

IMAGES IN PAEDIATRICS

Malignant vaginal tumor: About a case**Tumoración maligna vaginal: a propósito de un caso**

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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 hypcellularity and hypercellularity in a myxoid/oedematous stroma forming a cambium layer under the epithelium³). 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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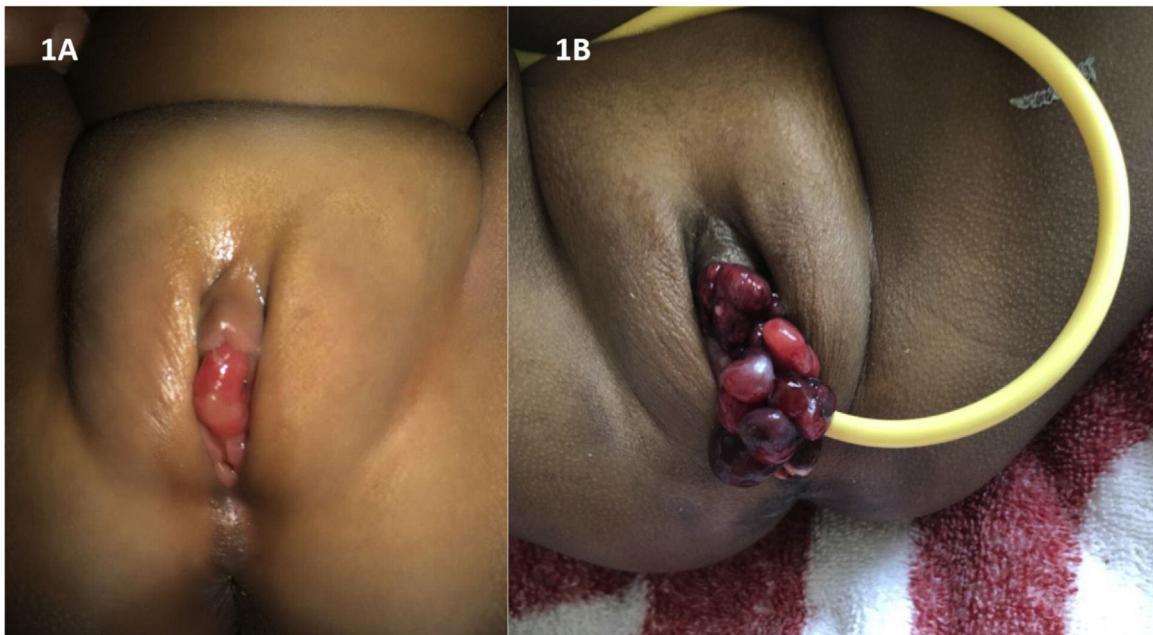


Figure 1 Image A shows a polypoid lesion of 2 weeks' duration. Image B shows the same mass 2 months later. Mass characterised by multiple dark red polypoid-tuberous lesions with a grape cluster appearance occupying the vulvar mucosa and protruding through the introitus.

References

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