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Adult Orbital Rhabdomyosarcoma: Case Report

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Abstract:- Rhabdomyosarcoma is an extremely aggressive malignant tumor that rarely occurs in adults. Its orbital location represents only 9% of all head and neck injuries. We report the clinical case of an orbital rhabdomyosarcoma in adults. Observation: This is a 23 year old adult; of Moroccan nationality, which had presented a right exophthalmia of progressive evolution with cervical lymphadenopathy. The clinical examination had objectified a right exophthalmia and upper internal jugular lymphadenopathy; the rest of the clinical examination was without particularity. Computed tomography revealed a right intra-orbital tumor process. Histological analysis revealed an embryonic rhabdomyosarcoma with lymph node metastases. The combination of ifosfamide, vincristine and actinomycin chemotherapy and 66 Gy orbital radiation therapy using the intensity modulation technique (IMRT) resulted in complete remission with a 6-month setback. Conclusion: Rhabdomyosarcoma is a rare tumor. Early management improves the prognosis of life and function.

Keywords:- *Rhabdomyosarcoma, Orbital Tumor, Radiotherapy, Chemotherapy*

I. INTRODUCTION

Soft tissue sarcomas account for less than one percent of all solid malignant tumors in adults. Therefore, it is considered a rare cancer in adulthood [1]. The preferred sites are the head and neck (35%-40%) followed by the genito-urinary tract and extremities. Orbital rhabdomyosarcoma (RMS) 9% of all head and neck injuries [2]. It is an extremely aggressive malignant tumor whose early diagnosis significantly improves survival and visual prognosis [3]. We report the case of an orbital rhabdomyosarcoma in a 23-year-old male adult who had ophthalmitis associated with cervical lymphadenopathy.

II. CASE REPORT

This was a 23-year-old male patient with no significant history of pathology who had a progressive right exophthalmia with cervical mass one month prior to admission. The physical examination revealed: a patient WHO to 1 with mandibular and internal jugular lymphadenopathy without inflammatory signs in sight. There was a decrease in visual acuity in the right eye;

- The rest of the somatic examination was without particularity.
- A nasofibroscopy with cavum biopsy had been performed and was found to be normal.
- Lymphadenopathy biopsy was performed and histological and immunohistochemical examination was in favor of lymph node metastasis of an embryonic rhabdomyosarcoma.

A cervical facial CT in favor of a voluminous tissue mass, centered on the inner wall of the orbit, poorly limited, heterogeneously enhanced and moderately after contrast injection, 46 x 40 mm in height and 50 mm in height with broad locoregional extension to the sinus of the face, endocavitary and endocrinal associated with voluminous lateral cervical and homolateral supraclavicular lymphadenopathy (Figure 1).

Given the size of the tumor and the local functional risk, the surgery was recused. chemotherapy was started according to the RMS 2005 protocol, which consists of 5 cures combining ifosfamide, vincristine and actinomycin (IVA). A treatment response assessment was performed after three cures showing a clear regression of the right facial tumour process and secondary lymph node locations. (Figure 2)

External intensity modulation radiotherapy (IMRT) was indicated after chemotherapy, on the tumour residue, in this patient at a total dose of 66 Gy in 33 fractions of 2, with good clinical tolerance to external radiotherapy. (Figure 3)

III. DISCUSSION

RMS is considered a disease of young children, occurring at an average age of 8 years [4]. However, few cases of rhabdomyosarcoma have been described as adults [5,6,7]. It's a malignant soft tissue and skeletal muscle tumor. Most of the available data comes from retrospective case series, and indicates that the first typical presentation is ptosis [5], while other cases describe atypical presentations such as eyelid nodule [8]. Intraocular locations are exceptional and develop from the ciliary body or iris [4-9].

Imaging is not pathognomonic, but provides arguments for diagnosis [10]: tissue density of the lesion, enhancement after iodine or gadolinium injection, osteolysis aspects of the orbital walls. It makes it possible to visualize the lesion, to locate it in the orbit, sometimes to affirm its original structure,

to measure it, to identify its relationships and to specify its orbital or encephalic extension.

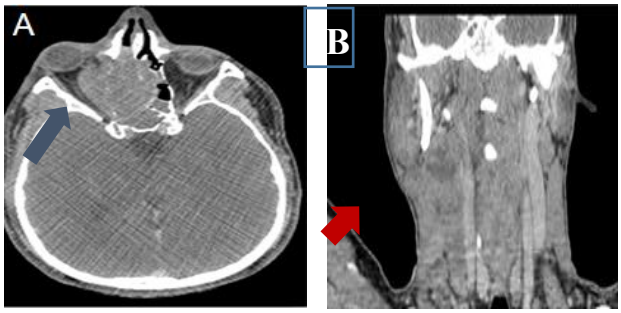

Figure 1: Axial (A) and coronal (B) slice with large tissue mass POC injection, centered on the inner wall of the orbit, poorly limited, heterogeneously and moderately enhanced after contrast injection, Measuring 46 x 40 mm and extended in height by 50 mm with broad locoregional extension to the sinus of the face, (blue arrows) and voluminous lateral cervical ADP and homolateral supraclavicular (red arrows).

From an histological point of view, rhabdomyosarcoma (RMS) is divided into three broad categories: embryonic, alveolar and pleomorphic. The contribution of immunohistochemistry is valuable in this type of tumor and helps to correct some erroneous diagnoses. Indeed, the distinction between rhabdomyosarcoma and other mesenchymal tumors is sometimes difficult especially when it comes to undifferentiated forms, as in this observation. [5,6,10].

Management, which may include chemotherapy, surgery, and/or radiotherapy, requires multidisciplinary consultation. The surgery is not systematic. The Intergroup Rhabdomyosarcoma Study Group I (IRSG I) gives priority to preserving the function over a complete resection [11]. Thus, due to the initial tumour volume and in order to avoid mutilating excision, it was decided not to use surgery in our patient. Rhabdomyosarcoma is a chemosensitive tumor. Current protocols combine three molecules when the tumour is not metastatic: vincristine, actinomycin and cyclophosphamide; vincristine, actinomycin and ifosfamide; vincristine, etoposide and ifosfamide. Superior efficacy of the combination of vincristine and actinomycin has been demonstrated [12]. Rhabdomyosarcoma is also a radiosensitive tumor, but requires high doses with no adverse effects. The main adverse events were radiation cataract (55%), dry eye (36%), orbital hypoplasia (24%), ptesis (9%), and radiation retinopathy (90%) [8]. In this child, irradiation was only initiated when imaging confirmed the existence of a tumor residue after 6 chemotherapy sessions. This technique does not alter patient survival and preserves a number of the adverse effects of radiotherapy [13]. Since the introduction of multimodal chemotherapy, surgery and/or radiotherapy, the survival rate of patients with rhabdomyosarcomas has improved significantly.

On the local level, a regression is observed in 80% of cases (20% of recurrences). At the regional level, no lymph node spread is observed in 94% of cases; 6% of cases develop lymphadenopathy as was the case of our patient. Finally, in general, 94% of patients do not have systemic diffusion compared to 6% of cases with visceral metastases. In total, 5-year survival is 94% embryonic and 74% alveolar [14]. Survival depends in particular on the metastatic nature of the disease. Added to this is the local functional risk, in particular of amblyopia and compression of the optic nerve: rhabdomyosarcoma is therefore a diagnostic and therapeutic emergency [12], especially since it is a tumour of brutal development [14, 15]. Differential diagnoses are numerous and are grouped into two categories: tumour and non-tumoral etiologies [10]. Tumor etiologies include cystic tumors (dermoid cysts, embryonic carcinomas), vascular tumors (capillary hemangioma or benign infant hemangioendothelioma, lymphangiomas), nerve tumors (orbital neurofibromas, plexiform neurofibroma, glioma of the optic nerve and chiasma), bone and cartilage tumors (fibrous dysplasia, aneurysm cysts, juvenile ossification fibroids, osteosarcomas, chondrosarcomas), histiocytic diseases (Langerhans cell histiocytosis or X histiocytosis, juvenile xanthogranuloma), orbital damage during hematology (lymphomas including Burkitt's lymphoma, tumor localization during leukemia), metastasis (neuroblastoma, Ewing's sarcoma), and finally, tumors propagated in orbit whose retinoblastoma is still important in countries where access to care is more difficult. Non-tumoral etiologies are mucoceles, meningocele, encephaloceles and microphtalmia with cysts, inflammatory pseudo-tumors, and infections (dacryocystitis, ethmoiditis, orbital cellulitis, orbital abscesses).

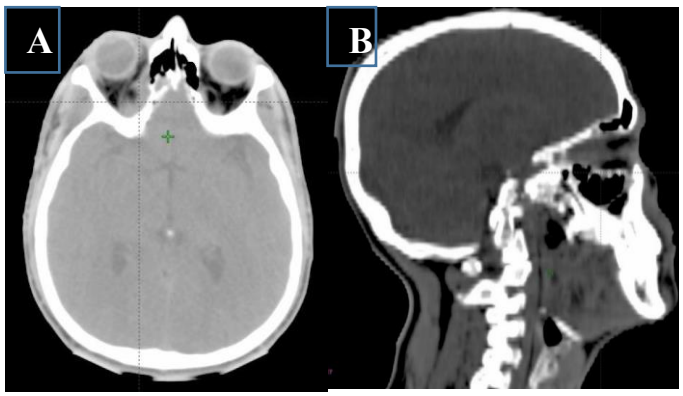

Figure 2 : axial (A) and Sagittal section (B): Sharp regression of the right facial process and lymphadenopathy

IV. CONCLUSION

This observation is atypical due to the occurrence of this tumour in adults; its orbital location, but also rhabdomyosarcoma is a diagnosis that must be considered in order to establish early management for a better vital and functional prognosis.

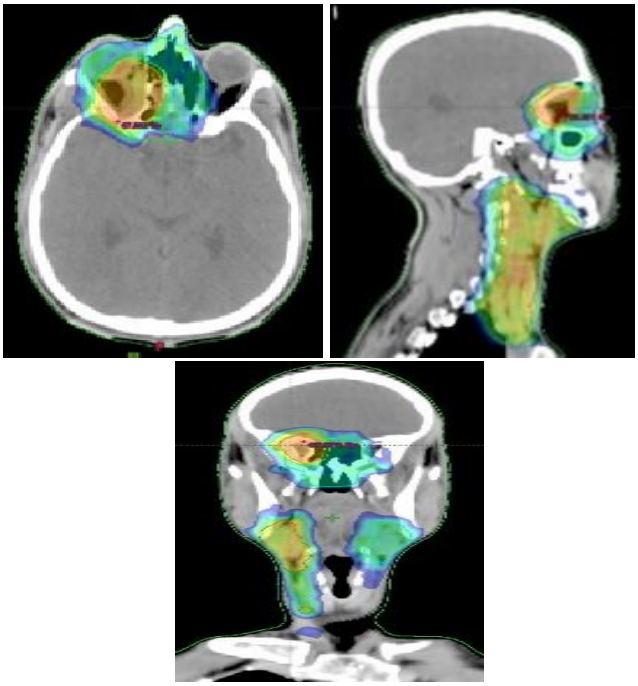

Figure 3: Dosimetric image of the tumour volume that received the total dose of 66 Gy

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