ORIGINAL RESEARCH PAPER

Oncology

CASE REPORT: TREATMENT OF PLEOMORPHIC RHABDOMYOSARCOMA IN ADULT

KEY WORDS:

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ABSTRACT

SUMMARY:

Rhabdomyosarcoma, a malignant tumor of the striated muscle, is highly variable in terms of frequency and type according to age, the presence of this tumor in adults being even rarer. The present case is about oncological management of pleomorphic rhabdomyosarcoma in adults with ICE scheme, obtaining a good oncological response and can be considered a good scheme for tumor relapse.

INTRODUCTION

Any malignant tumor originating from mesenchymal cells is called sarcoma. Mesenchymal cells normally mature to differentiate into skeletal muscle, smooth muscle, adipose tissue, connective tissue, bone, and cartilage. The term rhabdomyosarcoma (RMS) defines a tumor originating from immature mesenchymal cells that differentiate into skeletal muscle despite the fact that it often originates from sites where it is not ordinarily formed (for example: bladder)

Rhabdomyosarcoma, a malignant tumor of the striated muscle, is highly variable in terms of frequency and type according to age.

Due to its histology, it can be subdivided into 4 main forms: embryonic (60%), botryoid (botryoid sarcoma), which reaches 6%, alveolar (20%) and pleomorphic (1%), in total it accounts for about 3.5% of all childhood cancers.

Some tumors have mixed features. The embryonic and alveolar forms occur in children and adolescents and are among the most common malignant tumors in these stages.

The classic pleomorphic form occurs in adults, mainly in the extremities and is less common than the others. It usually forms inside a muscle and can also affect the skin.

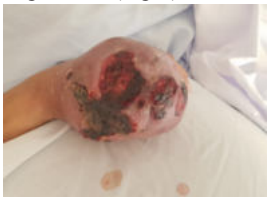
Rhabdomyosarcoma is one of the most malignant soft tissue sarcomas, with standard radical local resection or amputation; likewise, the 5-year survival rate is 30% in children and 40-50% in adults. In both groups, treatment is similar and consists of radical resection or its presumed equivalent (isolated perfusion of the limb, plus local resection or local resection followed by radiotherapy) to obtain local control, in addition to adjuvant chemotherapy with multiple drugs for the systemic control.

The present case is a relapse of rhabdomyosarcoma with transformation of alveolar in childhood to pleomorphic in adulthood with response to ICE treatment (ifosfamide, carboplatin,VP-16)

CLINICAL CASE

A 30-year-old patient with a diagnosis of right hand hypothenar alveolar rhabdomyosarcoma treated in pediatrics at the age of 10 years received chemotherapy scheme VAC (vincritine, actinomycin D, cyclophosphamide) subsequent local resection of the tumor, the histopathological report reported alveolar rhabdomyosarcoma (3 cm free edges, 40 % necrosis) subsequently received radiotherapy 41.4 g and to the surgical bed and continued with weekly chemotherapy according to the pediatric protocol until 102 cycles were completed, finally passing to controls.

In mid-2020, he attended due to the presence of a dependent tumor mass in the right hand (Fig-1).



Right-hand magnetic resonance imaging reported tumor lesion in the hypothenar region with bone destruction of the fifth metacarpal and tumor extension to adjacent soft tissues, no involvement of the proximal or distal joint, displacement of the flexor and extensor tendons of the distal phalanges.

The chest tomography reported a peripheral nodule in the left lung of (7.7mm). initially suggestive of metastasis.

The right hand tumor bed biopsy reported pleomorphic rhabdomyosarcoma.

This is a patient with a diagnosis of pleomorphic rhabdomyosarcoma (previously alveolar), with a good response to treatment, an ICE scheme was proposed, with which he received 4 cycles detailing clinical evolution favorable to treatment. As shown in the figures; (fig2 a / b, after the first treatment cycle) fig3 after the fourth treatment cycle.

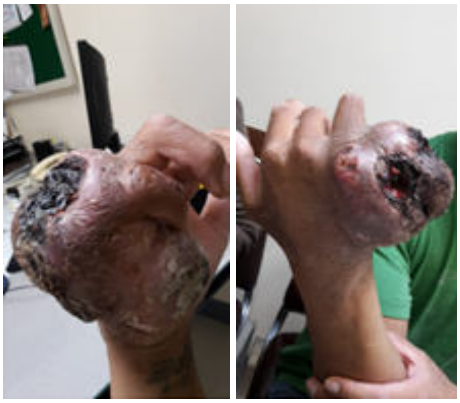
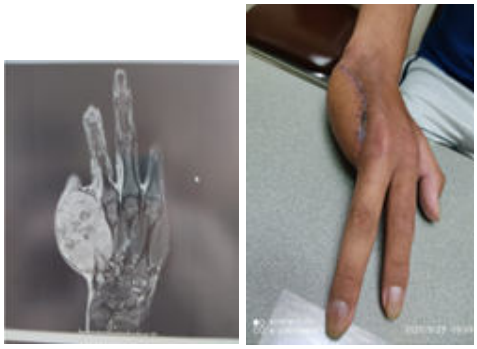


Figure 2 (a / b)

Fig 3

After chemotherapy, he went to surgery, due to bone destruction amputation was necessary.



The post-intervention patient has had a disease-free period of 6 months to date.

DISCUSSION

The first description of rhabdomyosarcoma was made by Weber in 1854, however, its "definitive" publication is attributed to Stout in 1946, 92 years later.

Rhabdomyosarcoma is very rare in adults. Five "large" series have been published that gather a total of approximately 400 cases of RMS in adults (including some cases in "children") evaluated in reference cancer centers in the United States and Europe over the last 20-30 years.

In patients with metastatic tumors, the most recently completed clinical trial attempted to assess the antitumor activity and actual therapeutic efficacy of one of three drug pairs (ifosfamide plus doxorubicin, vincristine plus melphalan, and ifosfamide plus etoposide) added to conventional chemotherapy.

Most patients with RMS receive chemotherapy treatments lasting 6 to 12 months (rarely longer, although depending on the severity of the side effects, treatment designed to last 10 months can sometimes be extended to 15 months). Chemotherapy is usually given in 2 to 5 (sometimes 10) "pulses" or "cycles" daily every 3-4 weeks. Some chemotherapy drugs can be given weekly.

The prognosis for children with recurrent / refractory sarcomas is poor. We determined the overall response rate (ORR) and overall survival (OS) of children with recurrent / refractory sarcomas who received ifosfamide, carboplatin, and etoposide (ICE) in three Children's Cancer Group (CCG) phase I / II trials.

For Veit Bucklein, patients with localized recurrence of soft tissue sarcoma (STS) after anthracycline-based chemotherapy have a poor prognosis, particularly when surgery is not possible. To facilitate resection and improve long-term tumor control, an intensified perioperative treatment consisting of ICE (ifosfamide 6 g / m 2, carboplatin 400 mg / m 2 and etoposide 600 mg / m 2) was applied in combination with regional hyperthermia (RHT) to maximize local control.

In conclusion, in this case we had a good response to treatment with ICE after the patient had received treatment in childhood, so we indicate that a treatment possibility.

The authors declare that they have no conflict of interest.

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