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Presentation and Surgical Management of Ischemic Priapism: A 5-Year Review

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ABSTRACT

Objectives: To review the clinical presentation and surgical interventions carried out on patients with ischemic priapism at a tertiary health facility in north central Nigeria.

Patients and Methods: Seventeen patients (median age 17 years, range 4-37) who were managed surgically and reviewed over a 5 year period. All patients had undergone distal shunt procedures and the outcomes on erectile function were examined in those who were sexually active.

Results: Most of the patients presented after 24 hours or more (>24 hours) following the onset of priapism. Sickle Cell Disease constituted 85% of all cases and we noted a bimodal age pattern amongst them (<12 years and >18 years). Intermittent penile massaging was employed after the shunt procedure to maintain detumescence in 12 patients after 24 hours of surgery. Twenty-five percent and 37% of sexually active patients maintained normal and partial erectile function respectively after surgical intervention regardless of delay in presentation.

Conclusion: This review highlights the fact that late and delayed presentation of priapism is still common in our environment and sickle cell disease remains a major predisposition. Appropriate surgical intervention using shunt procedures no matter how late, has the potential to preserve erectile function in some patients.

KEYWORDS: Priapism; Sickle cell disease; Shunts; Detumesence.

INTRODUCTION

Priapism is a persistent, usually painful, erection that lasts for more than four hours, occurs without sexual stimulation and is unrelieved by ejaculation. It was first described in 1845 by Tripe, as it is a relatively uncommon occurrence and as a result, there is scant contemporary literature on standardized treatment of males who experienced priapism. Two types of priapism, namely the ischemic priapism (also known as low-flow priapism) and non-ischemic priapism (or high flow priapism) have been well described. They both have specific and different causes and their management also vary from each other. The causative mechanisms even though not clearly understood, involve complex neurovascular factors. It may be associated with the use of intracavernous injections for erectile dysfunction, sickle cell disease (SCD), recreational drugs (cocaine and marijuana abuse), antipsychotic therapy, blood cancers like leukemia and perineal trauma. Adeyoju et al reported a high incidence (35%) of priapism among patients with sickle cell anemia which they identified as a common aetiological factor.

Badmus in an earlier study, also noted SCD as the commonest cause of priapism in southwestern Nigeria, whereas Aghaji in his series reported local aphrodisiac as the commonest predisposing factor of priapism in Eastern Nigeria followed by SCD. Whereas, use of intracavernous therapy is a common aetiological factor in the advanced countries, it is certainly rare in the poor resource settings like ours due to cost constraint and scarcity. Although, not all patients require immediate elaborate treatment, it is imperative to differentiate
those requiring urgent intervention and offer such accordingly.7 Surgical shunts try to correct the priapic state by literally diverting blood from the rigid corpora cavernosa into the corpus spongiosum. Late presentation to the urologist is common and could be the result of ignorance both on the part of the patient with priapism and some healthcare providers. We carried out this review to determine the aetiology, pattern of presentation and outcome of surgical interventions for priapism in our setting. It is our hope that this will help to address some of the challenges occasioned by the management of this condition in developing countries like ours, remind us of the various surgical options and outcomes of treatment, and to increase public awareness to avert late presentation and hence mitigate the sequalae of erectile dysfunction associated with priapism.

PATIENTS AND METHODS

A retrospective analysis of data was extracted from patient’s case notes and operation register of the University Of Abuja Teaching Hospital from 2008 to 2013. Demographic information on age, duration of symptoms and predisposing factors were documented. Following full clinical examination the patient’s blood and urine samples were taken for full blood count, genotype and urinalysis. Broad spectrum antibiotics were routinely commenced and patients were prepared for surgery. Three common types of distal shunt procedures via winters, Ebbehj and Al-Ghorab were carried out based on the surgeon’s discretion.

We improvised with a size 16 guage intravenous cannular in creating a winter’s shunt through the glans and then the corporal cavernosa. Ebbehj shunt was performed with the use of a size 11 blade pierced longitudinal through the dorsal glans skin along the direction of the two corpus rods and turned 90 degree to create the shunt. The outcome of erectile function, where applicable was qualified as able to achieve penetrative intercourse (normal ), erections not able to penetrate (partial) and no erections ( complete). No standardized method of assessment of erectile function was used.

RESULTS

A total number of 17 patients (n=17) who had distal shunt procedures over the 5-year period were reviewed. The age range was 4-37 years with a median of 17 years (IQR 22.3). However, the median age among patients who had priapism attributable to sickle cell disease was 11 years (IQR 12.9) and a bimodal age distribution pattern (children 0-12 years and adolescents >18years) was obvious among the sickle cell disease patients (Figure 3).

DISCUSSION

This review is amongst few reported series of priapism in our environment highlighting important issues regarding the mode of presentation and surgical management. We have assessed the outcome of procedures carried out in the treatment of priapism in our setting. Delay in presentation is common in our series as the median duration of symptoms prior to presentation is 24 hours. We have deduced that in our setting most patients are likely to present after 17-72 hours after the commencement of symptoms (Figure 1). Prior authors in this region have also reported late presentation of cases.2,3,5 This current study corroborates previous studies that sickle cell disease (Figure 2) is the most prevalent comorbidity (82%) associated with ischemic priapism.3,5 We have found a bimodal age distribution (Figure 1) among these subset of patients with peaks in the age ranges below 12 and above 18 years of age. The patient who had priapism following an overdose of cloxapine is on treatment for schizophrenia by the behavioral physician. Substance abuse with marijuana and leukemia are well-documented pre-disposing factors to ischemic priapism.2,3,6,9

![Figure 1: Boxplot Showing Median Duration of Symptoms Presentation to Intervention (24 hours) and the 95.1% Confidence Interval (17 to 72 hours). This Suggests that the Optimal Duration of Symptoms Prior to Presentation in our Environment (17 to 72 hours) is way beyond the Time before Onset of Damage Commences to the Corporal Tissues.](image-url)
Following distal shunt procedures, all our patients have achieved full detumescence, however, post-operative intermittent massaging was required in those with recurrent tumescence after the surgeries in 71% of cases (Figure 4).

Priapism has been reported to carry a high risk (<35%) of erectile dysfunction. Twenty-five percent of those who were sexually active before surgery reported normal erection adequate for penetrative intercourse despite delay in presentation (Figure 5). According to the suggestion of Aghaji and Badmus which state that despite the late presentation, vigorous surgical intervention should not be denied by the patients. Thirty seven point five percent had some erection inadequate for penetrative intercourse and another 37.5% had severe erectile dysfunction (Figure 5). Comparing the type of shunt procedure against the outcome on erectile function regardless of duration of symptoms, the winter shunt had the most likelihood of preservation of erectile function unlike the Ebbehoj and Al-Ghorab shunts (Figure 6).

This series highlights the fact that late and delayed presentation of priapism is still rife in our environment and sickle cell disease remains a chief predisposing condition. Offering appropriate surgical intervention by the different way of shunt procedures, no matter how late, has the potential to preserve erectile function in some patients.
LIMITATIONS OF THE STUDY

Our series has a small number of cases and due to poor follow-up attitude in our environment most of the patients were lost to follow-up thus limiting our duration of follow up to 3 months. Also a more objective method for assessing erectile function which is desirable could have been used but being a retrospective study we could only categorize the cases subjectively into full, partial and no erections.

CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

REFERENCES


Case Report

Congenital Penile Curvature in a Young Man: A Case Report and Review of Literature

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ABSTRACT

A varied level of penile deformation characterizes congenital penile curvature. It may to some extent affect the quality of the patient’s sex life. We report a case of congenital penile curvature diagnosed at adult age. A corrective surgery was performed by plication of the tunica albuginea of the cavernous body. A review of the literature underscores the rarity of publication on this condition and the treatment modalities.

KEYWORDS: Congenital penile curvature; Penileplication; Corpora cavernosa; Tunica albuginea.

INTRODUCTION

Congenital penile curvature is rare and affects 37 in 100,000 men.1 The prevalence of such condition, without hypospadias, épispadias or spongiosal hypoplasia rates of 4-10%.2 Very few cases have been reported in Africa. Several reconstructive techniques of surgery have been described and performed, including Nesbit3 operation or its variants pertaining to tunica albuginea plication. We present one case report of congenital penile curvature treated by simple plication of the corpora cavernosa.

CASE REPORT

This case report is based on a 24-year-old, married, father of a child, who complained of erectile dysfunction leading to difficulties in vaginal intromission due to a deviation of the penis. Physical examination showed a right lateral deviation of the penis and no palpable induration and plaques. A systematic intracavernous injection of Alprostadil 20 mg (erection test) determined the direction, the degree of curvature that was lateral right, at an angle of 85° with a slight clockwise rotation of 20° (Figure 1). The patient was suggested surgical treatment for penis stiffening and surgery began after his approval of the same. A tunica albuginea plication of the left corpus cavernosum on its lateral face, at the 1/3 proximal union 1/3 medium and the 1/3 distal union 1/3 medium, was made with non-absorbable single-strand 2/0 according to the technique described by Essed and Schroeder (Figure 2). The surgery performed was a simple procedure that lasted 32 minutes. Reviewed post-operatively on a regular basis until the 18th month, the patient was satisfied with the quality of his intercourse since the intervention and the score recorded on the basis of International Index of Erectile Function (IIEF) was 26. However, the patient reported a slight discomfort at the level of the plication area and a slight penile shortening subjectively.
DISCUSSION

Congenital penile curvature is most commonly reported in children and is usually associated with other penile anomalies. Penile curvature manifests itself more often in adolescents or young adults, at the time of puberty or first sexual intercourse. Isolated curves are rare: the incidence was estimated at 0.6% in a study conducted on 500 newborns in Israel. Congenital penile curvatures are manifested as a deviation of the penis during erection. The direction of penile curvature may be dorsal, ventral or lateral, left, or right. Most ventral curves are associated with hypospadias. The etiology is often determinate. The surgical treatment indication of the congenital penile curvature depends on several factors such:

- The degree of curvature, the threshold of 30° is classically admitted but a threshold of 20° can be retained;
- The orientation of the curve, the dorsal curves are better tolerated;
- The rotation of the curve;
- Psychological and sexual repercussions.

The procedure of surgical target the convexity of the albuginea of the corpora cavernosa. This is sometimes called “plastics of the corpora cavernosa”. Nesbit technique was the first method to be described in 1965 in which one or more excisions were introduced in the ellipse at the level of the albuginea, on the opposite side of the curvature. Many variations of methods derived from Nesbit technique have been proposed. These include methods of incision, but without excision or methods of plication. Tunica albuginea plication of corpus cavernosum was described in 1985 by Essed and Schroeder. One of the objectives of this functional surgery was to restore a straight and natural erection, to perform minimal surgical intervention so as to avoid possible complications or aggravating the previous condition. This technique is applied directly on the corpora cavernosa at the level of the convexity of the curvature; Wide reversing dots with single strand non-absorbable wire produce a plication. An artificial erection at the end of the procedure validates success of the surgical correction. No resection of albuginea is practiced. The procedure is performed for 20-30 minutes for which the patient needs to be hospitalized for 36-48 hours. The same principle had been used by Gholami and Lue and to put 16 knots.

In case of rotation of the corpora cavernosa, the technique of Shaer which consists of making a longitudinal incision on each corpora cavernosa and 2 lines of internal and external suture by overjet, could be used.

The satisfactory results of our study are consistent with the observations from the published series reporting the various techniques approaching the convex face of the curvature: in which 78 to 91% satisfaction is archived by the technique described by Essed and Schroeder. Gholami reported satisfaction in 96% of the cases and a complete recovery in 93% of the cases. Camerlo operated by introducing simple plication in 8 patients with a congenital penile curvature and obtained results with 100% satisfaction: no residual curvature, no penile shortening, no erectile dysfunction. The complications reported by the authors the procedure penile shortening, recurrence of the curvature, pain during erection, aesthetic sequelae, urethral wound, hematoma of the penis, and suture. Some authors have reported post-operative recurrences: 29.5% of the recurrences presented by Schulteiss reported in 61 patients operated by tunica albuginea plication.
CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

REFERENCES


Case Report

Renal Cancer Mimicking a Renal Cyst: A Case Report

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ABSTRACT

Introduction: Renal cystic diseases are common and are often less aggressive and benign in nature. Watchful waiting is the therapeutic choice for managing simple renal cysts. Some malignant tumors of the kidney may have features of a benign renal cyst. It is challenging to determine how to manage such affections of the kidney. Very few of these cases have been reported in the literature but do not have a standard therapeutic approach. The objective of this case presentation is to discuss useful ways in managing renal cancer presenting as a renal cyst.

Case presentation: A 78-year-old Moroccan male farmer with no significant medical history presented with a 2 year intermittent left flank pain, without macroscopic hematuria nor hydatiduria nor painful urination. Physical examination was unremarkable. Blood analysis especially that of hydatid serology was unremarkable. Abdominal ultrasound showed multiple simple cysts on both kidneys with a suspected left renal cyst that had internal echoes (Figure 1).

Abdominal CT scan showed multiple cysts on both kidneys with the largest of the cyst on the left kidney estimated to be 7x6 cm with slightly thickened wall classified as Bosniak II F or III, the other existing cysts were all classified as Bosniak I (Figures 2 and 3).

The patient underwent radical nephrectomy because of the pain and intra-operative
conditions, (the major cyst was very close to the vessels). Histopathology of the specimen confirmed papillary carcinoma of the kidney, Fürhman grade 2 cystic alterations, classified as pT2aNx.

Staging found no secondary localizations for this tumor. After a follow-up of 10 months the patient asymptomatic without any sign of recidivism.

DISCUSSION

The vast majority of cystic renal masses are benign. However, malignant tumors may have a cystic appearance. They are dominated by multilocular cystic renal cell carcinomas which are usually low grade.3

Clinical presentation for these lesions is not specific, as in the case of our patient who complained of lower back pain. Diagnosis is often made incidentally during radiological assessment performed for another reason.

Morton Bosniak classification (1986)1 has characteristics that distinguish benign cysts (categories I and II) from potential malignant cysts requiring surgical management (classes III and IV). A new category, called IIF (F for follow-up) was added in 19973 for intermediate lesions between types II and III, requiring regular monitoring through imaging. The relation between Bosniak score and the likelihood of malignancy has been widely demonstrated in the literature.3 However, there is no correlation between the Bosniak score and histological type, TNM staging or Fuhrman grade.3 Although, the lesions found in our patient were classified as type 1 and 2F or 3, nephrectomy was performed due to difficulty in preserving the kidney during surgery. Histopathology study of the specimen revealed the presence of cancer for a renal cyst classified Bosniak II. This implies how important it is to carefully assess the characteristics of multifocal Bosniak I and II cysts.

The Bosniak classification can be adapted to magnetic resonance imaging (MRI) which can at least correlate with histopathological findings.3 Predictive factors of neoplasia are similar to that used in CT scans.1

Contrast enhanced ultrasound of the kidney (CEUS) combines an intravenous injection of gas filled microbubbles to traditional ultrasound, this allows a dynamic study in real time of the enhanced lesion. Several authors describe a higher resolution CT scan with improved sensitivity.6
Renal biopsy in cystic tumors of the kidney is a controversial issue. We did not attempt to perform biopsy in our patient. Indications for renal biopsy remains restricted to Bosniak IV cysts with a tissue target clearly visible on imaging. Harisinghani et al report that a combination of biopsy and fine needle aspiration of cyst could prevent 40% of unnecessary surgery for Bosniak III cysts. In a series of 199 biopsies associated with a needle puncture for IIF and III lesions, Lang et al found a positive predictive value of 91% for malignancy and 100% for benign lesions.

Typically, non-symptomatic Bosniak I and II do not warrant treatment or special monitoring.

In case of symptoms related to the cyst, treatment can be proposed even for benign lesions as this was the case in our patient. Accepted treatment forms are percutaneous puncture and sclerotherapy and surgical resection of the prominent dome. We performed nephrectomy as it was technically impossible to preserve the kidney due it’s location.

Bosniak III and IV lesions, as well as evolved Bosniak IIF lesions, should be considered as potentially neoplastic and must undergo surgery by respecting good oncological practice. The choice between conservative surgery and extended surgery depends on the size of the cyst, its location and the patient’s general condition. Various surgical approaches can be used (open surgery, Coelioscopy or robot-assisted surgery) in our case we performed open surgery because our center is not yet equipped very well with laparoscopy.

CONCLUSION

Benign cystic lesions of the kidney may often not be solely benign as we have reported in our case. This implies a very careful interpretation of available imaging modalities especially that of abdominal CT scans for renal cysts as certain malignant features can be objectified. We recommend surgery without hesitation whenever there is doubt with respect to suspected or doubtful renal cystic lesions.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

COMPETING INTEREST

The authors declare that they have no competing interests.

REFERENCES


Free Split-Skin Grafting to Correct Radical Circumcision: An Alternative Solution for an Under-Reported Complication

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ABSTRACT

Circumcision is one of the most common surgical procedures worldwide, and like all other operations, there is a risk of complication. Our patient had an excessive skin excision circumci- sion which led to a symptomatic partially buried penis. A free split skin graft was successfully applied to correct a large circumferential skin defect with relief of symptoms, preservation of function and good cosmetic outcome.

CASE REPORT

An otherwise healthy 11-year-old boy underwent an elective circumcision at the age of five for phimosis with a normal healing. The boy began to develop pain associated with erections at the age of nine. He had presented his concern multiple times to his general practitioner, hospital and urologists for 2 long years. Despite his condition being correctly diagnosed, he was man- aged conservatively due to lack of a well-established intervention.

On examination, the flaccid length of his penis was 2 cm with the skin while under tension. The boy reported that his penis did not increase in length with an erection and this was associated with exquisite pain. The cause of the pain was attributable to the skin of the penis, for not being able to accommodate the penis during the erectile state (Figure 1).

A decision was made to release the skin of the penis, and cover the defect with a split skin graft from the inguinal region. This site was chosen for a donor site as it is hairless, and soft and closely resembling the skin of the penis.

The operation was performed with parent’s consent of the patient and broad-spectrum antibiotics were administered on induction. During the procedure, the skin overlying the shaft was released distally at the site of previous incision from the circumcision, resulting in a 3 cm skin deficit. A carefully measured section of split skin was harvested with a scalpel blade of size 21 from the inguinal region, we didn’t use the dermatome due to the difficulty to make the area flat to use this device. The graft was macerated and grafted onto the penis shaft. The graft was sutured with interrupted 4.0 Biosyn sutures and 4.0 Vicryl Rapide quilting sutures. The graft was dressed under pressure with Bactigras gauze. The skin donor site was closed primarily (Figure 2).
The patient was admitted for 48 hours under observation and analgesia. The dressings were left intact for 48 hours and when the dressing was taken down the graft was healthy and viable. At the time of discharge the graft and donor site were healing well. Community nurses managed daily dressings for a further three weeks. Analgesic requirement was minimal and managed in the community (Figure 3).

On follow-up at 3 weeks, the graft looked healthy, with no areas of breakdown or necrosis. The donor site was also healing well.

On follow-up at 6 weeks the graft had taken well, but there was evidence of keloid scarring at the suture lines of the proximal and distal sections of the skin graft, as well as keloid scarring of the donor site. There was no history to suggest a propensity to form keloid scar. The patient then started on daily applications of hydrocortisone cream to the graft site as well as the donor site (Figure 4).

After following the above mentioned procedures for 6 months, the graft had taken well, the symptomatic painful erections had resolved and the patient was pleased with the result and the overall cosmesis. The keloid scarring responded well to the steroid cream and did not interfere with the erectile function of the penis (Figure 5).

We continued follow-up of this patient for 2 years and the patient did not develop any contracture and the graft grew with the penile growth without producing any deformity. As the patient reaches puberty and the penis has a tendency to growth in size, the patient needs to be reviewed to check if the graft produces any contracture.

DISCUSSION

Male circumcision is a common surgical procedure having been performed on 30% of the worldwide population.\textsuperscript{1,2} It is subject to continue debate with varying public attitudes with some evidence supporting circumcision for hygiene purposes and prophylaxis of penile cancer. As with all surgical procedures, there is a complication rate and depending on the skill and training of the individual performing the circumcision, this can be as high as 15% with the median complication rate being 1.6%.\textsuperscript{3}

While a common procedure, if done incorrectly, it can have long-term, and debilitating complications, affecting sexual function, as well as significantly impacting the mental and emotional state of the individual.
Excessive excision of skin during circumcision results in a buried penis, leaving insufficient skin to accommodate the full length of the penis particularly during an erection. This can result in pain associated with erections, decreased penis length, impaired sexual function and poor aesthetic appearance. There is little data available on the rate of this complication, but the rate of complication is likely very under-reported and under-treated. This case report describes the use of split skin graft to correct a complication of excessive skin excision during circumcision.

There have been numerous techniques described to correct this complication including; healing by secondary intention, dorsal Z-plasty for small skin deficits, and skin re-surfacing. Techniques that have been described to correct this defect are complex and have high rates of failure, complicated and poor cosmetic results. The challenges with providing skin coverage for the penis shaft differ from other parts of the body in that the penis requires the ability to accommodate the elongation and engorgement during erection, as well as cosmesis, and preservation of sensation.

To correct a skin defect from a buried penis, the use of autologous free skin grafts from a hairless and subtle donor site have the benefit of being able to correct a greater defect than Z-plasty or scrotal skin advancement, while preserve the surrounding anatomy and give a greater cosmetic outcome. The use of split skin graft for skin resurfacing of the penis is not a frequently described method in the literature with only two cases reported in the last five years. However, as demonstrated by this case, it is a simple yet effective technique that can be utilised to correct a large skin defect as well as to maintain the function of the skin excised during the circumcision.

The donor site we recommend is the skin above the level of the inguinal region, easy to harvest and good quality skin due to its elasticity, softness and hairless, making this region ideal to replace the skin in the penile shaft with good cosmetic and functional effect. This region can provide grafts big enough to cover the whole shaft of the penis without leaving big and aesthetically scars in the harvesting area. The use of split skin graft decreases and diminishes the scaring and retraction of the graft compared with the full thickness.

This technique can be used in paediatric and adult patients that need a big size graft. We have been using this method in a Burmese patient with an indurated shaft of the penis due to injection of Benzene that covers the full length of the penis. A split skin graft from the groin was used to cover the full length of the penis, from base to glans with a good cosmetic and functional result and with a small lineal scar in the donor site.

CONCLUSION
Radical skin excision during routine circumcision is an under reported complication, one that can have profound effects on the individual including pain, self esteem and sexual function, and mostly left untreated, with a great impact in the individual. This complication can be avoided by taking extra precautions during circumcision. We can see in this case, free split skin grafting is an alternative method for correcting large skin deficit of the penis following excessive skin excision of circumcision, and is suitable for use in children and adults.

CONFLICTS OF INTEREST
The authors declare that they have no conflicts of interest.

CONSENT
Consent has been taken from the patient for purpose of using patient photographs for publication in print or on the internet.

REFERENCES
Rhabdomyosarcoma (RMS) of the Renal Pelvis in a Child: A Case Report

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ABSTRACT

Rhabdomyosarcomas (RMS) of the kidney is extremely rare. We are reporting one case of embryonal rhabdomyosarcoma localized in a child renal pelvis, focusing on the rarity of the site and role of imaging. An 8-years-old boy was admitted to the hospital with abdominal pain and hematuria. Ultrasonography revealed a right ureteropyelectasy including an echogenic image that extended from the renal pelvis towards inferior basin with no hypervascularization, simulating a clot. The urography-magnetic resonance imaging (Uro-MRI) showed two different components of the lesion: the first one simulating a clot; the second one detected in the papillary region and extended to the inferior calices having tumor-like features. A percutaneous ultrasound-guided renal biopsy revealed a botryoid embryonal rhabdomyosarcoma. The RMS is a complex childhood malignancy. This complexity is reflected in the radiological assessment that remains highly challenging and in the absence of treatment guidelines for unusual sites.

KEY WORDS: Rhabdomyosarcoma (RMS); Renal mass; Uro-MRI; Kidney biopsy.

INTRODUCTION

Rhabdomyosarcomas (RMS) are the most common soft tissue sarcomas in childhood and account for approximately 3% to 8% of all malignancies in children and adolescents with an annual incidence of rate between 4 and 5 per million in children younger than 18-year-old. The genitourinary tract is the site of the primary tumor in the 10-26 % of these cases and occurs more often in bladder and pelvic organs.1

These tumors are to be considered highly malignant neoplasms and arise from skeletal muscle progenitor cells (pluripotential mesenchymal cells of connective tissue).2-6 RMS of the kidney is extremely rare.

We report one case of the ERMS localized in renal pelvis in children, focusing on the role played by the imaging techniques and the difficulty of choosing therapy in this unusual site of origin of the tumor, compared with other pediatric cases reported in literature.

CASE REPORT

An 8-year-old boy was admitted to the emergency department of our hospital with a 15-day lasting recurrent abdominal pain, especially located in the right side, and hematuria. No history of lumbar trauma or stones was detected. The general physical examination was normal. Ultrasonography revealed the right ureteropylectasy of the right kidney including a corpusculated and echogenic image that extends from renal pelvis toward inferior basin with no hypervascularization. This image was suspected for a clot (Figure 1). From this image we suspected the presence of a clot. It was always detected with the same features in the other scans performed in the following days. For a better image definition we performed an urography-magnetic resonance imaging (Uro-MRI).
The image confirmed the lesion without vascularization in the right renal pelvis, while the T2 phase showed a mass, with a diameter of 15 mm, arising from the right inferior calices, in contiguity with the pelvic lesion, with intense enhancement after the administration of a contrast agent (Prohance 0.1 ml/kg), especially at the venous phase (Figures 2, 3 and 4). The radiologic assessment excluded metastases. Therefore, a percutaneous ultrasound-guided renal biopsy was performed. Three biopsies revealed a botryoid embryonal rhabdomyosarcoma.

The patient subsequently underwent an open nephroureterectomy (Figure 5) with a fat layer sampling, bone marrow biopsy, and placement of a central venous catheter. We decided to leave intact the bladder wall (and not to perform the ureterectomy until the bladder) because the tumor was completely located inside the kidney. Finally the anatomical pathological examination showed the actual size of the tumor: 5×2.5 cm (Figure 5). Histologically it presented undifferentiated oval cells with poor cytoplasm, hyperchromatic nucleus and irregular nucleolus. In minor components, there were elongated or round cells with eosinophilic cytoplasm, typical for the rhabdomyosarcoma. It was associated with inflammatory component. The mitotic activity was elevated. Immunostains were positive for Desmin, Myogenin, Actin, whereas negative for Cytokeratin, S100 proteins. The ureter, the fat layer and the Gerota’s band were free from disease. No evidence of hilar node involvement was detected. The post-operative course was regular. The infant underwent a chemotherapy according to EpSSG RMS 2005 (standard risk-subgroup B): Ifosfamide, Vincristin, Actinomicyn (13 cycles of chemotherapy). No recurrence or metastases were found after 5 years follow-up.

DISCUSSION

This is a rare case in pediatric population. This tumor can easily mimic other tumors of kidney in pediatric age group, as nephroblastoma or rabdoid tumor.7-8 Therefore, the RMS with renal localization in children, should be considered in the differential diagnosis with other renal tumor of childhood.

The ultrasonography is frequently used as radiological examination of children who were affected by tumors of the soft
tissues, since it is easily executable and well shows for visualization of the vascularization of a tumor.\(^9\) In our case, the ultrasonography examination provided a precise topographic description of the lesion that extends from renal pelvis toward inferior basin, and offered the initial suspicion of a tumor. But only the Uro-MRI revealed that the tumor was divided into two components: the first one protruding into the pelvis without specific radiological features (non-vascularised, located at the right renal pelvis simulating a clot) and the second one (approximately 15 mm long) arising from the papillary region and extended to the inferior calices with radiological characteristics typical of cancer (intense enhancement in the venous phase).

So our case confirms that the Uro-MRI is the gold standard imaging modality in RMS, although the imaging characteristics of RMS are relatively non-specific.\(^{10,11}\) Interestingly, some authors report that distinct renal tumors can correlate with imaging findings mimicking each other (the “claw sign”).\(^{12}\)

The RMS is a complex childhood malignancy with ubiquitous anatomic sites of presentation and varying histological types, each presentation with peculiar patterns of growth, clinical behavior and prognosis.\(^{13}\) This complexity is reflected in the radiological assessment that remains highly challenging. Prognosis is dependent on anatomic primary tumor site, age, completeness of resection, presence and number of metastatic sites, histology and biology of the tumor cells.

No specific treatment guidelines for renal RMS have been established. In our case the pathology, the postsurgical stage, the node stage, the size and age were favourable, but the rare site didn’t ensure absolute safety for low risk. For this reason, the case was classified as a standard risk, subgroups B (EpSSG RMS 2005) and the infant underwent a chemotherapy with Ifosfamide, Vincristin, Actinomicyn (IV A). Few cases are described in the literature.\(^{12,14,15}\)

The most important statistic revealed 6 cases of primary renal embryonal rhabdomyosarcoma (ERMS).\(^{16}\) Senga et al\(^{17}\) reported 15 cases of RMS of the kidney in Japanese literature. Small numbers of patients make it unwise to draw broad conclusions.

In our experience it was very important to perform an initial mass biopsy for a certain typing. Atypical localization of rhabdomyosarcoma should be considered as an unfavorable form. In addition, complete tumor removal and biopsy of contiguous lymph nodes are required in order to establish a radiotherapeutic treatment. The treatment of RMS requires a multidisciplinary approach, in which pediatric oncologists, radiologists, pediatric surgeons and pathologists altogether play a vital role.

**CONFLICTS OF INTEREST**

The authors declare that they have no conflicts of interest.

**REFERENCES**


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