

CASE REPORT

A case report of vulvar embryonal rhabdomyosarcoma in an adult pregnant woman

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Abstract

Embryonal rhabdomyosarcoma (ERMS) is the most common soft tissue sarcoma in children, but it is rare in adults, where cases are reported as individual cases or series. We present an ERMS found in the vulva of a 24-year-old pregnant woman who complained of a painless soft tissue mass in her left vulva. The mass grew rapidly within a month, and abortion, internal iliac artery embolization, and chemotherapy were performed. Despite an initially good response, cancer unfortunately progressed, and the patient passed away within one year after diagnosis. Adults with ERMSs tend to have poorer outcomes than children, and our case suggests that in some cases, pregnancy may accelerate the deterioration of the tumor.

Keywords

Adult; Embryonal rhabdomyosarcoma; Magnetic resonance imaging; Pregnancy; Vulvar mass

1. Introduction

Rhabdomyosarcoma (RMS) is a highly aggressive form of cancer developed from mesenchymal cells that have failed to differentiate fully into myocytes of skeletal muscle. Embryonal rhabdomyosarcoma (ERMS) and alveolar rhabdomyosarcoma are the more common of the two major sub-types of RMS. ERMS accounts for 60% to 70% of RMS; the five-year overall survival of localized of which exceeds 70% in children but is very poor in adult patients [1]. However, the prognosis in adults is unclear. We present a case of ERMS found in the vulva of an adult pregnant woman. The tumor increased in a short period, and the patient passed away one year after being diagnosed. We proposed that, in some circumstances, pregnancy may be responsible for accelerating the rapid progression of the disease, in addition to the tumor's aggressiveness.

2. Case Report

A 24-year-old woman, pregnant for more than four months, presented to the hospital for treatment when she found a painless soft tissue mass in her left vulva. The ultrasound imaging revealed a lesion was $4.6 \times 2.9 \times 4.4$ cm in size. The tumor rapidly grew over the subsequent 15 days, prompting her visit to our hospital for more evaluation and care. Laboratory values were average, including tumor-associated antigens such as the carcinoembryonic antigen, carbohydrate antigen 125, and alpha-fetoprotein. Physical examination revealed a hard mass on the patient's left vulva, with no ulceration on the surface of the mass and a slight increase in local skin temperature.

Magnetic resonance (MR) images revealed a mass in the left vulvar region, with an irregular shape and clear boundary, measuring $6.5 \times 5.5 \times 6.0$ cm. Fat-suppressed T2-weighted images (Fig. 1A, B) and T1-weighted images (Fig. 1C) showed a homogeneous, moderate-intensity mass. The mass on diffusion weighted imaging ($b = 800$ sec/mm²), Fig. 1D) and the apparent diffusion coefficient images (Fig. 1E) showed the mass with restricted diffusion. Dynamic contrast-enhanced T1-weighted images (Fig. 1F) showed heterogeneous enhancement. Fine-needle aspiration cytology showed a diffuse distribution of small dysplastic round and spindle striated muscle cells on hematoxylin-eosin staining (Fig. 2A). Immunohistochemical results showed a positive reaction for myogenin (Supplementary Fig. 1), sarcomeric actin (Fig. 2B), desmin (Fig. 2C), MyoD1 (Fig. 2D), recombinant cluster of differentiation 56 (CD56) and vimentin, and an adverse reaction for creatinine kinase, synaptophysin, neuron-specific enolase, chromogranin A, S100, and anti-human melanoma monoclonal antibody (HMB45). Twenty-six days after the mass was been discovered, the patient performed an induced abortion. The pelvic computed tomography images after induced abortion showed that the mass was irregular in shape, with unclear boundary, about $6.7 \times 6.3 \times 7.3$ cm in size, with uniform soft tissue density. The enhanced scan showed obvious uneven enhancement (Supplementary Fig. 2) increase.

We classified the ERMS into Stage 1, Group III, according to the rhabdomyosarcoma risk group classification by the Soft Tissue Sarcoma Committee of the Children's Oncology Group [2]. After discussion by a multidisciplinary team, the patient was treated with internal iliac artery embolization and 4 cycles of ifosfamide ($1.2\text{--}2.0$ g/m²) and cisplatin chemotherapy (21

days/cycle). After this treatment, the tumor volume was significantly reduced (Fig. 3). The patient received several courses of chemotherapy in another hospital after being discharged from our hospital. Unfortunately, one year after onset, the patient died of systemic bone and lung metastases.

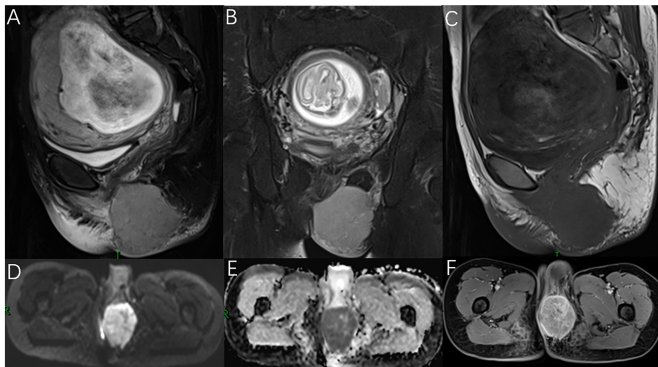


FIGURE 1. MRI of the vulvar mass before abortion and internal iliac artery embolization. Sagittal (A) and coronal (B) fat-suppression T2-weighted imaging, sagittal T1-weighted imaging (C), diffusion-weighted imaging (D), apparent diffusion coefficient map (E) and axial dynamic contrast-enhanced MRI (F) before iliac artery embolization and chemotherapy.

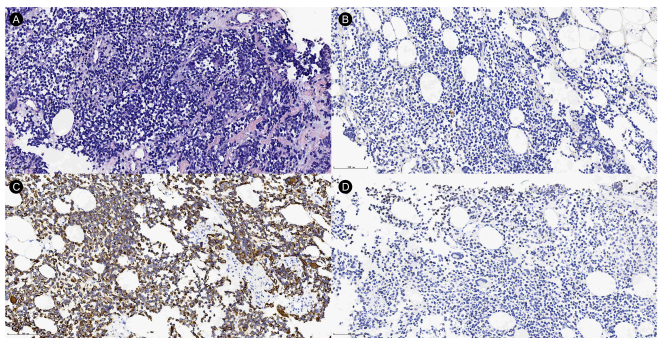


FIGURE 2. Hematoxylin-eosin staining and immunohistochemistry of the vulvar mass. Fine-needle aspiration cytology showed a diffuse distribution of small dysplastic round and spindle striated muscle cells on hematoxylin-eosin staining (A). Immunohistochemical results showed a positive reaction for sarcomeric actin (B), desmin (C), and MyoD1 (D).

3. Discussion

ERMS is one of the three pathological subtypes of RMS, the others of which are alveolar and pleomorphic. Embryonal and alveolar variants mainly occur in children, while adults typically develop pleomorphic variant [3]. We can find RMS in various parts of the body, and it occurs primarily in the head and neck (36%), genitourinary tract (23%), and extremities (20%) in children [4]. Nevertheless, RMS is uncommon in adults, especially in the genitourinary tract [5], and accompanying the pregnancy condition is very rare. We report a rare case of ERMS of the vulva in pregnancy.

Since the first vulvar RMS was reported in 1953 [6], the

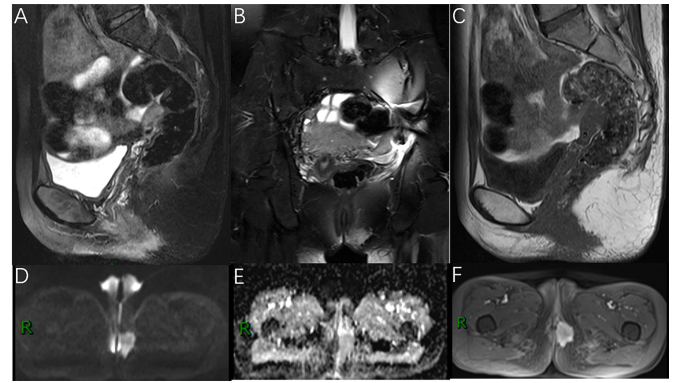


FIGURE 3. MRI of the vulvar mass after chemotherapy. Sagittal (A) and coronal (B) fat-suppression T2-weighted imaging, sagittal T1-weighted imaging (C), diffusion-weighted imaging (D), apparent diffusion coefficient map (E), and axial dynamic contrast enhanced MRI (F) showed the volume of the tumor decreased significantly after iliac artery embolization and chemotherapy.

literature on this rare disease predominantly comprises case reports and small case series. To our knowledge, only 4 cases of RMS of the genitourinary system complicating pregnancy have been reported, including three embryonal subtypes and one alveolar subtype. Of these four cases, two appeared in the uterine cervix and one in the vagina. The last case occurred in the perineum, in which the patient died of local and breast nodule progression seven months after the mass was discovered [7, 8]. In the other 3 cases, the patients were alive with no evidence of disease at 45 months, two years, and 54 months respectively. Both this patient, and the patient with perineal RMS, described above died of RMS within months due to rapid disease progression despite an excellent initial response to treatment.

More than half of the tumors in 9 cases of vulvar sarcoma enlarged during pregnancy, according to a review of the cases, suggesting that the increase of estrogen and progesterone during pregnancy may affect the progression of sarcoma [9]. In a patient with congenital vulvar lymphoma, symptoms began in adolescence and the tumor grew rapidly during pregnancy, thought to be due to increased hormonal secretion in stages [10]. Similar to this case, Xu H *et al.* [11] reported a case of aggressive angiofibroma of the vulva in a pregnant woman. The mass enlarged significantly during two pregnancies and decreased spontaneously postpartum, suggesting that pregnancy may promote tumor growth [11]. Herein we reported an extremely rare ERMS of the vulva in an adult pregnant woman, the tumor mass enlarged in a short time. We assumed that the reasons for the rapid growth of this tumor are as follows. First is the original aggressiveness of ERMS. In addition, maternal changes during pregnancy may also stimulate the tumor's rapid growth, such as the fluctuation of hormone levels, increased number of pelvic floor vessels, obstruction of venous blood, and lymph reflux of the pelvic floor. Finally, the patient did not undergo standard treatment for ERMS, VAC (Vincristine, Actinomycin D, and Cyclophosphamide) [2]. There is evidence that adult RMS is sensitive to chemotherapy and that diseases that respond

to chemotherapy develop metastases and are less likely to metastasize than disease that does not [12].

Although the embryonal subtype is the least aggressive, in two adult RMS series [12, 13], the progression of adult ERMS patients is rapidly, and the prognosis is worse than that of pediatric patients. In our case, the tumor showed intense aggressiveness and was prone to recurrence and metastasis, resulting in the patient's death within one year after being diagnosed with ERMS. These suggest that ERMS is highly aggressive in adults, even though the embryonal subtype is the least malignant. Therefore, when ERMS occurs in an adult, especially in a particular group of pregnant women, interventions should be performed as soon as possible, such as multimodal therapy, including surgery, combination chemotherapy, and radiation therapy to prevent the further development of the tumor. The use of chemotherapy drugs during pregnancy may lead to fetal malformations and intrauterine growth retardation, so when the patient wishes to preserve the pregnancy, clinicians are bound to face the dilemma of mother-infant management. Waiting for the fetal lung to mature may result in delayed or inadequate treatment for the mother. Unfortunately, there are no published data on the standard treatment of pregnant women with ERMS. In our case, clinicians implemented interventional chemoembolization and ifosfamide combined with cisplatin chemotherapy, which also achieved satisfactory results quickly. However, the patient developed systemic bone and lung metastasis and eventually died after the next few months, indicating that ERMS in adults is more aggressive than in children. Better treatment schemes are needed to improve the first-glance survival rate of adult patients with ERMS.

4. Conclusion

The vulvar EMRS is rare, significantly complicating pregnancy. In addition to the fact that the malignant nature of the tumor causes it to grow very fast, we emphasized that pregnancy may also influence the growth of cancer, which remains to be confirmed. The outcome of adult ERMS is often unsatisfactory, and clinicians should be aware of the possibility of ERMS and take rapid treatment measures when the tumor overgrows in a short time. However, adult ERMSs are currently treated according to pediatric protocols, and there is no standard treatment for pregnant patients. Further awareness and recognition of this rare malignancy are essential to optimize management and clinical outcomes.

AUTHOR CONTRIBUTIONS

JW—prepared the first draft. XJM, LJ, and HY—revised the manuscript. TL—completed the pathology review, LW—collected clinical data, SGL—edited and finalized the manuscript, and all authors reviewed and approved the final manuscript.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Ethical review is not required according to the ethics review committee of the Third Affiliated Hospital of Zunyi Medical University. The patient's husband provided informed consent and agreed to publication of the details of this case.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

SUPPLEMENTARY MATERIAL

Supplementary material associated with this article can be found, in the online version, at <https://oss.ejgo.net/files/article/1669274875630370816/attachment/Supplementary%20materials.pdf>.

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