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Paratesticular rhabdomyosarcoma in a 30 months old child at the Lagoon Mother and Child Teaching Hospital (CHUMEL) in Cotonou

Un rhabdomyosarcome paratesticulaire chez un enfant de 30 mois au Centre Hospitalier Universitaire de la Mère et de l'Enfant Lagune (CHUMEL) de Cotonou

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INTRODUCTION: Paratesticular rhabdomyosarcoma is a rare and aggressive tumor. We report here a case in a 30 months old boy. **OBSERVATION:** The parents would have noticed a small, painless and rapidly evolving testicular mass in the six months old child. Not having health insurance, they went to the hospital only five months later. This was followed by a total ablation of the mass (without orchidectomy) and an anatomopathological investigation concluding to a pleiomorphic rhabdomyosarcoma. No chemotherapy was performed and the mass recurred two months later. This time, its ablation was followed by a series of non-adapted chemotherapy sessions with irregular follow-ups leading eight months later to the child's admission at our department. He presented with a degraded general condition, associating an infectious and anemic syndrome, an important increase of the scrotal mass and the occurrence of a tumoral mass in the left iliac fossa. The tumoral nature of these masses was confirmed on abdominal ultrasound and scan. The tumor was classified stage II (AJCC Prognostic stages). The multidisciplinary medical staff indicated a total ablation of the two masses, followed by a new session of adapted chemotherapy, in the absence of radiotherapy means. The child died in the immediate surgery follow-ups due to cardiac arrest. **CONCLUSION:** In our context of insufficient technical facilities, only early detection and adapted imperative chemotherapy, would have enabled a durable remission in front of the paratesticular rhabdomyosarcoma.
KEYWORDS: Tumor; Paratesticular rhabdomyosarcoma.

INTRODUCTION : Le rhabdomyosarcome paratesticulaire est une tumeur rare et agressive. Nous rapportons ici un cas chez un garçon de 30 mois. **OBSERVATION :** Les parents avaient remarqué une petite masse testiculaire indolore et évoluant rapidement chez l'enfant de six mois. N'ayant pas d'assurance maladie, ils ont consulté à l'hôpital seulement cinq mois plus tard. Il s'en est suivi une ablation totale de la masse (sans orchidectomie) et un examen anatomopathologique concluant à un rhabdomyosarcome pléiomorphe. Aucune chimiothérapie n'a été réalisée et la masse a récidivé deux mois plus tard. Cette fois, son ablation a été suivie d'une série de séances de chimiothérapie non adaptées avec des suivis irréguliers aboutissant huit mois plus tard à l'admission de l'enfant dans notre service. Il présentait un état général dégradé, associant un syndrome infectieux et anémique, une augmentation importante de la masse scrotale et la survenue d'une masse tumorale en fosse iliaque gauche. La nature tumorale de ces masses a été confirmée par l'échographie et le scanner abdominaux. La tumeur était classée stade II (AJCC Prognostic stages). La réunion de concertation pluridisciplinaire a indiqué une ablation totale des deux masses, suivie d'une nouvelle séance de chimiothérapie adaptée, en l'absence de moyens de radiothérapie. L'enfant est décédé dans les suites opératoires immédiates en raison d'un arrêt cardiaque. **CONCLUSION :** Dans notre contexte de plateau technique insuffisant, seule une détection précoce et une chimiothérapie impérative adaptée, auraient permis une rémission durable devant le rhabdomyosarcome paratesticulaire.
MOTS-CLES : Tumeur ; Rhabdomyosarcome paratesticulaire.

INTRODUCTION

Rhabdomyosarcoma (RMS) is a malignant mesenchymal tumor characterized by the presence of cells with striated muscle differentiation similar to rhabdomyoblasts. Paratesticular locations account for 7% of all rhabdomyosarcomas in children (1). Embryonal and alveolar histological types are the most frequent in children, the pleiomorphic type exclusively occurring in adults (2,3). We report here a case of pleiomorphic paratesticular rhabdomyosarcoma in a 30 months old boy. Our aim in presenting this case was to emphasize the prognostic value of early diagnosis and adequate multidisciplinary management.

OBSERVATION

HG. was a 30 months old boy admitted for the management of a testicular mass fortuitously discovered by the parents at the

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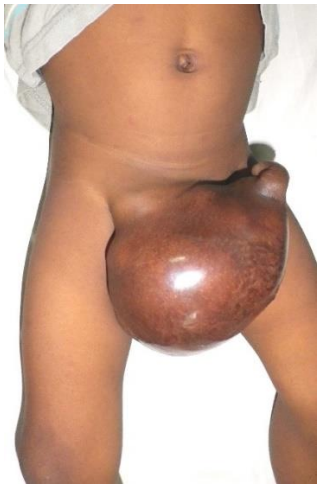
age of six months. It was then a small, painless and rapidly evolving testicular mass. A management had initially been started in a health facility which consisted of a first excision of the mass which would've been total and a second one six months later as the mass recurred. An orchidectomy had not been performed. The anatomopathological investigation concluded to a pleiomorphic rhabdomyosarcoma. He then received seven sessions of unsuitable and unsuccessful chemotherapy (Doxorubicin, Vincristine, Cyclophosphamide). Let's mention that in the pediatric oncology unit, there was a not yet published cohort wherein the RMS proportion was 3.7% with five cases out of 132 children. This was the context in which HG. was admitted at CHUMEL, two years after the disease onset. No particular personal or family history were noted. The patient's initial clinical examination revealed a general condition not significantly degraded, a normal skin and mucous membranes coloration, a good hydration and nutrition status with a weight of 11 kg. There was no infectious or edematous syndrome. The genitourinary examination revealed a shiny oval scrotal mass, 17x13 cm in size, a twisted and infiltrated penis, the presence of a left inguinal scar (Figures 1A and 1B) and a bladder globe.

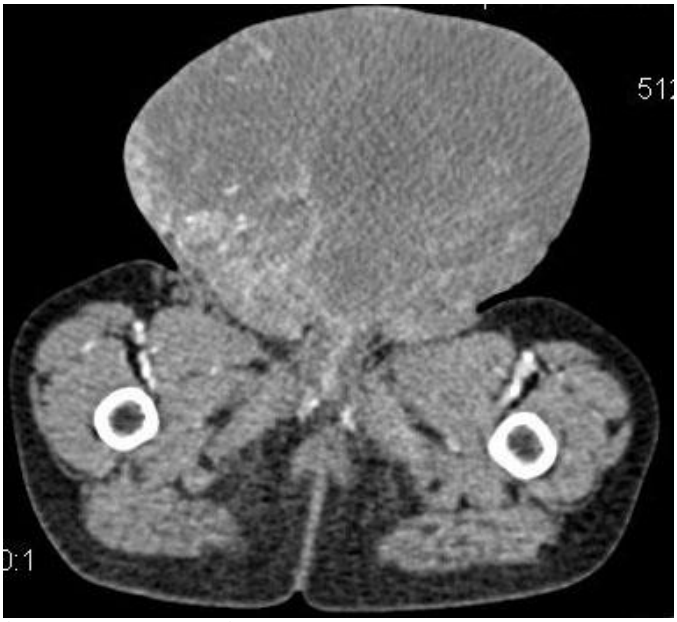


1A: Front view.

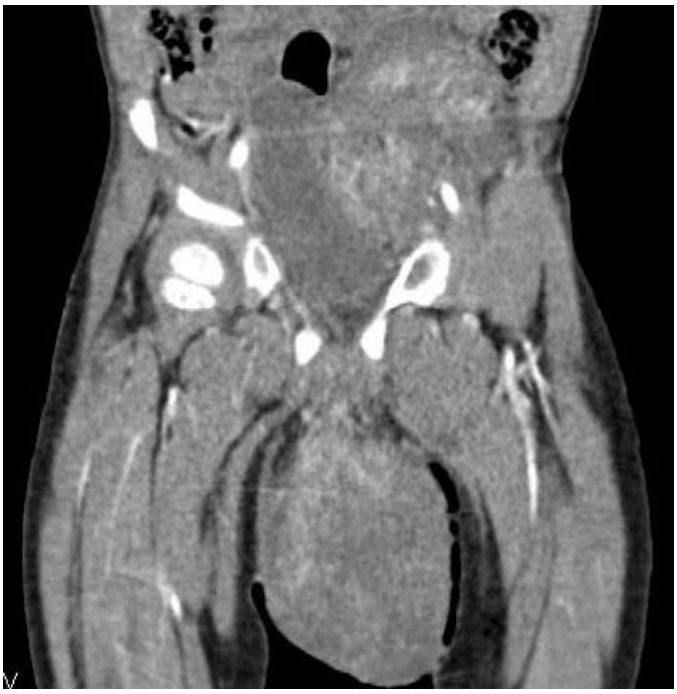
1B: Side view.

Figure 1: Scrotal mass on admission.

The hepato-digestive examination revealed a fixed oval mass in the left iliac fossa, hard, painless with a regular surface, measuring 3x2.5 cm. There was no hepatomegaly or splenomegaly. The lymph nodes areas were free. The thoraco-abdominal CT-scan performed as part of the extension investigation (Figures 2A and 2B) revealed a large intra-scrotal mass measuring approximately 140x125x127 mm, having an heterogeneous multi-lobulated shape with tissue (42 HU) and necrotic (30 HU) components, richly vascularized septa after contrast agent injection that persisted in the late stage; and a mass in the left iliac fossa with multi-lobulated shape, measuring 60x54x54 mm, heterogeneous, with a hyper-vascularized tissue and necrotic components, with an enhancement kinetics similar to the intra-scrotal mass, latero-deviating the bladder without invading it. No thoracic or bony location was observed. Proof-reading of the slide by another pathologist was consistent with the initial diagnosis of a pleiomorphic rhabdomyosarcoma. The tumor was classified as stage II (AJCC Prognostic stages) and had a poor prognosis (location, size, initial resection and type). The child was hospitalized, given analgesics (Paracetamol) and benefited from a urine diversion cystostomy.



2A: Transverse section of scrotal mass.



2B: Sagittal section.

Figure 2: Thoraco-abdominal CT-scan.

An additional investigation revealed a normochromic anemia (hemoglobin=10 g/dl), a discrete hyperleukocytosis with neutrophilia and thrombocytosis (539 G/l). The kidney function and the blood electrolytes were normal. The evolution was marked by a progressive degradation of the general condition associating an infectious and anemic syndrome, a progressive increase in the volume of the scrotal mass with necrosis areas, the tumor size increasing to 22x17 cm against 17x13 cm at admission (Figure 3) and an extension of the mass towards the hypogastrium. After a multidisciplinary meeting involving the surgical team, the pediatric team including the onco-pediatrician, the radiologists and the

anesthetists, a surgical removal of the mass was indicated, followed by a chemotherapy and a radiotherapy.

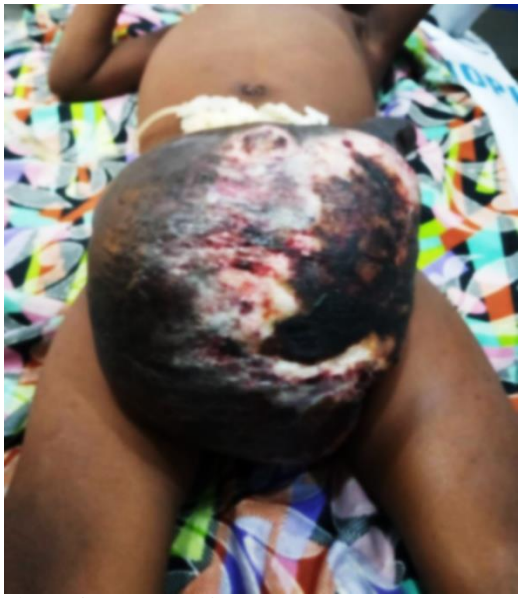
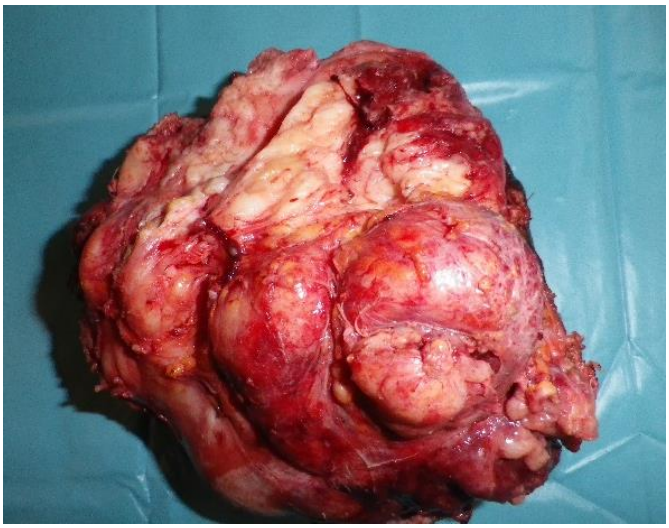


Figure 3: Evolution of scrotal mass at admission during hospitalization.

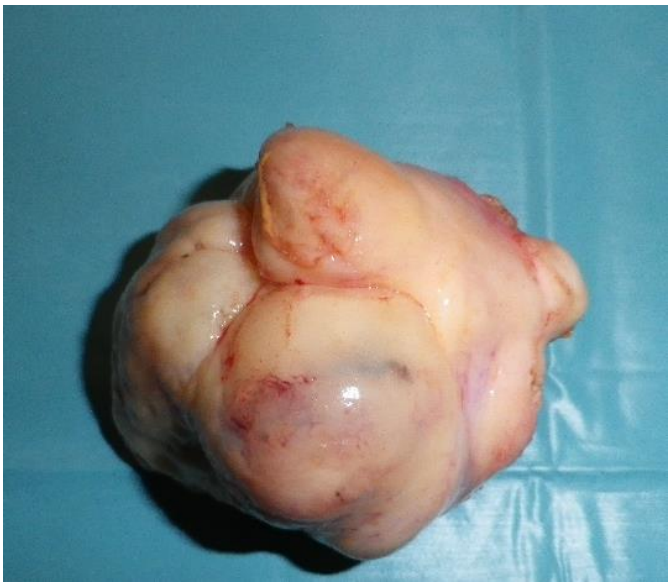
Per operatively, corpora cavernosa and spongiosa invasion was noted, and the urethra was catheterized with a CH8 urethro-vesical probe. Both were excised (Figures 4A and 4B). The hemostasis was satisfactory, and the patient had received two blood units' transfusions. The immediate postoperative incident was an irreversible cardiorespiratory arrest.

DISCUSSION

RMS develops from primitive mesenchymal cells that differentiate into striated muscle. It is rare in childhood and accounts for approximately 5% of pediatric malignancies (4). In our cohort, the proportion of RMS cases is 3.7%. This data is lower than those reported in the literature, but in our context, it only reflects hospital realities, given that many cases remain undiagnosed or not referred to hospitals for various reasons (5). It occurs in children between two and five years old and during adolescence (6). A case of paratesticular RMS in a 14 years old adolescent has been reported in Tunisia (7). Our child was six months old at the disease onset, which is early compared to peaks found in the literature (6). However, this child was seen in an adequate hospital structure at 30 months, that is, two years after the beginning of his disease. Paratesticular location accounts for 7-11% of childhood RMS and 10% of childhood testicular tumors (1–5). Other locations have been reported in studies from Africa and Europe: orbital, palpebral, parotid, auricle (2,8,9). Paratesticular localizations have been described in neonates, in a 23 years old adult and a 63 years old man (10–12). Paratesticular RMS develops from the mesenchymal tissues of the spermatic cord, the epididymis and testicular tunics. It was discovered in our patient following a painless intrascrotal mass. A cautious examination of an acute bursa was carried out and allowed to elimination of the main causes of large non-painful bursa, namely: hydrocele, cord cyst, epididymitis or inguino-scrotal hernia, which could lead to a detrimental diagnostic delay. The contribution of imaging is fundamental. At the initial evaluation, ultrasound was sufficient in studies done in Tunisia and France to suspect the diagnosis (1–7). The CT-scan was done to assess the extension as in our patient. Anatomopathological examination of the orchidectomy specimen confirms the diagnosis by specifying the histological type. There are essentially three histological types of rhabdomyosarcoma: the embryonic, the alveolar and the pleomorphic types. Their frequency varies with age. Embryonal rhabdomyosarcomas preferentially affect children under 10 years of age, alveolar rhabdomyosarcomas affect adolescents and pleomorphic RMS are found in adults over 45 years old. Embryonal subtypes are considered low-risk or standard-risk tumors, whereas alveolar are high-risk with a poorer prognosis (13). The pleiomorphic type seen in our patient had been reviewed by different pathologists who confirmed it. It is very rare in children and in the literature the cases observed were in adults (14).



4A: Scrotal.



4B: Abdominal.
Figure 4: Resected masses.

The management of RMS in children requires a multidisciplinary approach involving pediatric oncologist, pediatric surgeon, radiation therapist, radiologist and pathologist. Inguinal orchidectomy with high and first spermatic cord ligation is the standard treatment in localized forms (3). Treatment options after orchidectomy include

lumbo-aortic dissection, chemotherapy and radiotherapy. Multidrug chemotherapy is indicated in all cases. The treatment protocols duration varies from 18 to 24 months. These protocols include the VAC, IVA and VIE protocol (V: Vincristine; A: Actinomycin D; E: Etoposide; I: Ifosfamide and C: Cyclophosphamide). The combination of Vincristine, Actinomycin D and Cyclophosphamide is the most widely used and is based on the administration of several courses of treatment spread over five days and spaced two to four weeks apart (1). The therapeutic attitude in our patient was initially made of an ablation without orchidectomy, which did not correspond to the recommendations and would explain the early recurrence. Moreover, the recommended VAC chemotherapy protocol uses Actinomycin D and not Adriamycin as was the case before the referral. Adriamycin has a high cardiac toxicity, hence the interest in performing a cardiac investigation including heart ultrasound to evaluate the systolic ejection fraction. Also, the maximum cumulative dose not to be exceeded should be calculated and respected if the indication is made as in resistant forms. The unfavorable outcome in our patient could be explained by the initial non-adapted management of both surgery and chemotherapy, but also by the referral delay to an appropriate hospital with cardiac complications probably present. In our context of insufficient technical facilities, only an early detection, and an adapted imperative chemotherapy, would have enabled a durable remission in front of the paratesticular rhabdomyosarcoma.

CONCLUSION

Paratesticular RMS is a rare tumor which requires an early diagnosis, a precise assessment of extension, and a multidisciplinary management. In our context of insufficient technical resources, only a total removal of the tumor associated with an orchidectomy, and an imperative adapted chemotherapy, would have enabled a durable remission in front of the para-testicular rhabdomyosarcoma.

CONFLICTS OF INTEREST

The authors have declared no conflict of interest.

STATEMENTS

AS. Gbénou, G. Bognon, SF. Kunaba and M. Guédénon collected the data.
AS. Gbénou, G. Bognon, J. Akodjenou and E. Soho analyzed and interpreted the data.
AS. Gbénou, G. Bognon and SD. Kunaba wrote the manuscript.
AS. Gbénou, G. Bognon, J. Akodjenou and JM. Alao reviewed the manuscript.
All authors read and approved the final version of the manuscript.

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