Role of neoadjuvant chemotherapy in reducing surgical morbidity in a young child with a rare primary skull base Ewing's Sarcoma

GUSTAVO SIMIANO JUNG, VICTOR FRANDOLOSO, ERASMO BARROS DA SILVA JR, GIULIA XAVIER BORNANCIN, RICARDO RAMINA

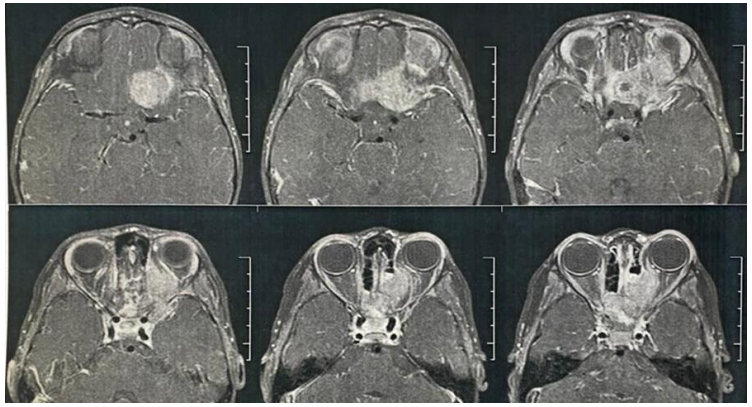
Instituição: Instituto de Neurologia de Curitiba - INC

Introduction:

Primary bone tumor account for 5% of all child and adolescent cancer. Ewing's Sarcoma is an aggressive form of childhood cancer that is the second most common bone tumor in this population. Skull base primary Ewing's Sarcoma is very rare and account for only 1% of all Ewing's Sarcoma. Before chemotherapy was introduce, about 10% of patients with Ewing's sarcoma survived. Progress since then has been dramatic, with 75% of patients with localised tumours now surviving. Achieve free surgical margins can be challenging in skull base ES specially when the orbit and, specially the periorbital tissue is involved. We present the first case, of a skull base Ewing's sarcoma treated with neoadjuvant chemotherapy to reduce surgical morbidity without compromising the surgical radicality,

The case

We present a case of a two year of age female with an anterior skull base Ewing's Sarcoma extending into the nasal cavity, left orbit and periorbital tissue. As presenting symptoms, she had fast progressing proptosis and decreasing visual acuity. A transfacial approach with orbit exenteration was proposed elsewhere. We decided to perform an endonasal-transethmoidal biopsy that confirmed primary skull base Ewing's Sarcoma positive to CD99 and EWSR1 rearrangement. The patient was submitted to "off-label" neoadjuvant chemotherapy alternating vincristine, adriamicine, ciclosfosfamide, ifosfamide, etoposide and vimblastine. After 6 cycles of chemotherapy, a very significant reduction in tumor volume was seen and a small residual lesion was located in the cribriform plate.



MRI before first 6 cycles of chemotherapy

Aiming local control, the patient was submitted to a combined (endonasal and transcranial) approach with extensive removal of the anterior skull base, dura mater, olfactory bulb and additional decompression of the left orbit and optic nerve. Pathology was positive for Ewing's sarcoma in the cribiform plate sample but free margins at twas confirmed

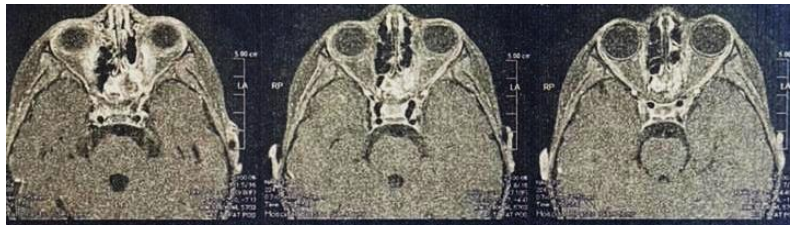
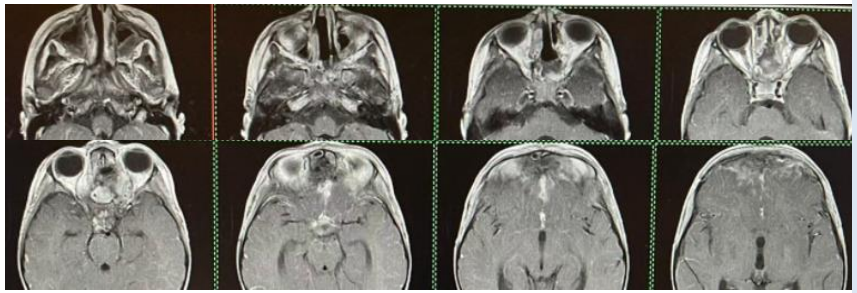


Image after first 6 cycles of chemotherapy (pre operative)

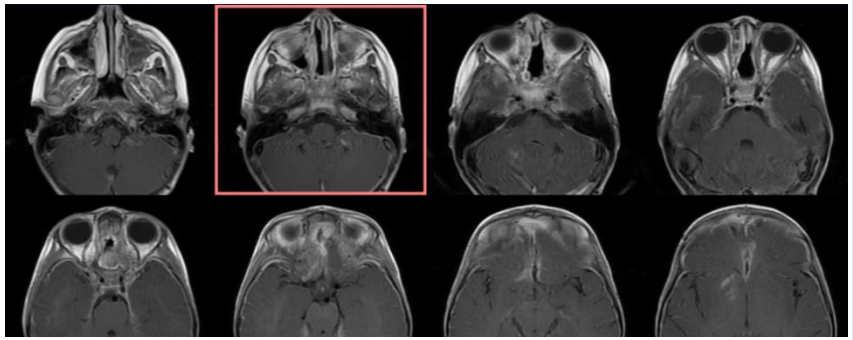
The patient completed 8 more cycles of chemotherapy and radiotherapy was precluded. She is one year free of disease presenting normal vision and only anosmia as permanent neurological deficits.



Post operative MRI.

Final comments

In this case we demonstrate the potencial benefit of neoadjuvant chemotherapy for a large fast growing Ewing's sarcoma, and its importance in reducing the surgical morbidity and mortality. No standard treatment are current available for primary skull base Ewing's sarcoma but similar to sinonasal undifferentiated carcinoma, neoadjuvant chemotherapy may play an important role in managing this challenge patients.



Control MRI aftter 8 cycles of adjuvant chemotherapy

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