METASTATIC MEDULLOEPITHELIOMA : 2 CASES-REPORT AT NATIONAL CANCER CENTER – BRAZIL

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Background

Medulloepithelioma is a rare embryonal neoplasm which presents in childhood as ciliary body mass and rarely tends to be locally aggressive and metastatic.

Methods

During the period of 1999 to 2018 two patients were admitted with medulloepitelioma at the National Cancer Institute, Brazil. Our aim was to perform a retrospective chart review to describe their presentation, treatment and outcome

Results

Patient1

A two-year-old girl with one year history of leukocoria, growing right eyeball mass and ipsilateral submandibular mass. Biopsy revealed medulloephitellioma (Figure 1). Enucleation showed intraocular lesion and neck dissection revealed metastatic submandibular gland and no lymph node infiltration. The patient received adjuvant chemotherapy with vincristine, ifosfamide, cisplatin, cyclosphosphamide and etoposide and local treatment with radiotherapy - 46Gy. The patient is alive without evidence of disease with a follow up of 216 months.

Patient2

An eight year-old female with 6 year history of inadvertent evisceration for blind painful eye. Six years later, she presented with large orbital mass and left cervical lymph node enlargement (Figure2). MRI of the orbits showed left extraocular tumor and large metastatic nodal mass to the parotid region (Figure 3). Chemotherapy consisted of ifosfamide, carboplatin and etoposide with partial regression of the extraocular extension. Modified exenteration, neck dissection and superficial parotidectomy revealed medulloepithelioma with scleral, extraocular soft tissue and optic nerve edge invasion(Figure 4). In 11 of 37 nodes showed metastatic deposits. orbital, parotid gland and left cervical radiotherapy (50.4 Gy) was performed. The patient is alive, disease-free two months after treatment(Figure 5).

In both patients it was observed diagnostic delay. They had good outcome, although the number of patients is too small, and in one patient the follow up is still short.

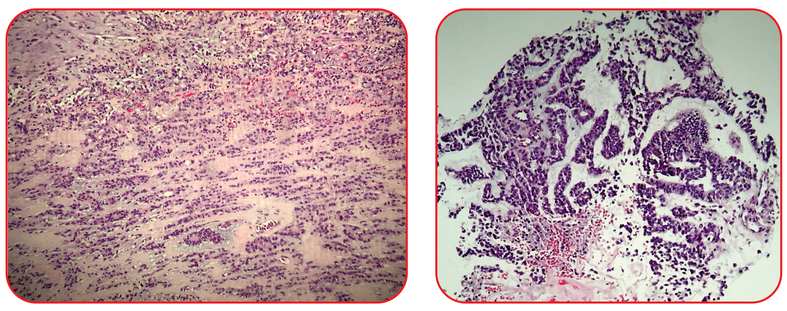

Figure1: Malignant Medulloepithelioma with metastatic lymph node

Figure 2 : Initial orbital and cervical mass at diagnosis

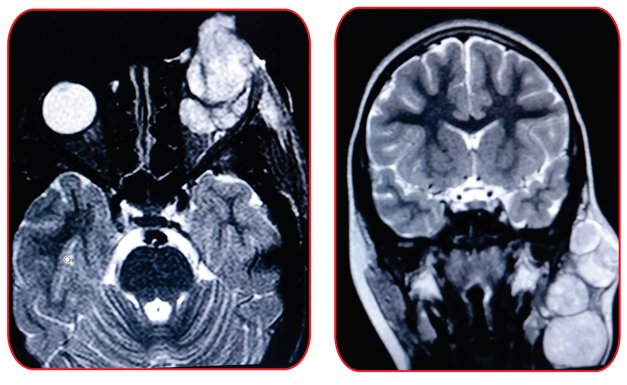

Figure 3: MRI images (sagittal and coronal sections)

Figure 4 : Surgical procedure of the cervical region and the orbit(images authorized by informed consent)

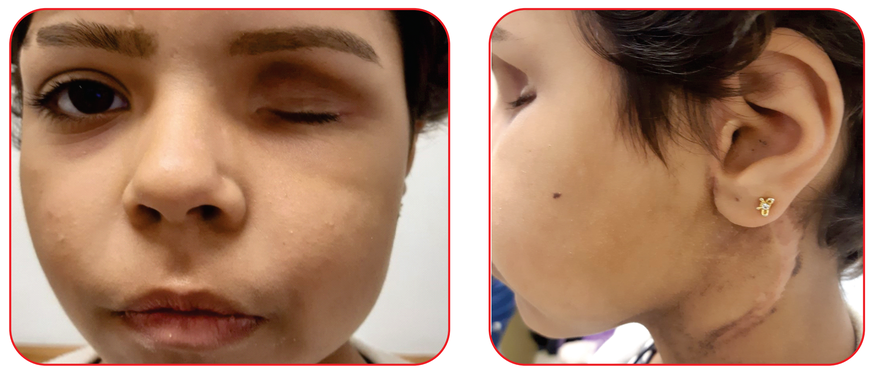

Figure 5: At the end of treatment (images authorized by informed consent)

Conclusions

Metastatic intraocular medulloepithelioma is a very rare disease and no standard treatment is available. Both cases highlight the importance of maintaining a high clinical index of suspicion for an unilateral intraocular tumor avoiding diagnostic delay.

Projeto Gráfico: Área de Edição e Produção de Materiais Técnico-Científicos / INCA