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CASE REPORT

Sarcoma Botryoides of the Bladder in a Nigerian Child: A Case Report

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Summary

Sarcoma botryoides of the bladder is a variant of the embryonal form of rhabdomyosarcoma that is characterized by protrusion of tumour cells and stroma into a body cavity like a bunch of grapes. Apart from the bladder, the embryonal form of this tumour can occur in other areas such as the vagina, nasopharynx, middle ear and common bile duct. One of its complications is obstruction of the bladder outlet, presenting as acute urinary retention and acute kidney injury. Pressure on the adjacent tissues and organs is also well recognized. It has an intermediate prognosis which depends on age, site and the extent of disease at diagnosis. We report a case of sarcoma botryoides in a 3-year-old Nigerian girl, confirmed by imaging and histology.

Key words: Acute Kidney Injury, Bladder, Embryonal tumour, Rhabdomyosarcoma, Sarcoma botryoides.

Introduction

Sarcoma botryoides is a unique variant of embryonal rhabdomyosarcoma, a rare tumour that occurs primarily in the urogenital region of infants and children. ^[1] In general, rhabdomyosarcoma accounts for 4.5% of all childhood cancer cases. ^[2] The term 'botryoid' in Greek means a bunch of grapes, which typically defines the clinical appearance of the tumour. ^[3] Sarcoma botryoides of the bladder is rare; hence most of the existing knowledge is based on case reports. It most likely originates from the

Wolffian duct. ^[3] It arises in the submucosal region of the bladder trigone and internal meatus and usually manifests as an expanding mass that may grow large before causing symptoms. It proliferates and tends to remain limited to the bladder, urethra and ureters without extensive infiltration until late in the disease. ^[3] Distant metastases to the regional lymph nodes, liver and lungs also occur late. ^[3, 4] Sarcoma botryoides of the bladder in children has been known for more than a century. The first published case report was in 1907. However, prior to that, three suspected cases were identified without

histologic confirmation was not done. [4] The clinical presentation mainly includes abdominal mass, straining to void, urinary retention, features of urinary tract infection and gross haematuria. [3] The diagnosis can be made by cystoscopy and histology [1], while treatment modalities include surgery and chemotherapy. [4] Although sarcoma botryoides in children can be cured, the recurrence rate is high. [3, 4] Mortality occurs mainly from complications such as uraemia and sepsis. [2,5] After a careful literature search, very few cases previously reported in recent times were encountered, the most recent being in 2012. [6] Therefore, this report is about a case of sarcoma botryoides of the bladder in a 3-year-old Nigerian girl that eventually died from the complications of the tumour.

Case Description

A 3-year-old girl presented to the Emergency Room with complaints of straining on micturition and defecation of two-months duration, abdominal swelling and fleshy mass at the perineum, which was noticed a week before the presentation. There was associated dysuria, dribbling of urine, and oedema of the face and lower limbs but no history of haematuria, fever, weight loss, or constipation. At presentation, she was conscious, in painful distress, afebrile and with a suprapubic mass, 18-week size, extending towards the umbilicus. The mass was firm, smooth, non-tender and not attached to the overlying skin. A fleshy, grapelike mass was seen protruding from the urethral orifice. Attempts at urethral catheterization were unsuccessful. Serum biochemistry showed elevated urea (70mg/dl), elevated creatinine (2.35mg/dl), and reduced bicarbonate (13.6mEq/L) concentrations. An abdominal ultrasound scan revealed thickened bladder wall with a hypoechoic cystic mass extending from the lateral wall into the bladder lumen with bilateral hydroureter and hydronephrosis.

Paediatric surgical and urology consults were sought, and the child had an emergency urinary diversion by inserting a nephrostomy tube in both kidneys. Facial and lower limb oedema resolved, and serum biochemistry normalized over three weeks of admission. The abdomen's magnetic resonance imaging showed heterogeneously grapelike intravesical soft tissue mass (Figure 1).

Histology also showed sheets of small, stellate, spindle cells with scant cytoplasm and eccentric small oval nuclei, generous amounts of eosinophilic cytoplasm and elongated tails of cytoplasm. The cells were disposed of in hypo and hypercellular patterns within the myxoid stroma. The cambium layer was present consistent with a diagnosis of botryoid embryonal rhabdomyosarcoma (Histopathological slides could not be accessed).

Chemotherapy was commenced on the fourth week of admission using intravenous vincristine, dactinomycin and cyclophosphamide. She developed complications (such as febrile neutropaenia and symptomatic thrombocytopenia) after the second course of chemotherapy, necessitating therapy suspension. Antibiotics and granulocyte colony stimulating factors were administered, and she was also transfused with platelet concentrates. The period between the fourth and seventh week was turbulent as she developed overwhelming sepsis caused by *Actinomyces israelii*, sensitive to meropenem. The antibiotic therapy was thereafter revised, and ceftriaxone was replaced with meropenem. During the seventh week on admission, the suprapubic mass was noticed to have increased in size to about 6cm above the umbilicus with recurrence of facial and leg swelling. The serum biochemistry at that point also showed worsened urea (160.8mg/dl) and creatinine (6.36mg/dl) concentrations.

The child later developed anuria and, consequently, acute pulmonary oedema from

renal failure. Nephrostomy tube insertion for urine diversion was repeated, but the child succumbed to the illness while preparing for urgent haemodialysis.

A written informed consent was obtained to use the child's clinical data in this report.

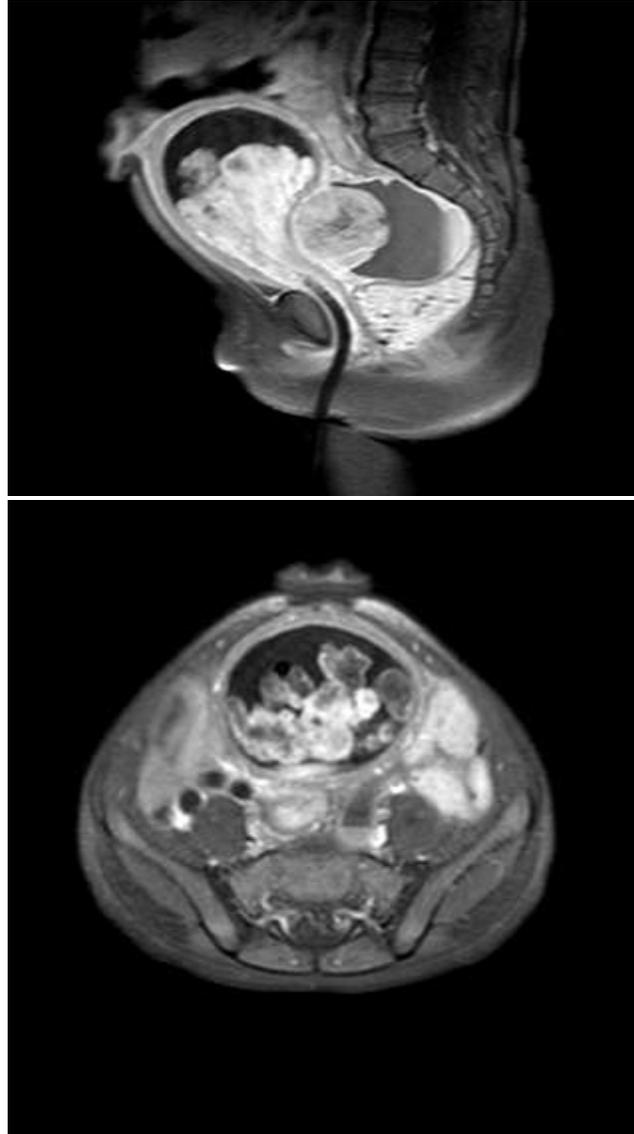


Figure 1: Axial and sagittal T1 Fat suppressed post gadolinium images showing heterogeneously enhancing grapelike intravesical soft tissue masses with extraluminal extension.

Discussion

Sarcoma botryoides has a distinctive feature of being more localized than other malignancies in children. [5] Children younger than four years of age are mostly affected, and males are three times

more affected than females. [3] Sarcoma botryoides accounts for about 10% of all rhabdomyosarcoma cases. [7] The index child presented late with features of urinary tract obstruction and renal failure, which significantly influenced the outcome. This child presented

with abdominal mass, straining on micturition, dysuria, urinary retention and constipation. Other possible modes of presentation include urethral prolapse, fever, urine dribbling and gross haematuria. Obstruction of the urinary tract may occur early, leading to impaired renal function. Prolapse of the 'grapes' may cause bleeding and gross haematuria, and the kidneys may be ballotable from obstruction to the ureters. Usually, the patients do not get cachectic or wasted.

Cystoscopy can diagnose sarcoma botryoides, which shows multiple translucent grapelike polyps arising from the anterior and lateral bladder walls at the bladder neck. [8] Ultrasonography may be inconclusive in diagnosis because the appearance of the mass may be variable. [9] Tissue biopsy may be unhelpful as the tumour usually appears histologically benign. [8] Conventional magnetic resonance imaging (MRI), such as T1 and T2-weighted imaging, is beneficial in diagnosis as it shows the extent of the tumour, composition and local spread. In contrast, multiparametric MRI may show the malignant or benign nature of the tumour. [10] MRI provides superior delineation of tumour size, location, and relationship to other organs, detects residual disease, and is recommended as the first-line imaging modality in patients with suspected sarcoma botryoides. [9] The benefits of MRI over computerized tomography scan include the absence of radiation exposure, a safer contrast agent and better contrast resolution of the tumour to normal tissue or organs. [9]

Radical surgical extirpation of the bladder and the urethra is the initial treatment modality for localized tumours. [5] Urinary diversion is made by ureterosigmoid transplantation. There is a likelihood of total cure after radical surgery if recurrence does not occur a year following surgical intervention. [5] Chemotherapy may be an adjunct treatment. [5] Chemotherapeutic agents

commonly used for other rhabdomyosarcomas, such as cyclophosphamide and dactinomycin, have been advocated, [5] but there is a lack of data on regimens available. Radiotherapy appears to be ineffective in treating the tumour and may stimulate it. [3] Indeed, radiotherapy may only be helpful at a lower dose if there is microscopic residual disease following resection. [3]

Conclusion

Sarcoma botryoides of the bladder is a rare subtype of rhabdomyosarcoma, and its prognosis is often poor due to its rapid growth. Therefore, prompt diagnosis and early intervention are essential for a better prognosis in the paediatric age group. Having structured health insurance would go a long way in encouraging the early presentation of children with malignancies for care.

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