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IMAGES IN PAEDIATRICS

# Malignant vaginal tumor: About a case

## Tumoración maligna vaginal: a propósito de un caso

**Mariona Morell Daniel***, **María Rosario Pérez-Torres Lobato**, **Miriam Morey Olivé**, **Luis Gros Subias**

*Servicio de Oncología y Hematología Pediátricas, Hospital Vall d'Hebron, Barcelona, Spain*

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We present the case of a girl aged 2 years presenting with a growing genital mass (Fig. 1A), leukorrhea and intermittent bleeding of 2 weeks' duration, without constitutional symptoms. The examination revealed a gelatinous polypoid mass with a grape cluster appearance (Fig. 1B). The tumour was surgically resected, the histological findings were compatible with botryoid rhabdomyosarcoma (desmin+, vimentin+, Ki67 30%, HHF35+) and metastasis was ruled out. The patient was treated per protocol (ISRG group III) with 15 cycles of chemotherapy with vincristine, actinomycin and cyclophosphamide and adjuvant pelvic floor radiotherapy, which achieved complete remission.

Rhabdomyosarcoma is the most frequent malignancy of the female genital tract in the paediatric age group.[1] The botryoid subtype is a variant of embryonal rhabdomyosarcoma that appears in girls aged less than 5 years in the vulvovaginal region, among others.[1,2] The differential diagnosis includes germ cell tumours, clear cell sarcoma, urethral prolapse and ureterocoele.[2] Imaging can help distinguish between these lesions, although the diagnosis is confirmed by histological methods (tissue positive for vimentin and markers of myoid differentiation, such as actin antibody HHF35, desmin and myoglobin, and histological appearance characterised by alternating areas of hypocellularity and hypercellularity in a myxoid/oedematous stroma forming a *cambium* layer under the epithelium[3]). Treatment consists of surgery, polychemotherapy and radiotherapy,[2] and the 5-year survival for localized tumours is of 80%.[1] Therefore, the presence of a rapidly growing mass in the genital region with a grape-like appearance should trigger suspicion in the paediatrician, given the importance of early diagnosis.

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* Corresponding author.
*E-mail address:* mariona.morell@gmail.com (M. Morell Daniel).

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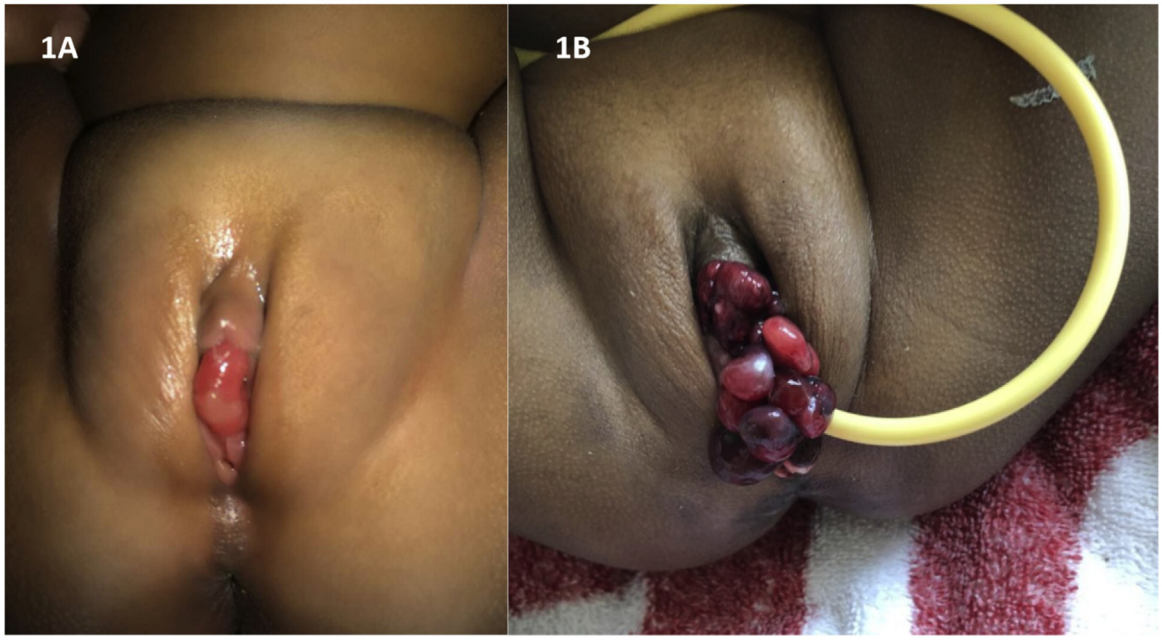


**Figure 1** Image A shows a polypoid lesion of 2 weeks' duration. Image B shows the same mass 2 months later. Mass characterised by multiple dark red polypoid-tuberous lesions with a grape cluster appearance occupying the vulvar mucosa and protruding through the introitus.

## References

1. Fernandez-Pineda I, Spunt SL, Parida L, Krasin MJ, Davidoff AM, Rao BN. Vaginal tumors in childhood: the experience of St. Jude Children's Research Hospital. J Pediatr Surg [Internet]. 2011;46:2071–5.
2. Raney RB, Anderson JR, Andrassy RJ, Crist WM, Donaldson SS, Maurer HM, et al. Soft- tissue sarcomas of the diaphragm: a report from the Intergroup Rhabdomyosarcoma Study Group from 1972 to 1997. J Pediatr Hematol Oncol [Internet]. 2000;22:510–4.
3. Bennett JA, Ordulu Z, Young RH, Pinto A, van de Vijver K, Burandt E, et al. Embryonal rhabdomyosarcoma of the uterine corpus: a clinicopathological and molecular analysis of 21 cases highlighting a frequent association with DICER1 mutations. Modern Pathol. 2021;34:1750–62.