

PERINEAL APPROACH IN VAGINAL YOLK SAC TUMOR SURGERY: CASE REPORT

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ABSTRACT

Objectives: Yolk sac (endodermal sinus) tumor arising primarily from vagina is extremely rare, which has no published standardized management approach. We present a case of yolk sac tumor of vagina, in which we directed a special attention towards chemotherapy along with an appropriate surgical approach to remove the tumor and preserve sexual and reproductive function.

Case study: Report a case of a vaginal yolk sac tumor in a 7-month-old female infant who was admitted to hospital with history of vaginal bleeding. Serum alpha-fetoprotein, computed tomography and pathologic biopsy samples confirmed a vaginal yolk sac tumor. The patient was given six cycles of chemotherapy along with vaginoscopic-assisted tumor resection and continued to follow up. After 8 months treatment and following up, the AFP level increased again and radiography showed the recurrent tumor. The patient was taken to surgery, where completely tumor resection was done through perineal approach. AFP returned to normal level after surgery.

Conclusion: Perineal approach appears to be simple and efficient to remove the tumor in vaginal position, which enable to preserve patient's reproductive anatomy and function.

Keywords: yolk sac tumor, vaginal tumor, fertility preservation, alpha-fetoprotein

I. INTRODUCTION

Yolk sac tumor (endodermal sinus tumor) (YST) was described firstly by Teilmann as a type of malignant germ cell tumor [4]. In children, YST usually emerges from testes or extragonadal organs. Vaginal YST is rare with several cases were published in literature. Vaginal bleeding is the most common symptom. Chemotherapy in addition to surgery is the current treatment for this tumor [6]. The prognosis of YST has been considerably improving during recent decades by the development of chemotherapy along with surgical procedure [3].

II. CASE REPORT

A 7-month-old with normal pregnant history,

presented to hospital with periodic vaginal bleeding in a week. Radiography (ultrasound and computed tomography) showed a non-heterogeneous mass located in the vagina behind the bladder, measuring 40x35x28mm. Serum alpha fetoprotein (AFP) was highly elevated at 6186 ng/mL and Beta-human chorionic gonadotropin was normal.

The patient was taken vaginoscopic biopsy under anesthetic. Pathology showed reticular structure with microcystic spaces, lined by flat or columnar cells, some papillae projecting into these spaces, occasional Schiller Duval bodies. Hence the diagnosis of yolk sac tumor was made based on examination, elevated alpha fetoprotein radiology and pathology.

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Fig 1. Recurrent tumor (Arrow)

The patient received 6 cycles of chemotherapy with CEB regimen (carboplatin, etoposide, and bleomycin), the serum AFP returned to normal (33ng/mL) after 3 cycles. When chemotherapy was complete, vaginoscopic-assisted tumor resection

was performed. In this operation, limited surgical field obstructed our observation and tumor resection. After 8-month following up, the tumor was recurrent with increased serum AFP to 235 ng/mL, CT scan showed a mass in left pelvis with 12x15mm in size (Figure 1). The patient was operated again through perineal approach to completely remove the recurrent tumor.

The patient was put in obstetric position. Perineal incision was performed at the fifth hour position near the vagina (Figure 2), continued to open in vaginal wall to approach the tumor (Figure 3). Surgical field was achieved through a lone star equipment, which showed the tumor in anterior lateral vaginal wall (from 12h to 4h), sized 10x15mm, near cervix, relatively clear margin (Figure 4). The tumor was resected entirely. Vaginal wall was recovered quite feasibly, and minivac drain was put para-incision. There was no complications in postoperative period. The patient was discharged after 5 days, and serum AFP returned to normal level after 2 weeks.



Figure 1. Perineal incision

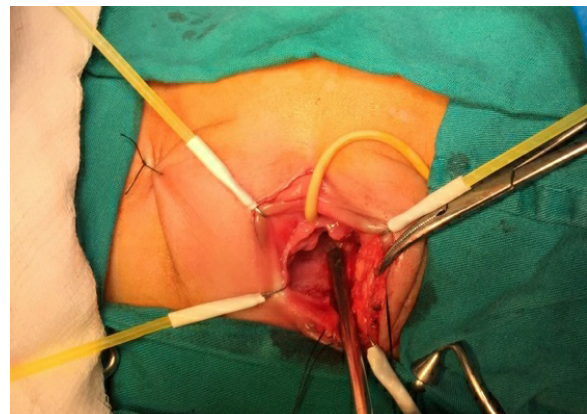


Figure 3. Expansion the vaginal wall to expose the tumor

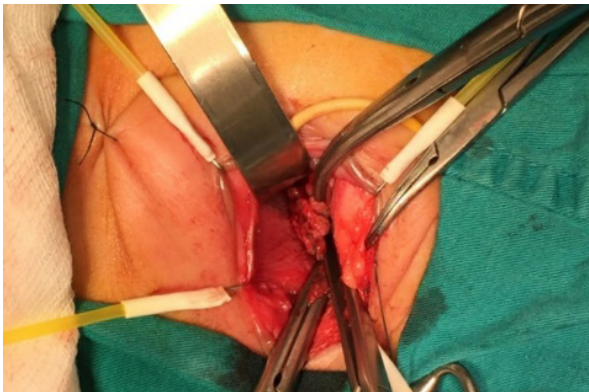


Figure 4. Complete tumor remove

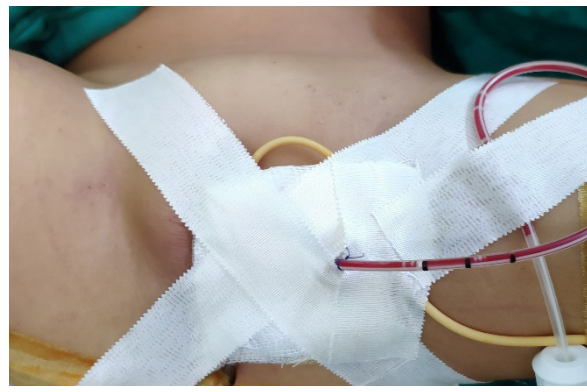


Figure 5. Minivac drain and bandaging the wound

III. DISCUSSION

Malignant germ cell tumor accounts for 3% malignant tumor in children, which is usually emerge from sacrococcyx, testes, and ovaries. YST which is atype of malignant germ cell tumor usually emerges from gonads. There are some rare cases in which these tumors emerge from extragonadal organs such as vagina, mediastinum, pituitary gland, cervix [1].

Primary vaginal yolk sac tumor is extremely rare with several cases was reported. The most popular symptom is vaginal bleeding, with or without a mass protruding from vagina [3]. The typical histiology is a reticular meshwork and papillae-like structures consisting of a central vascular core lined by a single layer of cells (Schiller-Duval bodies). These tumors are immunohistochemically positive for AFP. In addition, serum AFP level is a crucial marker for diagnosis and following up. [3], [6]

Chemotherapy in associated with surgery are commonly recommended management. With the development of chemotherapy, this protocol shows a good result, improves the survival prognosis, and more importantly preserves the sexual and reproductive function [3]. Operation is performed to resect the tumor and combined adjuvant chemotherapy. However, if surgery cannot remove the whole the tumor, there would be a risk of recurrent despite of an appropriate chemotherapy [6]. Hence, completed tumor resection takes an important role in treatment and prognosis.

Approach in tumor resection is still a challenge for surgeons. For the tumor in vagina, near the cervix, surgeons tend to approach the tumor through

abdomen or vaginoscopy [5].

In vaginoscopic-assisted approach, the narrow surgical field makes it difficult to evaluate the tumor's margin and to remove the tumor, leading to high risk of recurrent. We encountered this situation in the first operation. While in abdominal approach, surgeons sometimes have to removal the cervix, uterus or vagina to expose and remove the tumor. In this situation, it would be a challenge for surgeons in term of preservation the whole genital anatomy for the rest of patient's lifetime.

In the next operation, we decided to approach through perineum, expanded the vagina to expose the operative field. Once the tumor was clearly exposure, tumor resection was performed much easier and more feasible than the previous operation. The patient was discharged early without any postoperative complications.

After the surgery, AFP level returned to normal level. Long-term following up is necessary to determine the effective treatment. Through this case, we personally believe that perineal approach as mentioned above is an appropriate method in childhood vaginal tumor surgery.

IV. CONCLUSION

Vaginal YST is rare and highly malignant, the prognosis, however, is relatively good based on the advances of chemotherapy and appropriate surgery. Serum AFP should be considered in girls with vaginal bleeding to facilitate the diagnosis. Perineal approach is relatively simple and effective to remove the vaginal tumor, which enable to conserve patients' reproductive anatomy and function.

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