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RESEARCH ARTICLE

MULTIMODAL TREATMENT IN PEDIATRIC ORBITAL RHABDOMYOSARCOMA: ABOUT 8 CASES

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Abstract

Rhabdomyosarcoma is the most frequent mesenchymal tumor in children, composed of cells with histopathological characteristics of striated muscle at different stages of embryogenesis. The orbital location accounts for about 10% of cases. Its prognosis has improved thanks to the progress of chemotherapy and new radiotherapy techniques.The aim of our study is to report the clinical and therapeutic characteristics of rhabdomyosarcomas of the orbit in children, and to present different complications of the treatment.

Introduction:-

Rhabdomyosarcoma is the most common primary orbital malignancy in children. It may arise primarily in the orbit, sinuses, or nasal cavity and subsequently spread to the orbit. 40% arise in the head and neck region, and 10% arise in the orbit. In the majority of published series, the orbit is the most favorable site, with a 5-year survival of over 85%[1]. The therapeutic management has greatly improved from primary exenteration to a more conservative multidisciplinary approach consisting of removal of the mass in combination with chemotherapy and local radiotherapy[2].

Patients And Methods:-

This is a retrospective descriptive study of 8 patients collected over a period of 5 years (from January 2015 to December 2019), in the Radiotherapy Department of the National Institute of Oncology in Rabat. The data collection was carried out with the help of an exploitation form by consulting the medical file of each patient.

Results:-

The median age at diagnosis was 8.5 years with extremes ranging from 3 to 16 years, and the mean time to consultation was 4 months. The sex ratio was 3M/1F. The symptomatology was dominated by unilateral exophthalmia, Palpebral swelling, and local inflammatory signs are reported in four cases,The extension work-up including orbital MRI, thoraco-abdomino-pelvic CT and BOM were performed in all patients and none of them presented distant metastases.

The diagnosis was confirmed by anatomopathological examination, and the embryonic histological type was found in all patients.

All patients received primary chemotherapy, the protocols used were CEV, IVE, IVA. 2 patients underwent surgery and radiotherapy was indicated in all patients at doses that varied between 50.4 and 54 Gy.

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With a median follow-up of 18 months, the evolution was marked by the achievement of a complete response in 6 patients, and 2 local relapses requiring exenteration.

To date, 6 patients have good locoregional control and one patient has developed pulmonary metastases requiring chemotherapy.

The late side effects were marked by the presence of exophthalmos in half of the patients, installation of a minor cataract in 3 patients and a major cataract for one patient requiring surgery,and ptosis in 4 patients.

Table 1:- Initial treatmentstrategy.

First-line treatment	Total number of patients
Chemotherapy	
Protocol treatment	
IVA-CEV-IVE	2
CEV-IVE	2
VAC	4
Total number of courses	
<6	3
6-9	5
Surgery	3
No surgery	5
Radiotherapy	8
Type of irradiation	
Externalradiotherapy	8

Discussion:-

Rhabdomyosarcoma develops from embryonic mesenchymal cells,its main locations include the head and neck (40%), genitourinary tract (20%), extremities (20%) and trunk (10%)[3].

For orbital localizations, 76% are located in the orbital cavity,12% are conjunctival, 9% intraocular, and only 3% eyelid[4].

Embryonalrhabdomyosarcoma is the most common variant in the head and neck region, including the orbit, which is consistent with the results in our series. The alveolar and botryoid histologic types are less frequent, and the pleomorphic type is extremely rare in the orbit.

The management of orbital rhabdomyosarcoma is multidisciplinary and include chemotherapy, surgery and local treatment by radiotherapy, but must be discussed in a multidisciplinary consultation meeting.

Rhabdomyosarcoma is a highly chemo-sensitive tumor. The protocols currently used combine three molecules when the tumor is localized: vincristine, actinomycin and cyclophos-phamide; vincristine, actinomycin and ifosfamide; vincristine, etoposide and ifosfamide. In our series, chemotherapy was started as soon as the biopsy results were available or just after tumor removal for the operated patients, 3 to 9 courses (average 6 courses), the protocols used were CAV in 4 patients, alternating CEV-IVE in 2 cases and IVA-CEV-IVE in 2 cases.

Radiotherapy was performed in all our patients, it was a conformal external radiotherapy with a dose ranging from 41.4 Gy for tumors with microscopic margins to 54.4 Gy for macroscopic tumor remnant following the IRS II protocol.

The main late side effects of radiotherapy were radiation cataract (55%), dry eye (36%), orbital hypoplasia (24%), ptosis (9%), and radiation retinopathy (90%) [5]. In our series, exophthalmos was noted in half of the patients, minor cataract in 3 patients and a major one requiring surgery, radiation retinopathy in 4 patients and ptosis in 2 patients.

Based on the IRS I trial[6], 50 patients received radiotherapy treatment at doses between 50 and 60 Gy and showed late effects with decreased visual acuity (80%), cataract (92%), orbital hypoplasia (52%) and facial asymmetry (41%). Nevertheless, total radiotherapy doses of more than 45 Gy are still used, and preliminary data from patients treated in the IRS III trial from 1984 to 1991 suggest that new radiotherapy techniques and fractionation do not have a significant positive impact on the occurrence of long-term side effects[7].

The experience of the four international collaborative groups (Intergroup Rhabdomyosarcoma Study Group [IRSG], International Society of Paediatric Oncology [SIOP] Sarcoma Committee, German Collaborative Soft Tissue Sarcoma Group [CWS], and Italian Coop- erative Soft Tissue Sarcoma Group [ICG] studies) does not showa clear conclusions on the efficacy, or otherwise, of reduced-dose radiation therapy. To date, the efficacy of doses below 40 Gy has not been proven and a significant reduction in adverse events is not expected. Indeed, cataract formation is reported at radiation doses below 10 Gy.

Since the introduction of multimodal treatment by chemotherapy, surgery and/or radiotherapy, the survival percentage of patients with rhabdomyosarcoma has improved significantly, with a 5-year survival of 94% for the embryonal form and 74% for the alveolar form[8]. In our series we noted a 2-year survival rate of 87.5% and a 2-year local recurrence rate of 12%.

Conclusion:-

Any rapidly evolving orbito-palpebral swelling in children should raise the suspicion of Rhabdomyosarcoma; a biopsy should be performed without delay because the prognosis is strongly related to the diagnostic and therapeutic delay. Multimodal treatment including intensive chemotherapy and radiotherapy has changed the prognosis of this disease with a 5-year survival of 86% to 95%.

Conflicts of interest:

The authors declare no conflicts of interest.

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