ISSN: 0975-833X

Available online at http://www.journalcra.com

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

International Journal of Current Research
Vol. 13, Issue, 06, pp.17853-17855, June, 2021

DOI: https://doi.org/10.24941/ijcr.41540.06.2021

RESEARCH ARTICLE **OPEN ACCESS**

ADULT ORBITAL RHABDOMYOSARCOMA, A RADIO-CHEMOSENSITIVETUMOUR: A CASE REPORT

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ARTICLE INFO

Article History:
Received 27th March, 2021
Received in revised form
15th April, 2021
Accepted 20th May, 2021
Published online 30th June, 2021

Key Words:
Orbital Rhabdomyosarcoma,
Radiotherapy, Chemotherapy.

ABSTRACT

Introduction: Rhabdomyosarcoma is an extremely aggressive malignant tumor that rarely occurs in adults. Its orbital location represents only 9% of all damage to the head and neck. **Case presentation:** We report the clinical case of orbital rhabdomyosarcoma in a 23-year-old male patient who presented with progressive right exophthalmos with cervical lymphadenopathy. The introduction of a treatment combining chemotherapy based on ifosfamide, vincristine and actinomycin, and orbital radiotherapy of 66 Gy according to the intensity modulation technique (IMRT), made it possible to obtain a complete remission with a 12 month follow-up. **Conclusion:** Orbital rhabdomyosarcoma is a rare tumor in adults. Early care improves the vital and functional prognosis.

Citation: Mahomed Yessoufou, KantaKa3 Dondo Mara, Axel Attolou, Amina Bouziane, Touria Bouhafa, MeriemBoubbou, Boucar Ndong and Khalid Hassouni. *"*Adult orbital rhabdomyosarcoma, a radio-chemosensitivetumour: A case report*", 2021. International Journal of Current Research, 13, (06), 17853-17855.*

INTRODUCTION

Tissue sarcoma accounts for less than 1% of all solid malignancies in adults. Therefore, it is considered a rare cancer in adults (1). The sites of predilection are the head and neck (35%-40%) followed by the genitourinary tract and the extremities. Orbital rhabdomyosarcoma (ORS) accounts for only 9% of all head and neck cancers (2). It is a highly aggressive malignancy whose early diagnosis significantly improves survival and visual prognosis (3). We report a case of orbital rhabdomyosarcoma in a 23-year-old male adult.

Case presentation: The patient was 23 years old, male, with no significant pathological history, who presented for one month with a right exophthalmos of progressive evolution with a cervical mass. The physical examination revealed: a patient WHO 1 with sub-angulo-mandibular and jugulo-carotid lymph nodes without inflammatory signs opposite. We noted a decrease in visual acuity in the right eye with an unremarkable somatic examination.

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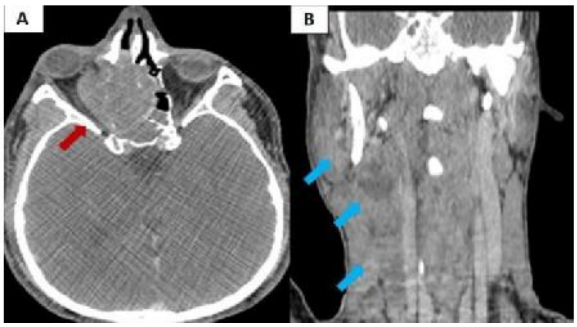
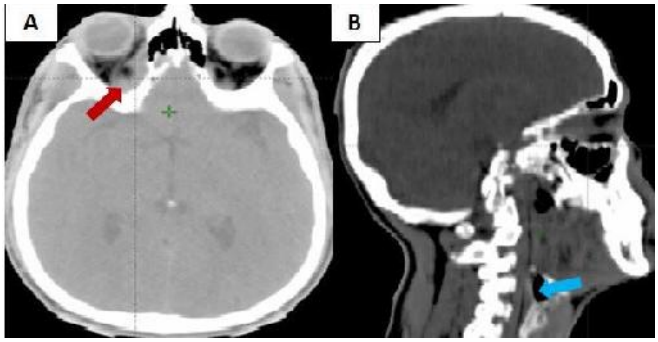
A nasofibroscopy of the cavum was performed and found to be normal. The biological work-up showed: correct renal function, normal liver function, haemoglobin 12.5g/dl, leucocytes 13,000/mm3, platelets 629,000/mm3, C-reactive protein 146mg/l and prothrombin 67%, alkaline phosphatase 2.5 times normal, albumin also normal and CPK 150 IU/l. A lymph node was biopsied and the histological examination and immunohistochemistry were in favor of a lymph node metastasis of an embryonal rhabdomyosarcoma. Cervical and facial CT scans showed a large tissue mass centered on the inner wall of the orbit, poorly limited, heterogeneously enhanced and moderate after injection of contrast medium, measuring 46 x 40 mm and extending over 50 mm in height, with wide loco regional extension to the facial sinus, endocavity and endocranium. It was associated with bulky laterocervical and supraclavicular homolaterallymph nodes (Figure 1). Given the size of the tumor and the local functional risk, surgery was rejected. The management adopted was chemotherapy according to the RMS 2005 protocol, which consists of 5 courses of ifosfamide, vincristine and actinomycin (VIA). An imaging check for response to treatment was performed after three courses showing clear regression of the right facial tumor process and secondary lymph node locations (Figure 2). Intensity modulated external beam radiation therapy (IMRT) was indicated after chemotherapy on the tumor residue in this patient at a total dose of 66 Gy in 33 fractions of 2 Gy,



➡: Large tissue mass, centered on the inner wall of the orbit, poorly limited, heterogeneously and moderately enhanced after injection of contrast medium, measuring 46 x 40 mm and extending in height over 50 mm with wide locoregional extension to the facial sinus.
➡ :Voluminous latero-cervical and supra-clavicularhomolateral lymph nodes.

Figure 1: Portal CT scan in axial (A) and coronal (B) sections of an adult patient with orbital rhabdomyosarcoma



➡ **:** Clear regression of the right orbital process
➡ **:**Clear regression of laterocervical lymph nodes

Figure 2. Axial (A) and sagittal (B) CT scan of an orbital rhabdomyosarcoma in an adult patient

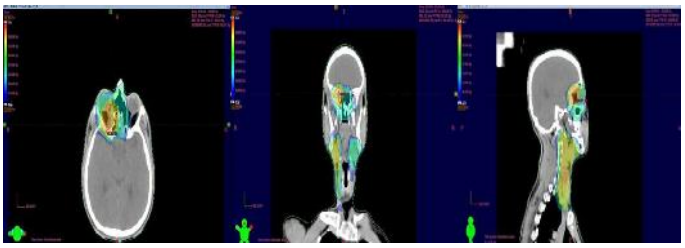


Figure 3. Dosimetric image of the tumor volume that received the total dose of 66 Gy in an adult patient with orbital rhabdomyosarcoma

with good clinical tolerance (Figure 3). After one year of follow-up, the patient showed no signs of recurrence.

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 in adults (5, 6, 7). It is a malignant tumor of soft tissue and skeletal muscle. Intraocular localizations are exceptional and arise from the ciliary body or iris (8). Most of the available data are from retrospective case series, and indicate that the first typical presentation is ptosis (5), while other cases describe atypical presentations such as eyelid nodule (9). Our patient presented with exophthalmos which is a fairly typical presentation of intraorbital masses but atypical of rhabdomyosarcoma. Imaging is not pathognomonic, but provides arguments in favour of the diagnosis: tissue density of the lesion, enhancement after iodine or gadolinium injection, aspects of osteolysis of the orbital walls.

It also allows the lesion to be located, the original structure to be confirmed, its volume to be assessed, its relationships to be identified, its orbital or encephalic extension to be specified and thus the pathway for the various therapeutic options to be determined (10). The main differential diagnoses on imaging are numerous and are grouped into two categories: tumor and non-tumor etiologies (10). However, the clinical context and histology guide the diagnosis. Histopathologically, rhabdomyosarcoma (RMS) can be divided into three main categories: embryonal, alveolar and pleomorphic. However, it is sometimes difficult to distinguish it from other mesenchymal tumors, especially in undifferentiated forms, as in this case. Hence the valuable contribution of immunohistochemistry in this type of tumor, which enables certain erroneous diagnoses to be made. Before discussing treatment, it should be remembered that orbital rhabdomyosarcoma is a diagnostic and therapeutic emergency because of the local functional risk, in particular amblyopia and optic nerve compression (11).

Management is multidisciplinary and may include chemotherapy, surgery, and/or radiotherapy. The Intergroup Rhabdomyosarcoma Study Group I (IRSG I) emphasizes preservation of function over complete resection (12). Rhabdomyosarcoma is a chemo sensitive tumor. Current protocols combine three molecules when the tumor 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 (11). Thus, given the initial tumor volume and extent, it was decided not to resort to surgery in our patient but rather to adjuvant chemotherapy and radiotherapy. Rhabdomyosarcoma is also a radiation-sensitive tumor, but requires high doses that cause side effects. The main side effects are radiation cataract (55%), dry eye (36%), orbital hypoplasia (24%), ptosis (9%), and radiation retinopathy (90%) (9). In order to minimize its adverse effects, our patient's irradiation was only undertaken when imaging confirmed the existence of a tumor residue after 5 courses of ifosfamide, vincristine and actinomycin (VIA). However, since the introduction of multimodal treatment with chemotherapy, surgery and/or radiotherapy, the survival of patients with rhabdomyosarcoma has improved significantly. Locally, regression is seen in 80% of cases (20% recurrence). Regionally, there was no lymph node dissemination in 94% of cases; lymph nodes appeared in 6% of cases. Finally, at the general level, 94% of the patients did not show any systemic spread, while visceral metastases appeared in 6% of cases. Overall, the 5-year survival rate is 94% for the embryonal form and 74% for the alveolar form (14). Survival depends in particular on the non-metastatic nature of the disease. The evolution of our patient after chemotherapy and external radiotherapy is favorable with a clinical and radiological remission after 12 months of follow-up.

CONCLUSION

Orbital rhabdomyosarcoma is a rare tumor in adults with a functional prognosis due to amblyopia and optic nerve compression. However, its diagnosis and management must be early because of its chemo- and radiosensitive nature as well as its low metastatic potential.

Abbreviations

RMS: rhabdomyosarcoma
IMRT: intensity modulated radiation therapy
Gy: Gray
IVA: ifosfamide, vincristine, actinomycine
IRSG I: Intergroup Rhabdomyosarcoma Study Group I

Conflicts of interest: None

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