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Sarcoma Botryoides a Rare Pediatric Pelvic Tumor: A Case Report and Literature Revue

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Abstract **Case Report**

Sarcoma botryoides, a variant of embryonal rhabdomyosarcoma, represents the predominant malignant tumor affecting the genitourinary (GU) system in children under 15 years of age. Among GU tract malignancies in this age group, the embryonal subtype of habdomyosarcoma is the most prevalent. In young and adolescent individuals, the cervix and uterus are affected; whereas in infants, vaginal lesions are more common. Imaging plays a crucial role not only in the initial diagnosis but also in long-term follow-up of genital rhabdomyosarcoma. Although the magnetic resonance imaging features of GU tract rhabdomyosarcomas have been escribed, there have been relatively few radiological reports in the literature regarding the botryoid variant. This article aims to illustrate and describe the imaging appearance of this rare tumor. We present a case of a one-year-old male who presented with a rapidly growing pelvic mass, crying and straining on micturition and difficulty in passing stool and urine. An abdomino-pelvic CT scan and MRI were performed. He had been examined under anesthesia and biopsy. Histology confirmed sarcoma botryoides.

Keywords: Sarcoma botryoides, Rhabdomyosarcoma, MRI, Pediatric pelvis masses.

INTRODUCTION

Sarcoma botryoides, also known as embryonal rhabdomyosarcoma, is a malignant neoplasm originating from embryonal rhabdomyoblasts. Its nomenclature derives from the Greek term "botryoid," meaning resembling a cluster of grapes, which aptly describes its clinical and radiological manifestation. This malignancy predominantly afflicts children and young adults, constituting 4–6% of malignancies within this demographic [1]. Typically localized to the female reproductive tract of infants, particularly the vaginal region, sarcoma botryoides infrequently manifests in the cervix or uterine fundus [2]. Vaginal bleeding is the most common presenting feature even though non-specific. It may also present as a polypoid or fleshy mass in the vagina, or more classically projecting from the introitus. Other forms of presentations include urinary symptoms especially when the tumor is anteriorly situated or tenesmus where there is posterior extension [3]. While magnetic resonance imaging (MRI) characteristics of genitourinary (GU) tract rhabdomyosarcomas have been delineated, there is a paucity of radiological documentation in the literature pertaining specifically to the botryoid subtype. The objective of this article is to elucidate and delineate the imaging manifestations of this uncommon tumor.

CASE REPORT

A one-year-old male who presented with a rapidly growing sacro-pelvic mass, crying and straining on micturition and difficulty in passing stool and urine. There was a positive history of anorexia, easy fatigability, as well as weight loss. No significant family or social history. The clinical review of other systems was unremarkable.

The examination revealed a diffuse abdominal tenderness and a sacro-pelvic mass was palpated.

An abdomino-pelvic CT (Fig-1) and a pelvic MRI (Fig-2) were performed and revealed a centropelvic compressive mass with involvement of the sacrococcygeal region, bone lysis and endocanal extension.

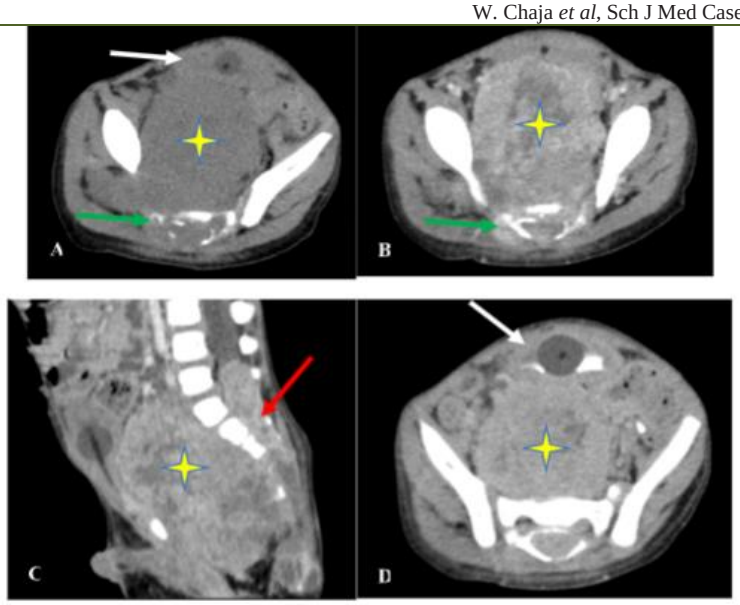

Fig-1: Axial image (A) of abdomino-pelvic CT scan without contrast, axial (B) and coronal (C) images with contrast in veinous phase and excretory phase (D) showing a centropelvic mass (stars), with involvement of the sacrococcygeal region, predominantly solid with delineated cystic areas, heterogeneous enhancement following contrast injection. This mass is causing mass effect on the catheterized bladder (white arrows) and a sacrococcygeal bone lysis (green arrows) with endocanal extension (red arrows)

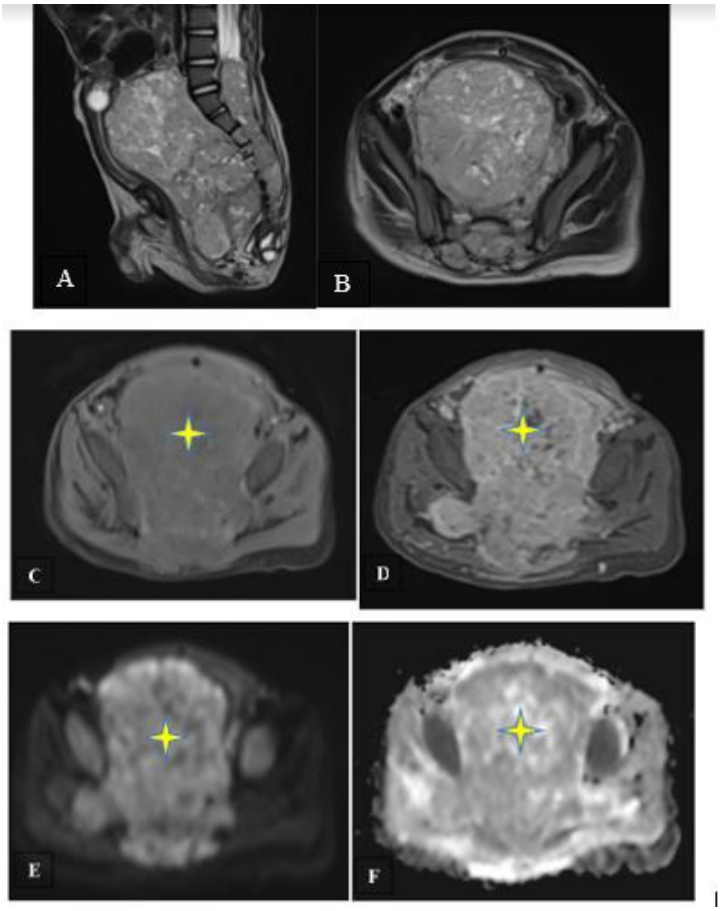

Fig-2: Pelvic MR imaging of the mass showing a centropelvic tumoral process (stars), with involvement of the sacrococcygeal region, on sagittal (A) and axial (B) T2 weighted fast spin echo, T1W fat suppressed axial image pre-contrast (C) and post-contrast (D), Diffusion-weighted sequences (E, F). The masse is demonstrating an heterogeneous high signal T2 (A, B), with cystic areas, intermediate T1 signal intensity (C), discreet high signal diffusion with ADC restriction (E, F) and heterogeneous post-contrast enhancement (D). This mass is causing mass effect on the catheterized bladder (white arrows) and an extensive sacrococcygeal endocanal invasion extending to the lumbar medullary canal (red arrows)

The patient underwent a chirugical exploration and biopsy. Histology demonstrated features of embryonal botryoid rhabdomyosarcoma (elongated spindly cells, with eosinophilic cytoplasm and cross-striations, inside a myxoid stroma which delimits a cambrium layer). A multidrug chemotherapy has been prescribed.

DISCUSSION

Rhabdomyosarcoma ranks among the most prevalent sarcomas diagnosed during childhood, comprising approximately 4% to 8% of all childhood malignancies before the age of 15 [4]. Notably, the genitourinary tract stands as the second most frequent site for rhabdomyosarcoma occurrence, trailing behind the head and neck region. In pediatric populations, rhabdomyosarcoma exhibits a bimodal distribution pattern, with an initial peak typically observed between the ages of 2 and 6 years, followed by a second peak occurring between 14 and 18 years of age. The International Classification of Rhabdomyosarcoma [5] categorizes tumors into distinct prognostic groups, including those with a favorable prognosis (such as botryoid and spindle cell rhabdomyosarcoma), intermediate prognosis (embryonal rhabdomyosarcoma), and poor prognosis (alveolar rhabdomyosarcoma and undifferentiated sarcoma). Botryoid rhabdomyosarcoma represents a variant of embryonal rhabdomyosarcoma, characterized by its growth in association with the mucosa of various organs, including the vagina, urinary bladder, or extrahepatic bile ducts.

The primary manifestation of sarcoma botryoides in the genital tract typically includes bleeding and pain attributed to the presence of an enlarging mass. Additionally, the observation of a polypoid mass protruding from the introitus is common. Patients may occasionally experience bone pain resulting from bone metastases; however, such occurrences are infrequent in the absence of clinical signs at the primary site. Sarcoma botryoides predominantly affects the vagina during infancy and childhood, the cervix during reproductive years, and the uterine corpus in postmenopausal women [6]. Rhabdomyosarcoma can disseminate through various routes, including direct extension, hematogenous spread to the lung, bone, liver, or bone marrow, or lymphatic dissemination to lymph nodes.

On gross pathology examination, sarcoma botryoides typically presents as an exophytic multinodular polypoid mass, resulting from cellular tumor proliferation pushing outward against the covering mucosal surface, hence the descriptive term "botryoid," meaning resembling a cluster of grapes. A defining histological feature of sarcoma botryoides is the identification of a hypercellular submucosal band composed of spindle-shaped tumor cells, known as the cambium layer, located just beneath the usually intact epithelium of the affected organ [7]. The area of the tumor distal from the mucosal surface exhibits lower cellularity compared to the cambium layer and is characterized by a more abundant myxoid stroma.

Before the introduction of ultrasound, intravenous urography (IVU) served as the primary diagnostic tool for evaluating pelvic or abdominal masses [8]. However, due to its noninvasive and painless nature, ultrasound has become the preferred initial imaging modality for assessing such masses in pediatric patients. Although the ultrasound appearance of rhabdomyosarcoma varies, it commonly presents as a polypoid mass with heterogeneous echogenicity [9].

Pelvic rhabdomyosarcoma also typically appears as a bulky mass with heterogeneous attenuation on computed tomography (CT), which is frequently utilized to monitor local disease progression and assess metastatic spread, particularly to the lungs [10, 11]. Recent studies have highlighted the efficacy of magnetic resonance imaging (MRI) in evaluating the local extent of pelvic rhabdomyosarcoma, as it offers superior visualization of tumor extension and residual disease, facilitating timely management. Rhabdomyosarcomas generally exhibit nonspecific low signal intensity on T1-weighted MRI sequences and high signal intensity on T2-weighted sequences, often with heterogeneous signal intensity due to hemorrhage at various stages [10, 12, 13].

Additionally, a distinct pseudocapsule, observable on both T1- and T2-weighted sequences, has been reported at the interface of the mass with adjacent fat [13]. The enhancement pattern of rhabdomyosarcomas is described as heterogeneous or speckled, corresponding to intratumoral septum-like structures observed histopathologically [13].

There is a scarcity of published literature discussing the appearance of the botryoid subtype of vaginal rhabdomyosarcoma. In this case detailed in this report, clear visualization of the tumor and its relationship with adjacent structures was achieved. The tumor presented as a septated cystic mass with high T2 signal intensity on MRI. Histopathological analysis revealed a superficial cellular layer and deeper portions abundant in myxoid stroma, corresponding to the observed "cystic" appearance on imaging. These myxoid stroma regions exhibited uniform brightness on T2 sequences and low signal on T1, indicative of the absence of hemorrhage, calcification, or proteinaceous debris. The mass exhibited a network of mildly thickened and nodular septa-like structures, showing avid enhancement following gadolinium administration. These "septations" likely represent deep infoldings of the surface epithelium with associated cambium layer of tumor, consistent with pathological findings [13, 14].

Complications of the disease include tumour rupture, invasion of adjacent structures, and metastases

to the lungs, cortical bones, and lymph nodes [15]. Liver and bone marrow metastases are less frequent. Metastatic disease is present in 10-20% of all patients with rhadbomyosarcomas at the time of diagnosis [16]. In the index case presented, there was no evidence of distant metastases. However, the tumour is locally invasive.

Children diagnosed with nonmetastatic rhabdomyosarcoma exhibit a notably favorable survival prognosis. Specifically, if the primary tumor originates entirely from a favorable site such as the vagina and is completely excised, the overall 3-year survival rate exceeds 90% [17]. Moreover, depending on the tumor stage, a multimodal approach involving chemotherapy, radiation, and surgery has demonstrated excellent survival outcomes while often preserving the full function of surrounding organs [18].

We advocate for MRI as the preferred imaging modality in patients with sarcoma botryoides or pelvic rhabdomyosarcoma, both for initial diagnosis and for post-treatment surveillance of residual disease. MRI offers several advantages over CT, including the absence of radiation exposure, utilization of safer contrast agents, and superior contrast resolution between tumor and normal tissues. The avoidance of ionizing radiation is particularly crucial in the pediatric population, where patients may have long life expectancies and thus face a heightened risk of radiation-induced malignancies compared to adults. Conversely, CT remains the modality of choice for evaluating distant tumor spread, including intraperitoneal metastatic disease [13].

CONCLUSION

Sarcoma botryoides is characterized by rapid tumor growth. We report a rare instance of the botryoid variant of embryonal pelvic rhabdomyosarcoma. We detail the imaging characteristics alongside pathological analysis. A multidisciplinary imaging approach aids in precise diagnosis and evaluation of tumor extent. MRI stands out for its superior delineation of tumor size, location, and relationship to adjacent organs, making it the preferred modality for assessing local disease.

BIBLIOGRAPHY

1. Dayyabu, A. L., Adogu, I. O., & Makama, B. S. (2014). Sarcoma botryoides a management dilemma: A review of two cases. *Int J Case Rep Images*, *5*(7), 15–20.
2. Behtash, N., Mousavi, A., Tehranian, A., Khanafshar, N., & Hanjani, P. (2003). Embryonal rhabdomyosarcoma of the uterine cervix: case report and review of the literature. *Gynecologic oncology*, *91*(2), 452-455.
3. Scaravilli, G., Simeone, S., Dell'Aversana Orabona, G., Capuano, S., Serao, M., Rossi, R., ... & Balbi, C. (2009). Case report of a sarcoma botryoides of the uterine cervix in fertile age and literature review. *Archives of gynecology and obstetrics*, *280*, 863-866.
4. Kobi, M., Khatri, G., Edelman, M., & Hines, J. (2009). Sarcoma botryoides: MRI findings in two patients. *Journal of Magnetic Resonance Imaging: An Official Journal of the International Society for Magnetic Resonance in Medicine*, *29*(3), 708-712.
5. Parham, D. M., & Ellison, D. A. (2006). Rhabdomyosarcomas in adults and children: an update. *Archives of pathology & laboratory medicine*, *130*(10), 1454-1465.
6. Van Rijn, R. R., Wilde, J. C., Bras, J., Oldenburger, F., McHugh, K. M., & Merks, J. H. (2008). Imaging findings in noncraniofacial childhood rhabdomyosarcoma. *Pediatric radiology*, *38*, 617-634.
7. Agrons, G. A., Wagner, B. J., Lonergan, G. J., Dickey, G. E., & Kaufman, M. S. (1997). From the archives of the AFIP. Genitourinary rhabdomyosarcoma in children: radiologic-pathologic correlation. *Radiographics*, *17*(4), 919-937.
8. Tannous, W. N., Azouz, E. M., Homsy, Y. L., Kiruluta, H. G., & Grattan-Smith, D. (1989). CT and ultrasound imaging of pelvic rhabdomyosarcoma in children: a review of 56 patients. *Pediatric radiology*, *19*, 530-534.
9. Maurer, H. M., & Ragab, A. (1991). Rhadbomyosarcoma. In Fernbach, D. J., Vietti, T. J., editors. Clinical pediatric oncology, 4th ed. St. Louis, MO: Mosby-Year Book, p 491–515.
10. Fletcher, B. D., & Kaste, S. C. (1992). Magnetic resonance imaging for diagnosis and follow-up of genitourinary, pelvic, and perineal rhabdomyosarcoma. *Urologic radiology*, *14*, 263-272.
11. Pai, D. R., & Ladino-Torres, M. F. (2013). Magnetic resonance imaging of pediatric pelvic masses. *Magnetic Resonance Imaging Clinics*, *21*(4), 751-772.
12. Shah, R. U., Lawrence, C., Fickenscher, K. A., Shao, L., & Lowe, L. H. (2011). Imaging of pediatric pelvic neoplasms. *Radiologic Clinics*, *49*(4), 729-748.
13. Huayanay Espinoza, J. L., Espinoza Figueroa, J. V., Velezmoro Díaz, V. E., Huanca Amesquita, L. S., Cruz Baca, R. C., Rioja Vega, M. A., & Guelfguat, M. (2024). Soft-Tissue Sarcomas of the Genitourinary Tract with Radiologic-Pathologic Correlation. *RadioGraphics*, *44*(2), e230138.
14. Jagannathan, J. P., Tirumani, S. H., & Ramaiya, N. H. (2016). Imaging in soft tissue sarcomas: current updates. *Surgical Oncology Clinics*, *25*(4), 645-675.
15. Staatz, G., Honnef, D., Piroth, W., & Radkow, T. (2008). Pediatric Imaging: Direct Diagnosis in Radiology. 1st ed. Thieme; 2008.
16. Idowu, B. M., & Abidoye, I. A. (2023). Botryoides Sarcoma of the Urinary Bladder: Case Report, 104-110.
17. McDowell, H. P. (2003). Update on childhood rhabdomyosarcoma. *Archives of disease in childhood*, *88*(4), 354-357.
18. Gruessner, S. E., Omwandho, C. O., Dreyer, T., Blütters-Sawatzki, R., Reiter, A., Tinneberg, H. R., & Bohle, R. M. (2004). Management of stage I cervical sarcoma botryoides in childhood and adolescence. *European journal of pediatrics*, *163*, 452-456.