# EXTRA NERVOUS SYSTEM METASTASIS DUE TO MIDLINE DIFFUSE GLIOMA: CASE REPORT AND LITERATURE REVIEW

Presenter: THOMAS MEDUNECKAS TOURINHO

Authors: THOMAS MEDUNECKAS TOURINHO, MARCOS DEVANIR SILVA DA COSTA, PATRÍCIA ALESSANDRA DASTOLI, TALITA HELENA MARTINS SARTI, EMMANUEL DE OLIVEIRA SAMPAIO VASCONCELOS SÁ, JARDEL MENDONÇA NICACIO, ANDREA CAPPELLANO, NASJLA SABÁ SILVA, SÉRGIO CAVALHEIRO Neurosurgery, Universidade Federal de São Paulo - Escola Paulista de Medicina



# **CASE PRESENTATION**

**HISTORY** A 7-year-old girl who has a progressively worsening headache and vomits in the last 20 days. She complained about low visual acuity but the ophthalmological exam did not show any alterations. No other complaints.

**PAST** Nothing significant.

**PHYSICAL EXAM** Nothing Significant.



*Initial MRI showing a diffusely expanded pons compatible with Diffuse Intrinsic Pontine Glioma (DIPG)* 



Initial MRI showing a diffusely expanded pons compatible with Diffuse Intrinsic Pontine Glioma (DIPG)



A magnetic resonance imaging (MRI) revealed a diffusely expanded pons compatible with Diffuse Intrinsic Pontine Glioma (DIPG).

#### TREATMENT

**Stereotactic Biopsy** 

Radiotherapy

#### DIAGNOSIS

Histological Analysis: Astrocytoma

#### ImmunoHistochemistry: p53 negative Ki 67 10-15%

NGS: HIST1H3B c.83A>T; p.Lys28Met(6p22.2)

# OUTCOME

The patient followed up with progressive neurological deterioration. A new MRI showed hyperintense lesions on midbrain, both thalamus and cortex of cerebrum.

Another treatment with radiation therapy and chemotherapy was performed.



MRI showing the progression of the disease



MRI showing the progression of the disease

#### OUTCOME

During an outpatient consultation, a new MRI was evaluated, which revealed the presence of multiple lesions in the spine, ischio and sternum.



MRI revealing the presence of multiple lesions in the spine, ischio and sternum

# OUTCOME

It was performed a transpedicular biopsy guided by CT in one of this vertebral lesions.

Histopathological analysis revealed that they were metastasis due to glial neoplasia.

## DISCUSSION

DIPG is an aggressive tumor of the brainstem and extraneural metastases are rarely reported and poorly characterized. DIPG most commonly affects the pediatric population. The mean age at diagnosis are 6-7 years. It represents approximately 20% of all pediatric CNS tumors (1). Unfortunately, DIPG has a poor prognosis, and the median survival is generally less than 1 year independently from the treatment received.

# DISCUSSION

These tumors are not surgically resectable due to their anatomic location, which limits tissue available for diagnosis and molecular study. However, recent studies have revealed molecular characteristics of diffuse midline gliomas that are distinct from hemispheric pediatric and adult gliomas (2, 3).

# **FINAL COMMENTS**

Patients with appropriate age and typical image in MRI of DIPG allow treatment without biopsy, but in reference centers the histopathological and genetic study warrant the development of other treatments. Despite of all the development in neurosurgical technology, DIPG continues to be a disease with small progress in overall survival. This uncommon presentation of metastatic dissemination of midline diffuse glioma is important to be presented due to the need to better understand its behavior.

#### REFERENCES

- Perrone MG, Ruggiero A, Centonze A, Carrieri A, Ferorelli S, Scilimati A. Diffuse Intrinsic Pontine Glioma (DIPG): Breakthrough and Clinical Perspective. Curr Med Chem. 2021;28(17):3287-3317. doi: 10.2174/0929867327666200806110206. PMID: 32767913.
- Huang, T.Y., Piunti, A., Lulla, R.R. et al. Detection of Histone H3 mutations in cerebrospinal fluid-derived tumor DNA from children with diffuse midline glioma. acta neuropathol commun 5, 28 (2017). https://doi.org/10.1186/s40478-017-0436-6
- Castel, D., Philippe, C., Calmon, R. et al. Histone H3F3A and HIST1H3B K27M mutations define two subgroups of diffuse intrinsic pontine gliomas with different prognosis and phenotypes. Acta Neuropathol 130, 815–827 (2015). https://doi.org/10.1007/s00401-015-1478-0