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RESEARCH ARTICLE

SINGLE INSTITUTION STUDY ON THE MANAGEMENT OF CHILDHOOD BLADDER AND PROSTATE RHABDOMYOSARCOMA

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Abstract

Rhabdomyosarcoma (RMS) is the most common type of soft-tissue sarcoma seen in the pediatric population, but it is rarely located in the bladder. Treatment for RMS is multifaceted, often involving a team of specialists to ensure the best outcome for the patient. The prognosis for RMS varies depending on the type and stage of the cancer as well as the age of the patient. This study reports on 8 cases of bladder rhabdomyosarcoma that were treated at the National Institute of Oncology in Rabat between 2008 and 2014.Through a review of these cases and the literature, this article discusses the diagnosis and treatment of this rare type of cancer.

Introduction:-

Rhabdomyosarcoma (RMS) is a type of cancer that affects the cells that form the muscles. In children, RMS is the most common type of mesenchymal tumor, accounting for 60-70% of all mesenchymal tumors. However, it only represents 5% of all solid tumors in children (1,2). The genitourinary tract accounts for 20% of RMS cases in children (3). The incidence of RMS of the urinary bladder in Morocco is not well-established, but it is considered a rare condition and requires a multidisciplinary approach for treatment and management

It is typically treated with a combination of surgery, chemotherapy, and radiation therapy.

Treatment for RMS is multifaceted, often involving a team of specialists to ensure the best outcome for the patient. The prognosis for RMS varies depending on the type and stage of the cancer as well as the age of the patient.

Materials And Methods:-

This article reports on 8 cases of bladder rhabdomyosarcoma that were treated at the National Institute of Oncology in Rabat between 2008 and 2014. Through a review of these cases and the literature, this article discusses the diagnosis and treatment of this rare type of cancer.

Results:-

The patients in the study were diagnosed at an average age of 3 years, with a range of ages from 1 to 15 years. The time between the onset of symptoms and seeking medical help was 8 months on average. The majority of patients, 7 out of 8, were male. The diagnosis was confirmed by pathology examination. 62.5% of the patients had embryonic type of tumor and 37.5% had alveolar type. 88% of the patients' extension scan was normal, however, one patient

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had lung metastasis at the time of diagnosis. All patients were given chemotherapy as first-line treatment, using protocols CEV, IVE, IVA. Unfortunately, one patient did not benefit from surgery.
Radiotherapy was delivered to 7 patients at doses ranging between 50.4 and 54Gy. 80% of patients experienced chemotherapy side effects, dominated by hematologic toxicity (neutropenia), as well as vomiting, diarrhea, and mucitis of different grades. Radiotherapy was well tolerated, only radiodermitis of grade 1 or 2 were reported. The evaluation of treatment effectiveness was based on clinical and paraclinical data according to the RECIST system.

Complete response was noted in 2 patients, progression under chemotherapy in one patient, 2 local relapses and 3 distant relapses.

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Complete response was noted in 2 patients, progression under chemotherapy in one patient, 2 local relapses and 3 distant relapses.

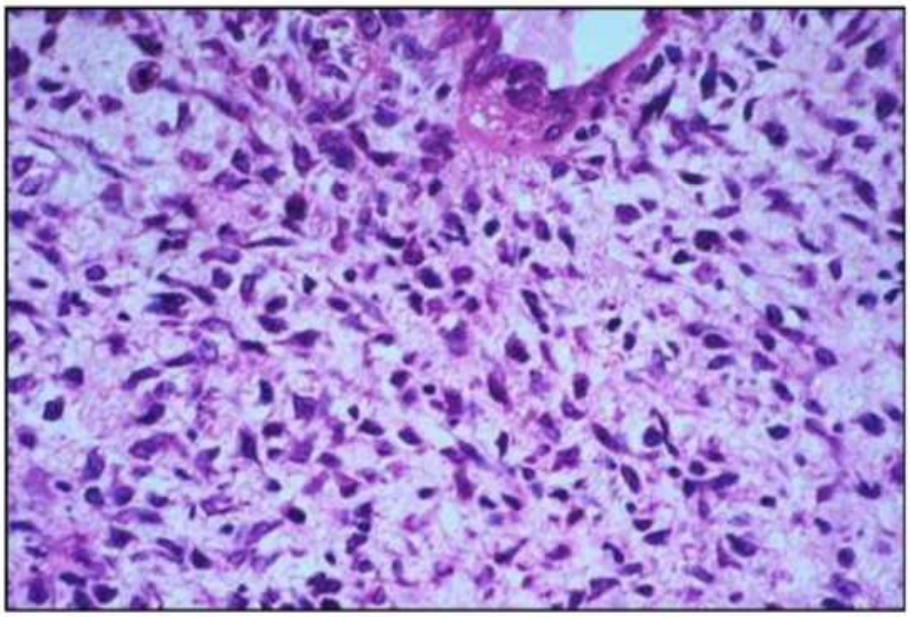
Discussion:-

Rhabdomyosarcoma (RMS) is the most common mesenchymal tumor in children and representing is the 3rd most common solid cancer in children, however it represents only 3% of all childhood tumors (4), it can occur in various locations in the body, but is more commonly found in the genitourinary tract, with the urinary bladder being one of the most common locations accounting for 20% of all cases.

Rhabdomyosarcoma is most commonly found in young boys, usually around the age of 2. This study showed that the majority of cases were boys, with an average age of 3 years. Clinical symptoms of RMS include urinary symptoms such as dysuria and hematuria, as well as a retroperitoneal mass, which were present in all patients in the study. Additionally, RMS can also cause acute urine retention in some cases.(5)

Anatomopathological examination shows that there are three main types of Rhabdomyosarcoma: Embryonic type (Figure 1), which is the most common and has a better prognosis, with an overall 5-year survival rate of 66%.

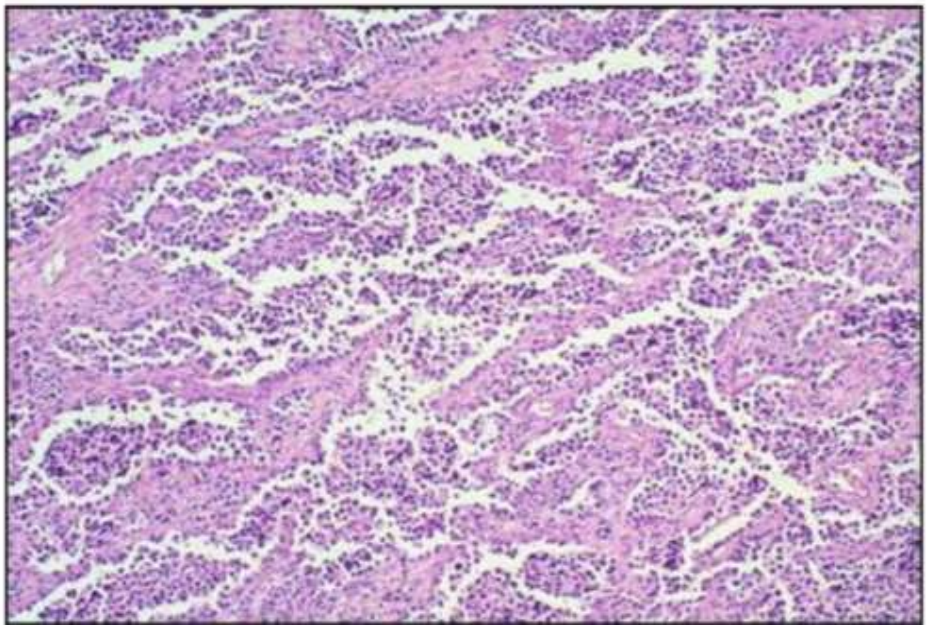
Figure 1:-



Microscopic appearance of an embryonic rhabdomyosarcoma (6)

Alveolar type (figure 2), which is more common in adults and tends to be more aggressive, with a worse prognosis and often diagnosed at a later stage, its overall survival for localized forms is 54%(7,8).

Figure 2:-



Microscopic appearance of an alveolar rhabdomyosarcoma(6)

The third type is pleomorphic, which is seen in both adults and children, but its incidence increases with age, and it has a poor prognosis. In this study, 62.5% of cases were of embryonic type and 37.5% were of alveolar type.

Additionally, studies have shown that pelvic scans reveal large nodules in clusters or intravesical mass, sometimes accompanied by hydronephrosis(figure 3).The majority of cases originate in the bladder base.(10)

Figure 3:-

Abdominopelvic CT: Hypervascularized intraperitoneal abdominopelvic tumor

Computed Tomography (CT) scans can be used to determine the extent of the disease using the TNM classification system for soft tissue sarcomas. (table 1)

In the study, CT scans showed that 50% of the patients had locally advanced tumors (stage III and IV).

Table 1:- TNM classification system for soft tissue sarcomas 2010 (11).

Primary tumor (T)	
TX:	Primary tumor cannot be assessed
T0:	No evidence of primary tumor
T1:	Tumor 5 cm or less in greatest dimension*
T1a:	Superficial tumor
T1b:	Deep tumor
T2:	Tumor more than 5 cm in greatest dimension*
T2a:	Superficial tumor
T2b:	Deep tumor

*Note: Superficial tumor is located exclusively above the superficial fascia without the invasion of the fascia; deep tumor is located either exclusively beneath the superficial fascia, superficial to the fascia with invasion of or through the fascia, or both superficial yet beneath the fascia

Regional lymph nodes (N)	
NX:	Regional lymph nodes cannot be assessed
N0:	No regional lymph node metastasis
N1**:	Regional lymph node metastasis

**Note: Presence of positive nodes (N1) in M0 tumors is considered Stage III

Distant metastasis (M)	
M0:	No distant metastasis
M1:	Distant metastasis
Anatomic stage/prognostic groups	
IA:	T1a N0 M0 G1, GX
T1b N0 M0 G1, GX	
IB:	T2a N0 M0 G1, GX
T2b	N0 M0 G1, GX
IIA:	T1a N0 M0 G2, G3
T1b	N0 M0 G2, G3
IIB:	T2a N0 M0 G2
T2b	N0 M0 G2
III:	T2a, T2b N0 M0 G3
Any T N1 M0 any G	
IV:	Any T any N M1 any G

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The treatment of Rhabdomyosarcoma requires a multi-disciplinary approach, including surgery, chemotherapy, and local treatment such as radiotherapy or additional surgery. Rhabdomyosarcoma is highly responsive to chemotherapy, with combination therapy being particularly effective, with a success rate of over 80% in most cases.(12)

The protocols used in the study varied depending on the time period of treatment. 75% of patients received chemotherapy using Protocol MMT95, while 25% were treated with Protocol MMT2005.

All patients received neoadjuvant chemotherapy, which consisted of 3 cycles with 21-day intervals. The main chemotherapy drugs used were Vincristine, Doxorubicin, Cyclophosphamide, Ifosfamide, Carboplatin, Epirubicin, Vincristine, Actinomycin D, and Etoposide, which were given at different doses and combinations depending on the patient's individual risk level.

The prognosis for RMS is generally poor due to the high risk of local relapse, so close monitoring and surveillance is recommended for these patients. The type of surveillance will depend on the initial treatment and is necessary to prevent long-term side effects as the patient grows.

Conclusion:-

Bladder RMS is a rare and serious type of cancer that requires a coordinated, multidisciplinary approach to ensure the best possible outcome. Early detection and treatment is key to improving the prognosis of RMS, and patients should be monitored closely for any potential side effects. With the right care, patients with bladder RMS can achieve long-term remission from this disease.

Conflicts of interest :

The authors declare no conflict of interest

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