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(CASE REPORT)

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# Vaginal Rhabdomyosarcome: Case Report

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## Abstract

**Introduction**: Rhabdomyosarcoma is a soft tissue sarcoma, most common in children, localised in the female genital tract and therefore rare in adults. Among its subtypes, it is most often distinguished as embryonal. It is an aggressive tumour, difficult to diagnose, is usually detected by CT or MRI and the diagnosis is confirmed by biopsy and treatment involving surgical excision, radiotherapy and chemotherapy. The authors present a case of embryonal rhabdomyosarcoma localised in the vagina, highlighting relevant aspects of the subject.

**Observation**: We report the observation of a girl aged 2 years and 8 months, with no notable antecedents, from a 2nd degree consanguineous marriage. For 1 month she presented with a rapidly progressive vaginal mass associated with bleeding. Examination revealed a reddish, grape-like mass in the vulva. Abdomino-pelvic ultrasound revealed a vaginal tumour, abdomino-pelvic MRI revealed an endovaginal mass delivered through the vulva, suggesting a botryoid rhabdomyosarcoma, and biopsy of the vaginal mass with morphological appearance and immunohistochemical profile of an embryonal rhabdomyosarcoma. The extension work-up showed a vulvovaginal lesional process, with no secondary location on the thoraco-abdominal CT scan. The patient was classified as group C according to the RMS 2005 protocol and received 6 courses of IVA + vaginal brachytherapy + 3 courses of IVA with a good clinical and biological evolution.

**Conclusion**: In view of the diagnostic difficulties and the risk of progression and rapid worsening of rhabdomyosarcoma, early positive diagnosis means that treatment can be started at an early stage with chemotherapy, surgery and radiotherapy.

**Keywords:** Embryonal rhabdomyosarcoma; Endovaginal mass; Immunohistochemistry; IVA treatment; Vaginal brachytherapy

## 1. Introduction

Rhabdomyosarcoma is a soft tissue sarcoma, most common in children, localised in the female genital tract and therefore rare in adults.

Among its subtypes, it stands out as the most common embryonal. It is an aggressive tumour, difficult to diagnose, are usually detected by CT or MRI and the diagnosis is confirmed by biopsy and treatment involving surgical excision, radiotherapy and chemotherapy.

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Vaginal Rhabdomyosarcoma is a rare vulvovaginal tumour and a highly malignant soft tissue sarcoma, composed of cells with round, oval or spindle-shaped nuclei and eosinophilic cytoplasm. The tumour may differentiate into striated muscle cells. It usually affects children, and manifests as a vulvar or vaginal mass that may be polypoid or cluster-shaped (embryonic subtype). It is associated with bleeding and ulceration.

## 2. Observation

We report the observation of a girl aged 2 years and 8 months, with no notable antecedents, from a 2nd degree consanguineous marriage. For 1 month she presented with a rapidly progressive vaginal mass associated with bleeding. Examination revealed a reddish, grape-like vaginal mass in the vulva (Figure 1). Biological work-up revealed normocytic normochromic anaemia, high CRP, BHCG <2, Alpha FP at 1.42, Cytobacteriological examination of urine negative, Diagnostic work-up, Abdominopelvic ultrasound revealed a vaginal tumour measuring 65×46×29 mm; Abdominopelvic MRI found an endovaginal mass delivered through the vulva, measuring 10×5.(Figure 2)

Biopsy of vaginal mass with morphological appearance and immunohistochemical profile of an embryonal rhabdomyosarcoma.

The extension work-up showed a vulvovaginal lesion, with no secondary lesions, and a thin right anterior pneumothorax on the thoracoabdominal CT scan. (Figure 3)

Trans-thoracic ultrasound and chest X-ray were normal; myelograms of both crests showed no metastatic cells or clusters in the crests.

The patient was classified as group C according to the RMS 2005 protocol on the basis of favourable histology + IRS 3 + absence of regional lymph nodes + age < 10 years + favourable site (vagina). The patient received 6 courses of IVA + vaginal brachytherapy + 3 courses of IVA with good clinical and biological progression. Disappearance of the intra-vaginal mass. (Figure 4)(Disappearance of the endovaginal signal anomaly with no sign of recurrence or tumour residue on abdomino-pelvic MRI)

**Figure 1** A vaginal mass delivered through the vulva in bunches of grapes

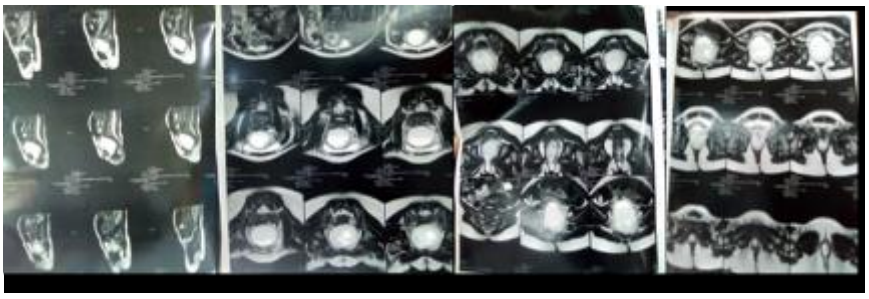

**Figure 2** Abdominopelvic MRI finding an endovaginal mass originating through the vulva

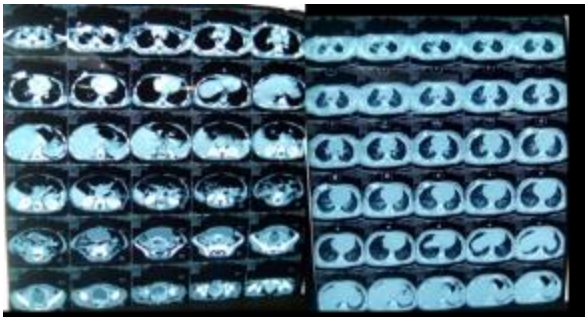

**Figure 3** Thoracoabdominal CT scan. Vulvovaginal lesions, with no secondary location

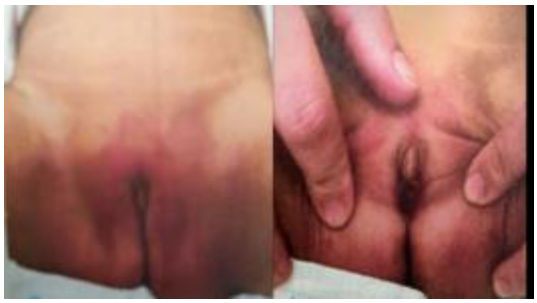

**Figure 4** Disappearance of the intra-vaginal mass.

## 3. Discussion

Rhabdomyosarcoma is a cancer arising from embryonic mesenchymal cells that tend to differentiate into skeletal muscle cells. It can develop in almost any type of muscle tissue, in any location, with highly variable clinical manifestations. It is usually detected by CT or MRI and the diagnosis is confirmed by biopsy. Treatment involves surgery, radiotherapy and chemotherapy.

Rhabdomyosarcoma is the third most common solid cancer outside the central nervous system in children (after Wilms' tumour and neuroblastoma). Rhabdomyosarcoma belongs to a group of tumours known as soft tissue sarcomas and is the most common cancer in this group. Two-thirds of rhabdomyosarcomas are diagnosed in children under the age of 7. The disease is more common in white children than in those with black skin.

Although rhabdomyosarcomas can occur in both children and adults, they differ significantly in terms of histological type, site of disease and overall outcome.

There are 2 main histological subtypes, embryonal and alveolar.

Although rhabdomyosarcoma can be found almost anywhere in the body, there are several areas of predilection.

Vaginal tumours may present as a mucosanguinous discharge with a polypoid mass protruding from the vagina.

Masses are assessed by CT and MRI. The diagnosis of rhabdomyosarcoma is confirmed by biopsy or excision of the mass.

The standard work-up includes a chest CT scan, positron emission tomography (PET), bone scan, bilateral myelogram and bone biopsy.

The therapeutic approach to rhabdomyosarcoma is multimodal, consisting of chemotherapy combined with surgery and/or radiotherapy.

The main aim of RMS treatment is to achieve local control of the tumour. Indeed, the evolution of these tumours, when they are not metastatic at diagnosis, is dominated by the risk of local recurrence.

At present, tumour removal as the first line of treatment should be reserved for cases where it can be completed without any functional consequences. It is therefore proposed mainly for small, easily accessible tumours.

Complete removal of the primary cancer is recommended where this can be done safely. Because cancer is sensitive to chemotherapy and radiotherapy, aggressive resection is not recommended if it is associated with organ damage or functional problems.

Regardless of the risk category, children are treated with chemotherapy. Chemotherapy is essential, and several drugs are effective in the treatment of SMI and are used in combination. Courses of VIA (ifosfamide, vincristine and actinomycin) are the most commonly used. Maintenance chemotherapy, given after IVA chemotherapy, has recently shown its value in RMS with a higher risk of recurrence.

Radiotherapy is generally recommended for children with massive or microscopic tumour residue after surgery and for children with intermediate-risk or high-risk disease.

Brachytherapy irradiation involves implanting radioactive products (iridium wires) in or near the tumour bed. This is a highly valuable technique because it delivers radiation to a very small volume at a high dose, and therefore reduces the risk of after-effects. However, it can only be used for tumours that are easily accessible (vagina, bladder, prostate, limbs, walls) and small in volume (< 5 cm). Children must be referred to a referral centre for this highly specialised treatment.

## 4. Conclusion

Given the difficulty of diagnosis and the risk of progression and rapid worsening of rhabdomyosarcoma, an early positive diagnosis means that treatment based on chemotherapy, surgery and radiotherapy can be introduced at an early stage.

## Compliance with ethical standards

### *Disclosure of conflict of interest*

No conflict of interest to be disclosed.

### *Statement of informed consent*

Informed consent was obtained from all individual participants included in the study.

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