoplasias1. Extramammary manifestation occurs in the apocrine glands and is a form of intraepidermal adenocarcinoma2. The most commonly affected regions are the vulva, perineum, perianal region, scrotum, penis, or pubic area4. Although surgery is the most effective treatment, a complete excision is challenging due to PDV intraepithelial reticular spread5. According to recent data, the rate of involved margins after primary surgery is up to 72%9. High-frequency ultrasonography (HFUS) is a technological advance that has been used for over 30 years in dermatology6-8. 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 evaluation9. 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 vulva7 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...
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 neoplasmatic 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 subepidermal 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. 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
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.

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