

Retiform hemangioendothelioma of the vulva: a case report

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Background: To report a rare case of retiform hemangioendothelioma (RH) originated from the vulva and review the relative references.

Case: A 38-year-old woman was admitted to our hospital with a vulvar subcutaneous nodule (diameter about 2 cm) for 2 years. The clinical examination findings, laboratory test results, including complete blood count and liver function test results were normal. The B mode ultrasound revealed that a hypoechoic nodule with a clear boundary was observed on the right side of the vulva. The size was measured to be 23 mm × 8 mm. An expanded resection with the excision range extending 1 cm and the depth reaching the fascia was performed. The histologic examination revealed that the elongated, arborizing, thin-walled vessels were abundant on the tumor, the inner wall of the vessels was arranged with monolayer cells with uniform morphology. Local infiltration of lymphocyte has been found around the vessels. Immunohistochemical examinations showed that the endothelial cells were CD31, CD34 and friend leukemia virus integration 1 (Fli-1) positive, the smooth muscle actin (SMA) was scattered positive, further confirming the diagnosis of RH. The patient was followed up for 15 months, no local recurrence was found. **Conclusions:** Vulvar region is the rare affected region of RH, which is lack of characteristic findings in clinical examinations and laboratory tests. The pathology and immunohistochemistry are necessary for the diagnosis of vulvar RH. Local expanded resection of tumor may be an effective method for the treatment of early-stage RH to reduce the risk of local recurrence.

Keywords

Retiform hemangioendothelioma; RH; Vulva; CD31; CD34; Fli-1

1. Introduction

Retiform hemangioendothelioma (RH) is a rare intermediate vasogenic tumor, which was first described in 1994 [1], with less than 40 cases reported in the worldwide [2]. Different from cutaneous angiosarcoma, the clinical manifestations RH are intermediate between a benign hemangioma and a high-grade angiosarcoma, therefore, the term “intermediary malignant tumor” was defined. Women in middle-aged are affected more commonly [3]. The limbs and trunk are the commonly involved organs of RH, follow by penis, scalp, jejunum and external genital area [4–6]. Distant metastasis is rare, however, local recurrence occurs in about 50% cases [7]. Herein, we report a case of RH in the vulvar area, and discuss the immunophenotypic features and treatment methods of RH in the vulva.

2. Case presentation

A 38-year-old woman with a medical history notable for caesarean section for 14 years, a uterine myoma (diameter about 6 cm) for 5 years, and a vulvar subcutaneous nodule with a diameter about 2 cm for 2 years prior to presentation. Physical examination revealed that a mass with approximately 2 cm in diameter on the right vulva. The margin of the mass was well-defined, the texture was in a medium quality and fair mobilizable activity. There were no tenderness or ulceration on the surface. No abnormality was found in the laboratory tests including carbohydrate antigen 125, cervical screening for human papillomavirus (high-risk type) and liquid-based cervical cytology. Transvaginal ultrasound showed that a hypoechoic area of 57 mm × 47 mm in the uterine wall and an anechoic area of 24 mm × 10 mm in the right ovary were observed. The B mode ultrasound revealed that the hypoechoic nodule with a clear boundary and the size of 23 mm × 8 mm was observed on the right side of the vulva. Furthermore, the blood flow signals were also observed (Fig. 1). The initial diagnosis was uterine myoma, right ovarian cyst and right vulvar nodule. We considered that the vulvar nodule might be leiomyoma, lipoma, endometriosis or malignant tumor. Although we did not perform a pre-operative biopsy for the vulvar nodule to determine the pathological properties, we performed a local resection of the vulvar nodule. The Intraoperative frozen pathology reported that the vulvar nodule was between benign or borderline tumor, with abundant positive tumor cells at the incisal edge. Afterwards, an expanded resection was performed, with the excision range extending 1 cm and the depth reaching the fascia. Meanwhile, laparoscopic myomectomy and right ovarian cystectomy were performed. After the surgery, the histologic examination of the vulvar nodule revealed that the elongated, arborizing, thin-walled vessels were abundant in the tumor, which formed rete-testis-like networks. The inner wall of the vessels was arranged with monolayer cells with uniform morphology. The cytoplasm of the cells was sparse, the nucleus was prominent, and the vascular lumen appeared as nail-like structure. Therefore, the presumptive diagnosis of RH was confirmed. The immunohistochemical staining results showed that the CD31, CD34 and Fli-1 in the endothelial cells, the SMA (smooth

muscle actin) in the vascular wall, and the Ki-67 (5%) were positive, further verifying the diagnosis of RH of the vulva (Fig. 2). Meanwhile, the pathological diagnosis was uterine leiomyoma and right ovarian follicular cyst. A regular physical examination has been performed after operation, no local recurrence has been found during the 15 months follow-up.

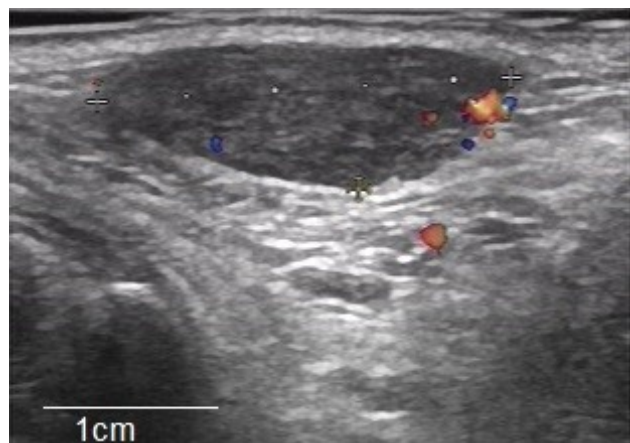


Fig. 1. Ultrasound image of the vulvar nodule: B mode ultrasound showed clear boundary and blood flow signals.

3. Discussion

The etiology of RH is still unknown [7]. Although several cases suggested its association with human herpesvirus-8, nonepidermal malignant tumors, previous radiation treatment and prior lymphangioma lymphedema, the exact associations have not been explicitly established [8–11].

RH has no specific clinical, laboratory or radiographic characteristics. However, in this case, B mode ultrasound showed clear boundary and blood flow signals, which may provide more information available for the diagnosis of RH. The diagnosis of RH is completely depended on the histopathological findings. The typical RH tumor tissue is characterized by a network of elongated and arborizing vessels, which similar to the normal testicular structure. The inner wall of the blood vessel is arranged with a monolayer cells, which have uniform morphology, hyperchromatic nuclei, sparse cytoplasm, and generally no atypia or nuclear mitosis [12, 13]. Immunohistochemical staining results are featured by positive factor VIII-related antigen, CD31, CD34 and D2-40, and negative or weakly positive SMA [2, 4, 14–16].

Surgical resection for early-stage RH is the most common treatment. However, the standard routine of the operation has not been determined. According to Emberger *et al.* [15], it was necessary to expand the surgical range to 3 cm out of the edge of the tumor, because the high local recurrence rate of RH. In this case, the expanded resection with only 1 cm extended cutting edge was performed due to the vulvar position. The patient has been monitored for 15 months after

surgery, and no local recurrence has been found, suggesting that the feasibility of 1 cm resection of early-stage RH of the vulva.

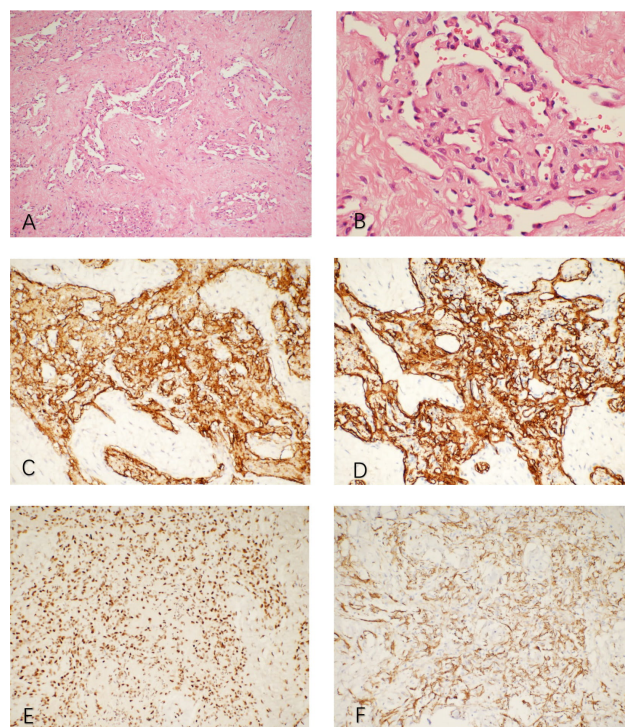


Fig. 2. The characteristics of tumor histopathology. (A) Elongated, arborizing, thin-walled vessels (H&E staining, $\times 100$). (B) The inner wall of the vessel was lined by a single layer of cells, with equal size and single morphology (H&E staining, $\times 400$). (C) Endothelial cells are immunohistochemically reactive for CD31 (immunohistochemistry staining, $\times 200$). (D) Endothelial cells are immunohistochemically reactive for CD34 (immunohistochemistry staining, $\times 200$). (E) Endothelial cells are immunohistochemically reactive for Fli-1 (immunohistochemistry staining, $\times 200$). (F) The smooth muscle actin (SMA) in the vascular wall was scattered positive (immunohistochemistry staining, $\times 200$).

4. Conclusions

In conclusion, RH of vulvar area is rather rare. The diagnosis of RH is clinically and radiologically challenging, and the pathology examination is necessary. Local expanded resection of tumor may be an effective method for the treatment of early-stage RH to reduce the risk of local recurrence.

Abbreviations

Fli-1, Friend leukemia virus integration 1; RH, Retiform hemangioendothelioma; SMA, Smooth muscle actin.

Author contributions

HJW and ZCD were major contributors in writing the manuscript, XY performed the histological examination, HJW, ZCD and YY analyzed and interpreted the patient data. All authors read and approved the final manuscript.

Ethics approval and consent to participate

All participants have given their consent for use of the medical data.

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Conflict of interest

The authors declare no conflict of interest.

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