Case Report

BOTRYOIDES SARCOMA OF THE URINARY BLADDER: CASE REPORT

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Abstract

Background: Children are at a higher risk for developing rhabdomyosarcomas than adults. Rhabdomyosarcomas are the most prevalent malignant tumour of the urinary bladder in the paediatric age group. Genitourinary rhabdomyosarcoma is reported seldomly in sub-Saharan African children.

Case presentation: This is a case report of botryoid sarcoma in two boys who were both two-year-old. Recurrent haematuria, crying and straining on micturition, inability to pass urine, and painful lower abdominal swelling/suprapubic mass were the presentations.
One of the boys had been misdiagnosed with posterior urethral valves (PUV) at a peripheral hospital before presenting at our facility. Unfortunately, one child was taken away against medical advice, while the other died of the disease complication.

Conclusions: Imaging plays a vital role in diagnosis, staging, stratification, assessment of response to therapy, and monitoring for recurrence in urinary bladder rhabdomyosarcoma. Physicians need to avoid the potential pitfall of misdiagnosing this tumour for PUV on account of similar clinical presentations.
Keywords: Embryonal Rhabdomyosarcoma, Sarcoma Botryoides, Urinary Bladder Tumour.

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INTRODUCTION

Rhabdomyosarcomas (RMS) are more common in children than in adults.[1] The genitourinary tract is the second most common primary site of RMS after the head and neck in children.[2] Genitourinary RMS may arise from the urinary bladder, prostate gland, vagina, uterus, cervix, urethra, seminal vesicles, ductus deferens, spermatic cord, testes, epididymis, penis and pelvic sidewall.[3,4] RMS is the most common malignant tumour of the urinary bladder in the paediatric age group, constituting 50% of all RMS cases involving the genitourinary tract.[5]

Most genitourinary RMS is of the embryonal histologic type. When they arise from or protrude into hollow organs such as the urinary bladder or vagina, they are called sarcoma botryoides ("bunch of grapes appearance").[6] Its rapid growth and the ensuing local invasion lead to symptoms of bladder outlet obstruction or rectal compression.

Urinary bladder rhabdomyosarcoma is reported seldomly in Nigerian children.[7,8] This report describes the radiological diagnosis of the botryoides variant of this rare disease in two Nigerian children. The parents gave consent to publish these cases.

CASE PRESENTATION

CASE 1

A 2–year old male patient presented with a three-day history of inability to pass urine, crying and straining on micturition, with painful lower abdominal swelling. There was no history of haematuria or dribbling of urine. No

associated vomiting, constipation, or diarrhoea was noted. No history of pyrexia or trauma to the lower abdomen was noted. He had a similar episode of urinary retention about seven months prior to the presentation, which was diagnosed as PUV, at a peripheral hospital. The family, social, and past medical histories were non-contributory.

On general examination he was irritable, not pale nor cyanosed, and apyretic with normal hydration status. Abdominal examination demonstrated a tender suprapubic swelling with no definitive palpable mass or organomegaly. The external genitalia was normal. The rectum was empty on digital rectal examination, but an anterior mass was felt bulging into the rectum. The chest, cardiovascular, and central nervous system examinations were normal. The clinical diagnosis was bladder outlet obstruction secondary to possible pelvic/urinary bladder mass.

The serum urea was 11 mmol/L (Normal = 2.5 – 5.8 mmol/L), but creatinine, potassium, and other blood electrolytes were within normal limits. Urinalysis showed significant proteinuria. Urine microscopy culture and sensitivity identified Klebsiella species. Full blood count showed leucocytosis with neutrophilia; haemoglobin concentration was normal.

Abdominal ultrasound (USS) demonstrated a polypoid, immobile mass with a heterogeneous echopattern appearing to arise from the trigone of the bladder and protruding into the urinary bladder **(Fig. 1)**; measuring 7.2 x 4.2 x 4.8 cm (L x AP x TS). The bladder wall was thickened (measured 5.1 mm). The urine within the bladder contained numerous mobile, low-level internal echoes suggestive of infection and/or haemorrhage. The mass showed minimal vascularity on Doppler insonation, with grade III bilateral hydronephrosis **(Fig. 2).** There were no features of ascites or organ metastasis.

MRI of the abdomen revealed a hypointense lesion of the bladder on the T1-weighted image **(Fig. 3A)** and heterogeneously hyperintense **(Fig. 3B)** on the T2-weighted image with extrinsic compression of the rectum and loss of the cleavage plane between the bladder lesion and the rectum (rectal invasion). There was bilateral hydronephrosis but no ascites, loco-regional nodal disease nor distant metastasis.

Urethrocystoscopy showed multiple polypoid masses at the neck of the urinary bladder, thickened bladder wall and foul-smelling urine; the urethra was normal with no posterior urethral valves. Histopathology examination of the biopsy sample taken at cystoscopy showed features of botryoid embryonal rhabdomyosarcoma. Histologic examination showed elongated spindle-shaped tumour cells, with cross-striations and increased mitotic activity, embedded in a myxomatous stroma. A cambrium layer, a subepithelial condensed layer of tumour cells that is separated from the overlying epithelium by the loose myxoid stroma, was present. The presence of a cambrium layer is the hallmark of botryoid rhabdomyosarcoma.

Unfortunately, the patient's family refused further medical intervention and left against medical advice.

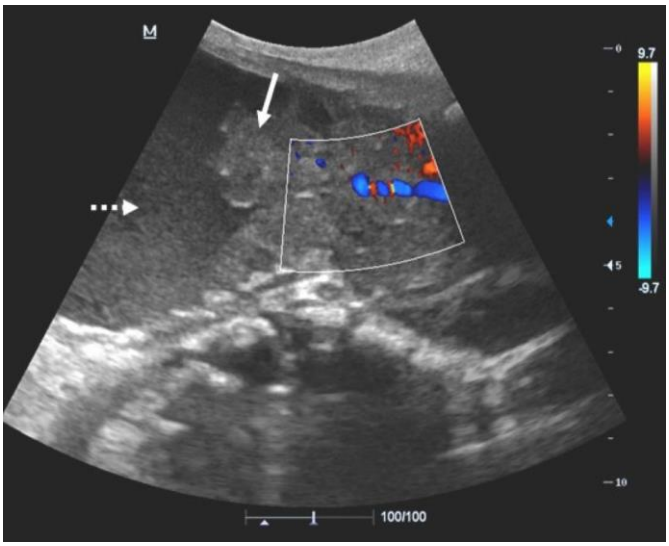

Fig. 1: Longitudinal duplex ultrasound of the urinary bladder showing an irregular, heterogeneous, polypoid, vascular mass (solid arrow) projecting into the urinary bladder lumen from the region of the trigone; Mobile low-level echoes noted within the bladder (dashed arrow)

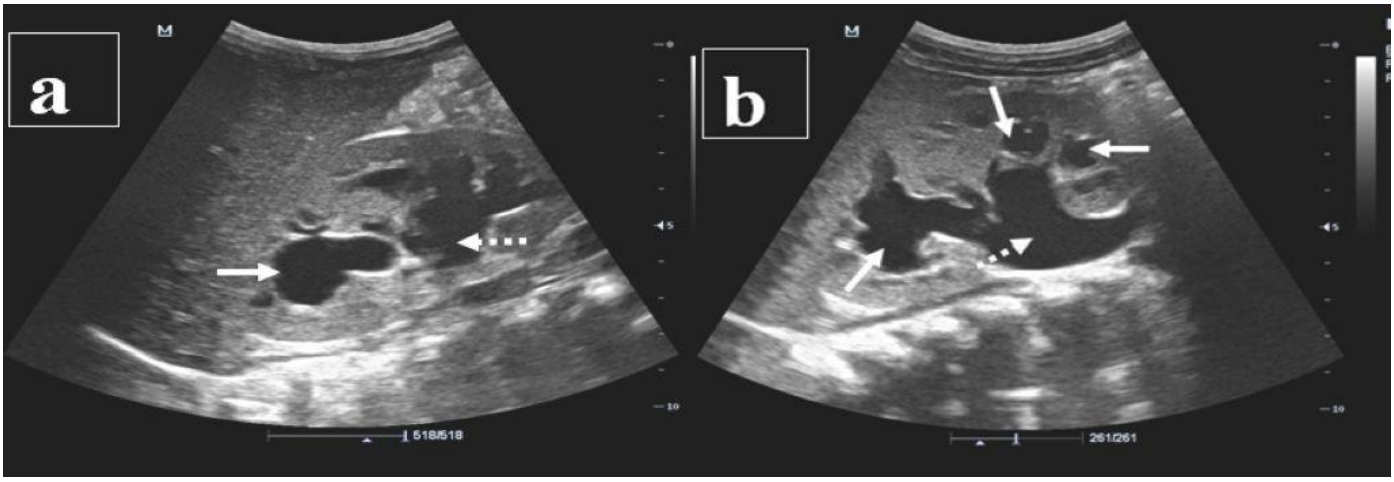

Fig. 2: Longitudinal ultrasound of the right (a) and left (b) kidneys demonstrating marked dilatation of the calyces (solid arrows) and renal pelves (dashed arrows)

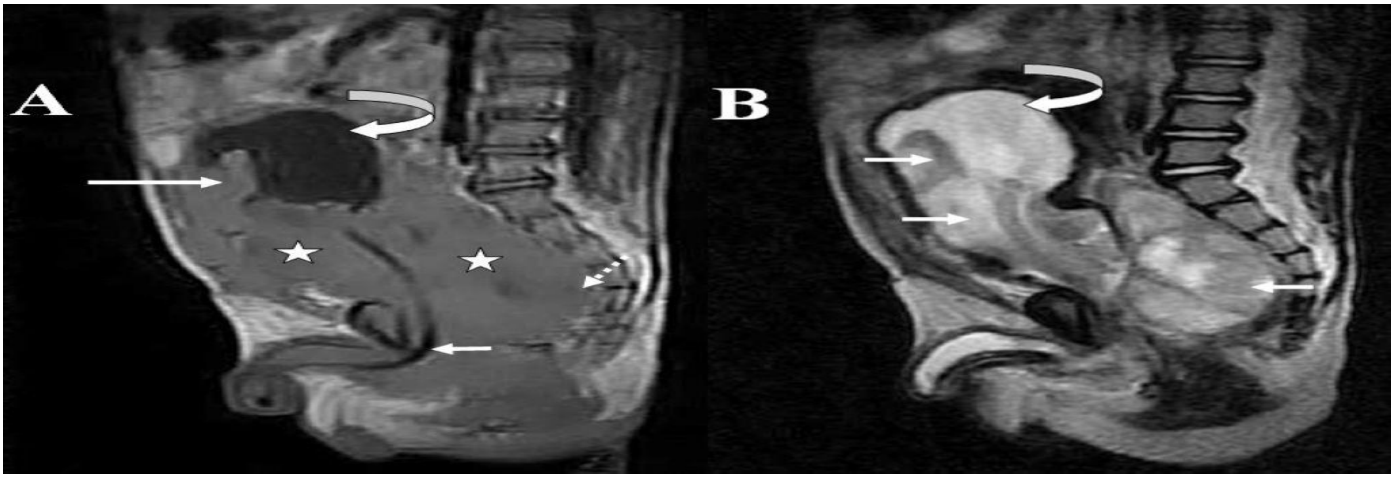

Fig. 3: Sagittal T1weighted sequence (A) and Sagittal T2 weighted sequence (B) of the pelvis showing a hypointense mass (stars) with a polypoid projection into the urinary bladder lumen (long solid arrow) and posterior bulge into the rectum (dashed arrow). The curved solid arrows indicate retained urine within the bladder

CASE 2

A 2-year-old male patient presented with difficulty with micturition, recurrent haematuria, terminal dribbling of urine and suprapubic mass of three months duration. There was no history of reduction in urinary output, hiccups, frothiness of urine, skin rash or fever. 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 general examination was normal. On abdominal examination, a suprapubic mass was palpated, measuring approximately 7 cm x 5 cm, and was non-tender, firm, and smooth-surfaced with no differential warmth. There was no hepatosplenomegaly nor palpable kidneys. A clinical diagnosis of bladder outlet obstruction possibly secondary to a urinary bladder tumour, was made.

The full blood counts were within normal limits. Urinalysis showed proteinuria of 2 pluses. Urine microscopy showed numerous pus cells. The electrolyte, urea and creatinine were all normal. Cystoscopy was attempted but was not successful.

Abdominopelvic ultrasound (**Fig. 4**) revealed a heterogeneously minimally hyperechoic soft tissue mass with a lobulated outline arising from the bladder base measuring6.47 cm x 5.69cm x 4.20cm (L x AP x T). No significant vascular uptake was demonstrated on colour

Doppler interrogation. Bilateral obstructive uropathy was noted.

An Intravenous Urogram (**Fig. 5**) demonstrated a large filling defect within the urinary bladder, appearing to arise from the bladder base. A right-sided duplex collecting system was noted, with bilateral hydronephrosis.

A radiological assessment of bladder outlet obstruction secondary to a bladder mass, possibly a rhabdomyosarcoma was made. There was no evidence of distant metastases on further imaging. The patient underwent urinary bladder 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). Post-operatively, the patient developed acute renal failure and subsequently demised on the third post-operative day.

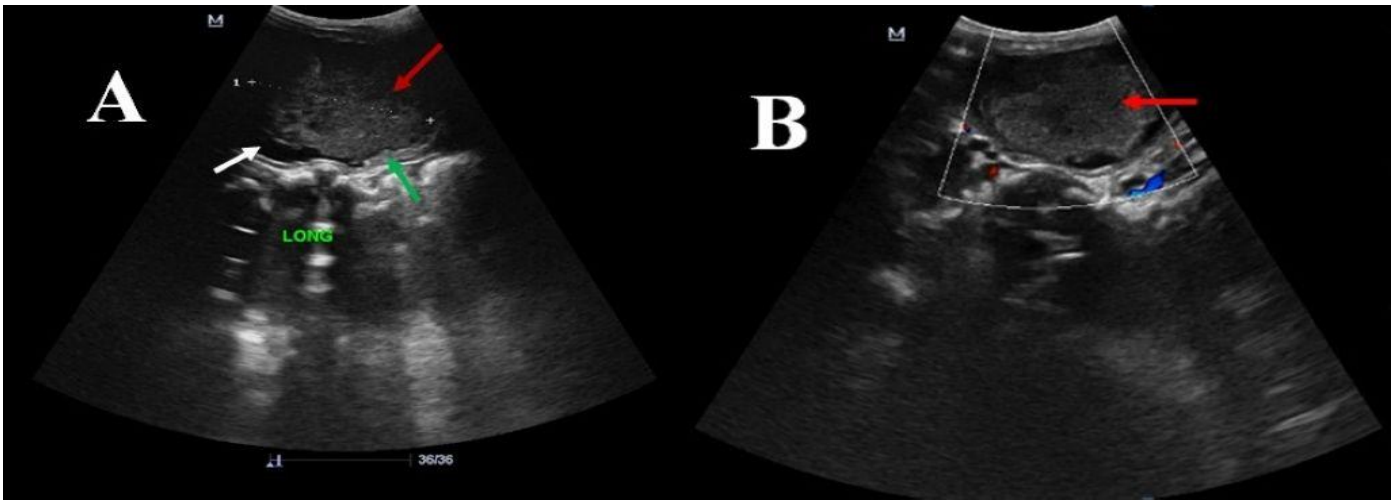

Fig. 4: Pelvic ultrasound, longitudinal (A) and transverse (B) views, showing a huge, lobulated, minimally hyperechoic mass arising from the bladder base (red arrow) in keeping with a rhabdomyosarcoma. The bladder wall is thickened (green arrow). No significant vascularity on Doppler insonation (B).

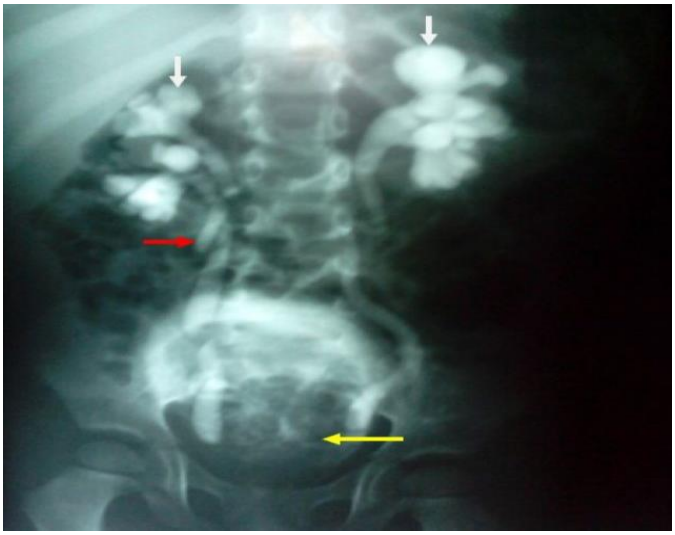

Fig. 5: An intravenous urogram (KUB phase) showing a large filling defect arising from the bladder base (yellow arrow). Right sided duplex collecting system (red arrow). Bilateral hydronephrosis noted (white arrows).

DISCUSSION

Three percent of all childhood cancers are rhabdomyosarcomas. Urinary bladder RMS is a malignant mesenchymal (non-epithelial) tumour arising from the submucosal region of the trigone, and occasionally from the dome of the bladder.[6]

Genitourinary RMS constitute 15 to 30% of all paediatric RMS with 0.5 - 0.7 cases per million yearly. Genitourinary RMS has a bimodal peak age of incidence: 2-6 years and 14-19 years.[9] Generally, the disease affects more males than females, with a male to female ratio of 2–4:1.[3,10–12] African Americans are affected four times as often as whites.[11]

The aetiology of urinary bladder RMS is unknown.[4] Embryonal, alveolar, and pleomorphic variants are the major histological types of RMS. The embryonal type constitutes 50-60 % of cases.[13] The embryonal type was confirmed histologically in both index cases. The tumour

often goes unnoticed until its mass effect causes urinary and/or excretory symptoms.[5] Most patients present with bladder outlet obstruction, which causes abdominal pain and distension, constipation, dysuria, frequency, and features of urinary tract infection.[2] The botryoides form may present as a palpable suprapubic mass.[6] Painless gross haematuria is unusual.[2] Flank pain secondary to hydronephrosis may also occur.[6] Symptoms of bladder outlet obstruction/acute urinary retention, abdominal pain and distension, suprapubic swelling, and evidence of urinary tract infection were present in the index patient.

The roles of imaging include diagnosis, staging, detection of complications, and follow-up (monitoring response to treatment). Ultrasonography (USS) is the usual imaging method for making an initial diagnosis.[9] Bladder RMS are often seen as solid, lobulated mass at the bladder base (trigone) or fundus.[3] They appear as hyperechoic, exophytic/pedunculated/polypoid soft tissue mass extending into the vesical cavity (botryoid or "bunch of grapes" appearance), or as focal wall thickening.[3,9] Polypoid lesions often contain cystic areas due to haemorrhage and/or necrosis. Infiltrating tumours produce irregular, nodular wall thickening[9], which may be isoechoic to the bladder wall. Bladder wall deformity and rigidity, with reduced bladder capacity (contracted bladder), may be seen with deep pelvic invasion. Hydro(uretero)nephrosis and invasion of perivesical structures also can be noted when the mass obstructs the ureters.[4,9]

CT and MRI are required for evaluating and staging the extent of the disease and detecting the presence of metastases.[14] MRI is generally preferred to CT for its better soft tissue resolution and avoidance of ionising radiation in children.[4]

MRI is the gold standard for imaging pelvic RMS because of its excellent soft tissue resolution.[14] Similar to other soft-tissue tumours, RMS are hypo-/isointense on T1W images. On T2-W images, they tend to be iso-/hyperintense. Heterogeneous signal intensity representing haemorrhage in various stages of evolution may also be identified.[2] A pseudocapsule may also be seen.[3] RMS generally enhances heterogeneously with gadolinium.[2] The tumour can be mostly cystic in very rare situations. Dynamic series is helpful in assessing the vascularity of the tumour and differentiating residual disease from fibrosis after chemotherapy.[4,14]

On intravenous urogram (IVU) or MCUG, bladder RMS appears as a large, irregular, and often lobulated filling defect in the bladder. This classic sarcoma botryoid (grapelike) pattern is seen in 25% of cases.[11] IVU is still valuable in centres where cross-sectional imaging is not available.

Urinary bladder RMS may create a confusing clinical and imaging picture sometimes, with documented reports of misdiagnosis for PUV[7], which possibly was what happened at the peripheral centre where the case 1 patient presented with acute urinary retention.

Complications of the disease include tumour rupture, invasion of adjacent structures, and metastases to the lungs, cortical bones, and lymph nodes.[2,3] Liver and bone marrow metastases are less frequent.[2] Metastatic disease is present in 10-20% of all patients with RMS at the time of diagnosis.[2] In the index cases presented, there was no evidence of distant metastases. However, the tumour had invaded the rectum in case 1.

Once rhabdomyosarcoma is diagnosed, the pre-treatment TNM staging and post-biopsy and resection clinical grouping systems developed by the Intergroup Rhabdomyosarcoma Study Group (IRSG), which is now a part of the Children's Oncology Group Soft-Tissue Sarcoma Committee (COGSSC), are used to stratify the risk of rhabdomyosarcoma (risk stratification).[6,14] IRSG staging system assesses the tumour size (≤5 cm or >5 cm), invasiveness (if it is localised to the anatomic site of origin), nodal status (whether regional nodes are implicated), and site of origin (the bladder and prostate gland are unfavourable sites). Data from both IRSG and COGSSC grouping systems are used to assign the patient a risk level (low, intermediate, or high).[14] Risk stratification allows for customised patient-specific treatment plans for selected patients with the benefits of avoidance of irradiation in selected low risk patients and chemo-radiotherapeutic dosage reduction.[15]

The aim of treatment is complete tumour removal, as a common risk factor for recurrence is the residual soft tissue mass at the primary site.[4] Initial chemotherapy, followed by CT and MRI re-staging, cystoscopy and biopsy are all part of the treatment. Radiotherapy is

initiated if there's a good response to the aforementioned treatment; otherwise, extirpative surgery (cystectomy) is done.[4,5]

Cystectomy is no longer the initial procedure of choice, but reserved for those not responding to initial chemotherapy or those relapsing after initial success.[5] The 3-year survival rate after chemotherapy is 60–90%, while the 5-year survival rate after radical surgery is 14-35%.[3] Definitive treatment had not been instituted in the index cases before case 1 left against medical advice and case 2 died.

Vesical and prostatic RMS tend to recur at about 2 to 3 years after the start of treatment.[5] In general, the embryonal botryoid variants have a better prognosis than tumours with alveolar histology.[4,5] Metastatic disease is associated with a very poor prognosis (survival rate < 30% at 5 years).[4] For prognosis, the primary source site is also significant, with bladder or prostate tumours having a worse prognosis than the other genitourinary areas.[4,14]

CONCLUSION

In conclusion, imaging plays a vital role in diagnosis, staging, stratification, assessment of response to therapy, and in monitoring for recurrence. MRI and USS are the imaging methods of choice. It is also essential for doctors to avoid the potential pitfall of misdiagnosing this tumour for PUV on account of similar clinical presentations.

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